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Marie-Struempell disease : with special reference to early diagnosis

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MARIE-STRUMPELL DISEASE

WITH

SPECIAL REFERENCE TO EARLY DIAGNOSIS

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INTRODUCTION

The idea for this particular problem presented itself to me several years ago after having suffered from considerable pain, progressive crippling of posture, and resultant delay in obtaining my education. This disabling disease continued over many years with periods of remissions and exacerbations but with no positive diagnosis made or any effective treatment found to stop its course or even to decrease the misery. Finally after six years of agony, loss of much school time, and almost permanent crippling, a diagnosis was made of Marie-Strumpell disease and corrective measures taken which fortunately turned out favorable. After going through so much suffering myself, I began to wonder if an earlier diagnosis couldn't be made and thus alleviate the many years of suffering for patients in the future. In this thesis an attempt will be made by examination of the literature, case histories and personal observation to seek a method of reaching an earlier diagnosis in Marie-Strumpell disease and substantiate its acceptibility.

Before one can attempt to solve any problem, he must first have a working knowledge of that problem and understand all of its intricacies. Therefore a brief presentation will be made of what is known of the disease up to the present time.

HISTORY AND NOMENCLATURE

The pathological process in spondylitis involves articular bone and any alteration in bone structure will leave a permanent record of the disease as long as the skeleton or casts of it remain; for this reason spondylitis was found to be among the oldest of known diseases.

It is known that spondylitis can attack animals as well as man. Sir Armand Ruffer found evidence in the skeletons of a dinosaur and a crocodile from the lower Miocene period that these animals had been severely crippled by spondylitis. Spondylitis was apparently very common among the Egyptians as mummies and skeletons dating as early as 5000 B.C. showed evidence of the typical spinal changes of this disease. In two tombs at Kom-El there were found four skeletons that exhibited spondylitis deformans. Even the sacred monkeys of Thebes did not escape. (1) It was noted in reading that this particular type of spinal arthritis was designated variously as: Marie-Strumpell arthritis, Strumpell-Marie's disease, Bechterew's disease, ankylosing spondylitis, spondylose rhizomelic, spondylitis ankylopoetica and spondylitis adolescens, differentiating it from other types of arthritis.

The first accurate account of the disease was considered by many writers to be that of one O'Connor in 1691 who gave a very lucid description of the condition

known as ankylosing spondylitis. Rudolf Virchow suggested the term arthritis deformans for cases of ankylosing spondylarthritis in the early part of the past century. The Russian neurologist Vladimir Mikhailovich von Bechterew in 1893, from observation of 500 cases of spinal rigidity, noted several different characteristics from the original entity and thought them to be worthy of classification as a separate disease which now bears his name. His type of spinal arthritis was one in which there was a progressive stiffening of the vertebral column beginning in the thoracic region and extending downward. There was marked thoracic kyphosis which was accompanied by signs and symptoms of nerve involvement as evidenced by paralysis and atrophy of the muscles of the back, more marked in the upper portions. There was also found to be diminished sensation in the cutaneous nerves, reflex changes, and psychic involvement. Bechterew thought that the other joints of the body were uninvolved. (2)

In 1897 Adolf von Strumpell and in 1898 Pierre Marie described a somewhat different form of ankylosing spondylitis which has come to be known as Marie-Strumpell disease. Their findings first manifested themselves in the lower spine and then progressed dorsalward. There was usually prostration with the patient being confined to bed generally in severe pain. Stiffness progresses rapidly and the

pathologic changes which start in the lower back, soon rather uniformly involve all the portions of the spine. Marie believed the ankylosing process to be limited to the joints of the spine and the roots of the limbs which led him to designate the affliction spondylose rhizomelic. He thought the other joints of the body to be uninvolved in the process. (2)

The term ankylosing spondylitis designates inflammation of the vertebrae resulting in abnormal immobility and consolidation of the joints. Spondylitis ankylopietica refers to the poker spine found as the end result of this malady. This disease in a vast majority of instances is found in the young and otherwise healthy males which lead to its being entitled spondylitis adolescens, a term suggested by S. Gilbert Scott.

In 1925 R. Lawford Knaggs from his work in pathological anatomy added two more descriptive names to the already long list. He called Bechterew's disease spondylitis muscularis due to the muscular atrophy which is present. He and other workers believed that this condition depends first upon the impaired muscular power primary to the changes in the vertebrae. Knaggs classified the Marie-Strumpell type of spondylitis as spondylitis ossificans ligamentosa. He thought the characteristic lesion to be the ossification of the spinal ligaments which is more or

less general in this type. (2, 3)

From the nomenclature it is seen that this particular type of spinal arthritis is known by many titles, also that most of the terminations have a reference to ankylosis. It is in the final stages of this disease, however, that either bony or fibrous ankylosis of the spinal column appears. Thus it appears that the term merely stresses the unfortunate fact of medicine's helplessness to prevent this unfortunate terminal complication. As one who has suffered this disease I wondered why I had to go through so many years of pain with no relief nor a definite diagnosis from what I might be suffering. However after reading the literature on the subject I was no longer surprised as most of the articles seemed to be limiated to a full and detailed description of the disease only in its final stages. None of them gave any sign by which the condition could be recognized at its onset or roentgenographs which might demonstrate the early stages of ankylosis of the sacroiliac joints, a condition which is always present in the late stages.

ETIOLOGICAL ASPECTS

The question of the etiology of ankylosing spondylitis has not been too satisfactorily settled. It is agreed by most authors that the disease is infectious in nature, but the causative agent or its method of infecting is still unsettled.

Bechterew believed the main etiologic factor to be an underlying hereditary predisposition with trauma as the exciting cause of the disease; this trauma being ineffective in producing the disease without the presence of such a hereditary tendency. Marie considered his disease to be not directly infectious but probably of toxic origin resulting from the infection and thought that the Neisserian infection might have some role in the cause. He also indicated that there might be the possibility of a nutritional basis for the disease.(2)

V. A. Oppel believes that in a given disease manifested by arthritic changes the spinal manifestations are mere incidents and that the same physical and chemical factors are operative in the selection of the spine as the site of the involvement as of other parts of the body. He is of the opinion that of these factors; trauma, occupational posture, strain, or pre-existing local infection may play a part. Arthur Streindler holds the view that spondylarthritis is a systemic disease with the spinal symptoms as merely the cardinal symptoms. Oppel and Hall

advocate the theory that an endocrine imbalance on the basis of parathyroid dysfunction with resultant calcium metastasis is the main etiologic factor in certain types of ankylosing spondylarthritis of obscure etiology. Hall does not hold that this calcium metastasis is purely a defense reaction on the part of Nature to preserve the painful joints from destruction by immobilization but rather that the calcium metabolism is not under normal control, or is unduly stimulated so that if Nature is trying to protect the diseased areas she carries the process entirely too far. He believes, then, if infection plays a major part in the etiology of spondylitis it would seem quite probable the toxins from the same infectious process might act as parathyroid stimulants and thus parathyroidism becomes the most important factor in producing the ankylosing changes. (2,4,5)

Blair is of the opinion that the pathological changes which occur in spondylitis adolescens are caused by an absorption of cartilage brought about by a general metabolic change. He reasons that it is possible to suppose that absorption would begin in an area of low vitality. It is known that disease or destruction of a joint produces immobilization in that joint. While this immobilization is Nature's method of relieving pain, it also accelerates the absorption of cartilage and the atrophy

of bone. The process spreads until ultimately all cartilage is absorbed and bony ankylosis supervenes. Thus he concludes; the sacro-iliac joints contain large amounts of cartilage, normally there is very little movement in these joints; therefore the tissues here are of low relative vitality and so it is the sacro-iliac joints that are the first to be attacked by the fixation process in spondylitis adolescens. (6)

Jacques Forestier believes that the primary focus in ankylosing spondylitis is in the genito-urinary system or in the low part of the bowels and that the toxic products excreted by this focus are drained into the lymphatic system of the pelvis and from this place alongside the spine. In his pathogenesis he points out that in the male the lymphatic vessels from the prostate and the seminal vesicles pass in front of each sacro-iliac joint and extend upward in the posterior part of the abdomen on both sides of the spinal column just in front of the apophyseal joints with which they have many connections. In the female the lymphatic vessels of the uterus and the vagina follow approximately the same path but conversely the lymphatic vessels of the Fallopian tubes and of the ovaries are much more laterally situated in the pelvis and when they come up into the abdomen they lie on the ventral aspect of the large blood vessels while in the

male they are directly applied on the vertebral column behind the aorta and the vena cava. Knowing that the draining of the uterus and vagina is far easier than that of the ovaries and tubes, it is understandable why the occurrence of ankylosing spondylitis is so infrequent among females. It also quite satisfactorily explains the slow progression of the disease; first to the sacro-iliac joints and later ascending along the different segments of the spine. (7)

Scott believed that the process, which occurs in the sacro-iliac joints, is infective in nature. He had four suitable cases trephined by an experienced orthopedic surgeon using the technique of Smith-Petersen. Specimens were removed for investigation from the trephined sacro-iliac joints. The subsequent laboratory report revealed that the results were unsatisfactory as the bacterial investigation was nullified by contamination. The contents of the sacro-iliac joints also gave a negative result to the animal test for bacillus tuberculosis, this test being made as Scott always thought, and still thinks, that some mild grade of tuberculosis may eventually prove to be responsible for the infection of the sacro-iliac joints.(8)

The conception that this disease is infective in character, being produced by a variety of microorganisms and develops as a complication of various diseases, as

gonorrhea, tuberculosis, bacillary dysentary, and others is the one that seems to have survived the test of time so far. However there were some authors that would attempt to supplant this idea in favor of a rather new one. They believe that it is a diffuse vertebral polyarthritis of an inflammatory origin involving the synovial membrane of the vertebral joints becoming a wide-spread periarticular process with secondary ligamental ossification; the inflammatory process evidenced by an increased sedimentation rate. (9,10,11)

It was not the purpose of this paper to attempt to settle the question of the etiology of this disease or its mode of infection, so we have just pointed out those that have survived the test of time and some of the more likely possibilities of recent investigations. Inasmuch that this writer is attempting to prove that the very first sign of involvement of ankylosing spondylitis is in the sacro-iliac joints I am more or less in favor of those etiological theories concerning the sacro-iliac joints but in the words of Scott there is as yet no satisfactory answer to the problem of the source of the primary infection in spondylitis.

PATHOLOGICAL PROCESSES

While the nature of the disease has not yet been absolutely determined, it is generally accepted that its pathological process starts as an atrophic arthritis of the joints between the facets of the vertebrae which after a primary stage becomes involved by bony ankylosis. (7) Joseph L. Miller is of the opinion that this pathological process begins in the synovial membrane and the only synovial membrane in the spine is in the small lateral articulation of the transverse processes and ribs, which is usually referred to as the small articulations of the spine. He states that while it is not conclusively proved, it is generally believed that these articulations are first involved in this disease. (12) As the disease is chronically progressive in character further pathologic changes develop rather slowly. Secondarily these later changes are ossification of the intervertebral ligaments, especially the anterior longitudinal ligament but the posterior also becomes involved in a good number of cases. The periphery of the intervertebral disc may become ossified as well as extension of the spongiosa of the vertebra through the central portion of the disc. In the roentgenograms, the classical picture of the bamboo-spine results if the pathological changes are completed and the involved spine structurally resembles a long bone. (7, 12)

Louis Ramond states, "Rhizomelic spondylosis is characterized essentially by a practially complete fusion of the vertebral column. The writer presents a typical case, in which the head, neck, trunk and thighs form one rigid unit. Walking is possible, or rather, a sort of shuffling along with tiny steps and the support of two canes, the head rigidly stretched forward, and the knees slightly flexed. To be put to bed, the patient stands with his back turned towards the bed, and the nurses place him, first transversely, then horizontally, using his ischia as fulcrum and pivot. Established in bed, he remains supine, unable to sit up or be propped up by cushions in a semi-reclining position.

Examining the various segments, one finds that the cervical segment is markedly flexed forward, the head slightly raised but rigidly fixed in this position, and the entire dorsal and lumbar segments form a rigid trunk, without any lumbar lordosis and only an indication of dorsal kyphosis, where the cervical segment begins to bend forward. The thorax is flat. The upper arms are soldered to the sides of the chest, and the movements of the scapulohumeral articulations are considerably limited. The elbows, wrists and hands are perfectly free. The pelvis is flattened and heart-shaped. The hips are immovable and the thighs are fixed in semiflexion, external rotation

and slight abduction. The knees, ankle joints and feet are free. The patient is emaciated and shows a certain degree of muscular atrophy, especially in the region of the fused joints, but otherwise, no organic taint.

The condition started, about sixteen years ago, with dull pains in the hips and sacro-iliac regions. These pains did not radiate, or exacerbate on coughing or on fatigue and, for a long time, did not disable the patient for his difficult profession of electrician. The hips were the first to be incapacitated; in 1937, the vertebral column and the neck followed suit.

The x-rays show: Sacro-iliac line on both sides, abolished; sacrum and ilia, completely fused; vertebral bodies, very poorly opaque, manifestly affected with osteoporosis, but with the contours conserved (no diabolo form, no parrot beaks, or other osteophytic deformities); presence of calcifications of ligamental and other peri-vertebral fibrous structures, especially at their osseous insertions, giving the lumbar segment the peculiar aspect of a twisted trunk ("wine press screw", or "bamboo stick"). The scapulohumeral, coxofemoral, and other joints show no radiologic changes." (9)

Cecil in his section on rheumatoid arthritis in his textbook gives almost the same picture as does Ramond. He says that ankylosing or Marie-Strumpell spondylitis is

a disease of mostly young men and that the typical Marie-Strumpell spine is rarely encountered in the female. As does Ramond he states that ankylosing spondylitis usually makes its first appearance in the sacro-iliac and the lumbosacral joints with the small articulations of the entire vertebral and costovertebral joints often affected. The disease progresses upward with the longitudinal ligaments calcifying and eventually ending with complete ankylosis of the entire spine. His cases present characteristic appearances having a flattened lumbar curve and the dorsal curve exaggerated with flat and rigid chests. The deep muscles are in spasm and the patient flexes his body at the hips as the entire spine is held rigid. There is marked atrophy of the trunk muscles and poor chest expansion. The radiographic examination also presents the same type of condition as Raymond found. Early, there may be no changes to be seen but in advanced cases there is demineralization of the bone, obliteration of the small intervertebral articulations and calcification of the intervertebral ligaments giving rise to the typical bamboo spine. Cecil also found the sacro-iliac joints affected in 90 per cent of the cases. (13)

Thus with examples from both European and American Medical Annals it is found that the diagnosis of Marie-Strumpell disease is made upon the classical picture established in the past century by Marie and Strumpell. In it is seen the final result of this disease and it is not a pleasant one to behold and realize that medicine has been able to do little or nothing to prevent or even arrest this crippling disease. This clinical picture prevailed up until recent years and evoked but little interest in its cause, probably accounted for by the fact that in the past little could be done in the way of treatment in this disease. The best that could be hoped for was that the disease might "burn itself out" before the victim became quite crippled. But as Samuel Gee says in his Clinical Aphorisms, "Therapeutics must begin before physical signs have developed, for if you wait for physical signs, you wait too long;" and the fact that so much depends on the early recognition of the disease before irreparable damage has been done to the spine caused men interested along these line to make further investigation. Several men in particular have worked on the problem and have brought forth their contentions that an earlier recognition can be made of this disease. A hypothesis will be constructed not only from their works with an attempt being made to contain all of

the important diagnostic criteria by which they arrived at an early recognition of this disease, and from observations made by myself while I had this condition, and case histories. After this supposition has been set up, it is the purpose of this thesis to try to substantiate it through the courtesy of Dr. Herman Johnson and the University of Nebraska, College of Medicine in extending the use of their files to find these earlier signs and symptoms in known cases of Marie-Strumpell disease.

Scott from his investigations concluded that the onset of spinal symptoms signified not the commencement of the disease but the final stages of a long-standing pathological condition. (14) However, though Marie had reported that the sacro-iliac joints had undergone a bony ankylosis in his first description of the pathological changes occurring in the disease; it was not until the last decade that the well-known appearance of bamboo-spine became associated with this fusion by roentgenologists.

Loring T. Swaim and John G. Kuhns were among the first to mention this fact. They observed that it was often first noticed in the sacro-iliac joints and extends upward. (10) In 1935, C. W. Buckley in his survey of more than a hundred cases stated that he found complete bony anklylosis of the sacro-iliac joints in the majority of them. In nine of these cases the joints were not fused

but some gross changes had taken place. He also reported a few cases in which he had detected these sacro-iliac changes at an early period of the evolution of the disease and previous to any changes of the joints of the facets as seen roentgenologically. (15) Jacques Forestier reviewed 153 cases of ankylosing spondylitis and found only two cases in which the sacro-iliac joints did not appear to be roentgenologically involved. However he was not certain that one of the cases was a true ankylosing spondylitis, the other one had normal sacro-iliac joints roentgenographically but he pointed out that the spine was free from the typical changes of the disease. (7) Scott as a result of his work in the disease at the British Red Cross Rheumatism Clinic in 1936 reported that all 110 cases of spondylitis adolescens he had examined so far had shown indication of bilateral infection of both sacro-iliac joints usually in the form of ankylosis radiographically. (14) In 1942, Scott states that this particular type of spondylitis can easily be identified by one single characteristic; the sacro-iliac joints always present pathological changes radiographically. Bony ankylosis completely obliterating the sacro-iliac joints in his experience was peculiar to this disease and as far as he knew had never been found in any other condition.(8) I believe that one can safely reply in the affirmative to

the question: "Are sacro-iliac joints always involved in the course of Marie-Strumpell syndrome?"

Even through the poker-back spine came to be associated with bony ankylosis of the sacro-iliac joints and possible calcification of spinal ligaments, this association aroused but little interest and not much attempt was made to explain the relationship, if any, to the disease. Scott attempted to obtain a complete radiographic record of those pathological changes which precede or gradually lead up to the ankylosis of the sacro-iliac joints. However here he ran into some difficulty, he assumed that he would find the early or active stages of the process associated with the early clinical symptoms of the disease. It was found that in a number of cases where the spinal symptoms had only been present for the short period of six months, the sacro-iliac joints were already completely ankylosed. From this, his conclusion was, "that the onset of the infection in the sacro-iliac joints did not coincide with the onset of spinal symptoms of spondylitis." Blair also found, at the time of first examination, the previous clinical history to be short - a month or six months - and the roentgenograms usually showed advanced changes in the sacro-iliac joints. This indicated to him as well as to Scott that these changes apparently caused no pain, disability or other symptoms to be referred to the affected

sacro-iliac joints during their active stage. In this disease the symptom of pain appears to become localized to the affected area only in the final stages of ankylosing. It is not until this ankylosing process begins that pain or stiffness of the back is complained of. (6,8)

This finding, however, did not really contribute much towards solving the problem of what were the early changes that preceded the ankylosing of the sacro-iliac joints. Nothing of importance could be demonstrated that would lead to the answer of the problem. Then Scott noticed in the course of taking the clinical history of these patients that when he would ask them the question, "Did you experience any rheumatism before your back got bad?", the answer was always in the affirmative. The majority would reply that they have had attacks of wandering rheumatic pains every since they were boys. This observation brought out many questions: "Could these early attacks of pain actually indicate the period during which the pathological process in the sacro-iliac joints was in its most active state? Was this the period that actually preceded the ankylosing process in those joints? Did these attacks indicate a much earlier stage of the disease than the spinal stiffness and pain of the text-book descriptions?" Scott sought the answers to these questions by seeking out all those patients attending the British Red Cross

Clinic and who gave a history of recurrent attacks of rheumatism over a period of years and were actually being treated for aches and pains which could not satisfactorily be accounted for. His radiographic survey of the sacro-iliac joints showed in most of these cases definite pathological changes in these joints. Here was evidence, not only of an active stage involving the sacro-iliac joints that preceded their ankylosing but also evidence of certain clinical symptoms representing a hitherto unknown prodromal stage of this disease which Scott designated as the pre-spondylitic syndrome. The assumption made from this discovery was that it meant a definit percentage of patients under about 25 years of age giving a history of recurrent attacks of wandering rheumatic pains extending over a number of years might be expected to develop typical spondylitis sooner or later. (8)

The confirmation of the above assumption also proved difficult. It was hard to obtain a complete series of radiographs which showed in one patient the whole development of this condition. The reason being that the sacro-iliac joints were rarely radiographed in the early or active stages of this disease as there were no symptoms referable to this area at this time and also the pathological process takes five to seven years to develop. In order to obtain a complete record of an established case of spondylitis it was necessary to get hold of early radiographs

that might have been taken during this period and which by chance happened to include the sacro-iliac joints. Scott at the time of his writing was only able to secure a complete radiographic record of the pathological process in nine patients out of 300 cases. These serial records of the nine cases were of considerable importance as they clearly demonstrated radiographically the active stages of the pathological process in the sacro-iliac joints that lead up to ankylosis. They recorded those changes in the sacro-iliac joints that occurred several years before the onset of spinal symptoms. They also confirmed the suggestion that changes in the sacro-iliac joints always preceded the onset of spondylitis and that sooner or later those persons showing similar changes in the sacro-iliac joints will develop the clinical symptoms now associated with ankylosing spondylitis; namely, pain and stiffness of the spine. Blair remarked in his observations that pronounced changes had already taken place in the sacro-iliac joints and often without the patient realizing anything was wrong before the symptoms of back pain had first appeared. Forestier found in all of his cases that the patients complained of pains and stiffness in the spine with muscular contracture, lumbo-sciatica, and impairment in the chest expansion, but as a rule no pain at the site of the sacro-iliac joints. He associated these clinical

signs with some definite roentgenographic signs in the sacro-iliac joints as being pathognomonic of ankylosing spondylitis. (6,7,8) Forestier's observations may have preceded those of Scott's slightly in the sequel of events in making an early diagnosis of this disease but it may be said the fundamental fact remains; that, changes of some sort can actually be detected by means of a radiographic examination several years before there is any suspicion clinically that the patient may eventually develop spondylitis symptoms.

What are these diagnostic changes detected radiographically in the sacro-iliac joints? For an understanding of these changes it was thought necessary to have in mind a clear picture of the normal sacro-iliac joint. The sacro-iliac articulation is an amphiarthrodial joint which is formed between the auricular surfaces of the sacrum and the ilium. A thin plate of cartilage covers the articular surface of each bone which is thicker on the sacrum than on the ilium. There is usually a small cavity containing a small amount of synovial-like fluid between these surfaces, especially in advanced life giving the joint the characteristics of a diarthrosis. The fine interosseous fibrous section is confined to the upper and posterior part. The opposing surfaces here are very irregular and dense fibrous tissue

binds the two bony surfaces firmly together. (16)

Radiographically these two sections can be distinguished. The anterior and posterior fissures outline the cartilaginous portion. Examined radiographically the whole outline of the joints appear pear-shaped. The cartilaginous section representing the 'pear' and the fibrous section being the 'stalk'. This pear-shaped shadow actually does not outline the joint in its entirety but for all practical purposes any deviation from the normal is betrayed sufficiently by changes occurring in this radiological outline. (8) Plate 1.

However correct interpretation is never easy in these early cases, for not only is there great variability of the roentgenographic films of the sacro-iliac joints in normal persons but, congential deviations from the normal occur quite frequently and must be considered in making an early radiological diagnosis of this disease. However, these congenital variations are usually confined to the cartilaginous section of the spine and are also usually found in conjunction with other congenital abnormalities of the spine. Scott believes that the diagnosis of osteoarthritis based on the presence of bony spurs at the lower extremity of the sacro-iliac joints is very unlikely. He does not believe them to be osteophytes which are characteristic of osteoarthritis, but rather a

congenital variation as these 'thorn-like' bony projections are by no means uncommon in this position and have been found in young individuals where the symptoms of arthritis of these joints are entirely absent. The sacro-iliac joints are seldom attacked by disease. Tuberculosis is possibly the most common, if not the only, infective condition found. These joints may also be involved secondarily in the form of metastases from malignant disease, but it would appear that these joints are singularly immune from infection. Difficulty is also encountered in the interpretation of radiographs of the sacro-iliac joints of patients under seventeen whose bony development is not yet complete. The edges of the wings of the sacrum are still semi-cartilaginous, consequently the shadows of the sacro-iliac joints appear to be much broader than in the adult. Instead of the pear-shaped shadows of the fully developed joint they are less defined and appear as single broad fissures. (8)

Blair in examining the sacro-iliac joints noted in the roentgenograms that the first abnormal change was a widening and an irregular fuzziness of the sacro-iliac joints. There occurs, nearby, areas of localized bone absorption or a general atrophy of the adjacent sacrum or ilium ensues which, however, is usually 'spotty' in character. The cartilage is then absorbed and the joints

as such disappear with bony ankylosis taking place. Increased density of the bones in the sacro-iliac region usually follows this. Examination of the remainder of the spine reveals that the same process is taking place. The posterolateral articulations disappear as their cartilage is absorbed and the bodies of the vertebrae show considerable halisteresis. The supporting ligaments also are involved as they are seen to have become calcified and later ossified as remission takes place. The other soft tissues in the body do not appear to be involved by this calcification and ossification. The intervertebral discs show more or less calcification but do not lose width. (6)

The roentgenographic changes in the sacro-iliac joints that Forestier described dealt with the joint space, the contours of the bones and the structure of the subchondral bone with the latter two elements being the more important. In fact the joint space as a rule is not thinned nor becomes irregular by cartilage or bone erosion as in a true arthritis. He emphasizes that the changes in some cases do not, at the beginning, affect the entire length of the joint but only a part of it, with the rest of the joint space appearing to be absolutely normal. Later on the entire joint becomes abnormal with the rapid disappearance of the joint space

altogether without passing through a stage of diminution of its width. (7)

Forestier classified his cases into three stages to make his description more clear. His first stage consisted of a pseudo-widening of the joint space. The contours of the subchondral bone become woolly and hazy on a segment of one or both joints. The clear-cut outline of the joint space on the iliac side disappears entirely and there is a loss of calcium alongside the margin of the bone. It is this marginal decalcification that results in the appearance of a widening of the joint space. After several months or a year the development of the first stage ends in a mottled appearance of the cancellous bone both on a wide area of the lateral portion of the sacrum and of the ilium. Some small spots appear very translucent, others look hypercalcified or spotted---in French 'aspect tigre'. The joint space itself is hardly visible on the mottled surface. The third stage is a far remote and a terminal one with the changes in the greatest majority of cases affecting the whole length of both sacro-iliac joint spaces and are frequently accompanied by ossification of the iliolumbar ligaments. There is more uniformity in the calcification of sacro-iliac region and the appearance of thin ossified fibers transversely across the joint space

often accompanied by osteosclerosis in the adjoining regions. The bony densification tends to become more and more marked as the disease progresses. This is the stage in which there is a loss of joint space and synostosis. (7)

Scott also concluded that the very earliest signs of sacro-iliac joint involvement occurred under three guises. In his first stage the anterior or posterior fissures suffered a loss of definition. The whole joint undergoes a general broadening and blurring with the fissures tending to fuse into a single irregular shadow. The cartilaginous section undergoes a mottling also. These slight but significant changes are frequently overlooked in this early stage or, even if detected, are easily passed over as of no consequence. In the second stage it is found that the usual well-defined, narrow, pear-shaped linear shadow now has become broad and indefinite with the fissures being obliterated. The now broadened shadow of the joint has a "rosary" appearance due to the small rounded cavities that may be present and these cavities tend to coalesce to form larger cavities. The radiological features of the third stage show the fissures to be obliterated and replaced by irregular areas of rarefaction. There is osteosclerosis in the neighborhood of the joint if resistance is high while if

resistance is low there is absence of osteosclerosis and presence of osteoporosis or decalcification. It is possible in this stage to gauge with some considerable accuracy the probable resistance of the patient from the bone reaction; that is, the sclerosis around the joint. If there is a marked degree of sclerosis of the bone in the neighborhood and bony ankylosis of the joint is rapidly completed, it means a high resistance and consequently a favorable prognosis. If, however, the disease makes rapid progress owing to a low resistance, the sacro-iliac joints are obliterated and replaced by irregular areas of rarefied bone, usually with some degree of sclerosis in the neighborhood, frequently displayed as a 'halo' around the joint. The presence of decalcification of bone or localized osteoporosis denotes a high degree of virulence which while an unfavorable sign does not indicate a bad prognosis as Scott found that these patients responded to his therapy just as satisfactorily as those with normal bone density. The earliest radiological evidence of onset in spondylitis is not decalcification of bone, generally the bones of the pelvis and lumbar spine as Buckley, Miller and other authorities believed. The loss of bone calcium must be looked upon as a late manifestation and attention placed on changes in the

sacro-iliac joints. (8,12,17)

The clinical history plays a major role also in the making of an early diagnosis of ankylosing spondylitis for unless the physician or surgeon, as the case may be, recognizes the prodromal symptoms from the history, the radiologist is not apt to have the opportunity of examining the sacro-iliac joints in the initial stages of involvement. It would seem important, therefore, to enumerate those clinical signs and symptoms which should rouse suspicion of early spondylitis in the mind of the physician, and which call for a radiological examination of the sacro-iliac joints. These early symptoms may antedate the onset of spinal symptoms by several years. (8)

I would like to present my own observations made while undergoing this disease in enumerating the clinical signs and symptoms. Up until 1935 I had been well, active and healthy, when in the winter I came down with what my local doctor called sciatica. Severe knife-like pains envolved the right gluteal area and radiated downward into the posterior and lateral aspects of my right thigh. These pains were made worse by movement. Treatment consisted of local application of heat and massage for a period of two months after which time the pain disappeared. This temporary remission gave me a sense

of security for I was apparently free from pain except for an occasional stab of pain in my right hip and for some fleeting pains across my shoulders. I went on this way until 1936 when another seizure came along. This time the pain seemed to be referred to my right hip. The movements of that joint were painful and limited, which made walking a very difficult procedure. At times the pain seemed to radiate into my left buttock and thigh but there was no pain in my back. My general health at this time was becoming poor and there was some wasting of the thighs and muscles of the buttocks. Under symptomatic treatment I again apparently recovered. I went on again suffering no severe attacks but rather recurrent episodes of wandering rheumatic pains across my shoulders, round my ribs, down my legs to the knees but mainly in my hips. I also noticed that my chest was becoming flattened anteroposteriorly with expansion impaired and marked tendency towards abdominal breathing. 1940 found the pain to be rather constant in my hips and I was beginning to notice some painful spinal stiffness. Walking and stooping became extremely difficult and I noticed that I was frequently obliged to get up several times in the second part of the night to relieve the muscular spasm in my spinal muscles, sometimes spending the rest of the night sleeping sitting up in a

chair. During the first part of the night I seemed to obtain relief from these lumbar pains by bed-rest. Any movement in bed was extremely difficult, having to raise myself up and then ease over into a new position. The muscles of my body seemed to be in the state of marked hypertonicity or irritability as a slight blow would cause a ripple of progressive contraction to travel some distance along the fibers. Even to cough or sneeze was very agonizing and I tried to avoid these luxuries as much as possible. In spite of the rather acuteness of this attack I had several partial remissions in which the symptoms abated somewhat but all during this time I was gradually losing weight and was excessively fatigued all of the time. By the time a diagnosis was finally made in 1941 I presented a sorry-looking picture. I was markedly underweight with a general sharpening of my facial features, flat chested, and very round shouldered due to a slight forward bowing of cervical and dorsal portions of my spine with a loss of the lumbar lordosis. My back was stiff to movement and x-ray revealed a fusing of the sacro-iliac joints. I was quite discouraged as to the future for not only had all methods of treatment constantly failed but also no one had satisfactorily diagnosed my condition. With the diagnosis finally made, orthopedic therapy was carried

out with satisfactory results.

It was found in the literature that these vague and diffuse pains in the early stages of the disease were variously labelled sciatica, muscular rheumatism, lumbago, colitis, pyelitis, fibrositis, or even 'growing pains'. The age of the patients varied between 20 and 35 with the onset of the first mild symptoms occurring between the ages of 18 and 25 years, Scott thought the age of early onset to be lower, around 12 to 14 years. The disease was observed almost exclusively in the male with it being a rarity among women. The sedimentation rate is usually increased, sometimes to a considerable degree. (6,7,8)

The individuals attacked by this disease had all been healthy young persons prior to the onset of their pain. Some had been subjected to a vigorous examination for foci of infection such as; removal of tonsils, suspicious teeth, sinus operations, and afterwards followed by all forms of physiotherapy and various supports. But in spite of the varied treatment the course of the disease was not shortened, it progressed on to complete ankylosis. (6)

Scott found a few variations in the life-history of the spondylitic. After the pathological process has commenced it may pass through the progressive stages until

the disease has reduced the victim to a cripple with a 'poker-back' in the course of a few years. The process may become permanently arrested at any stage without producing definite spondylitic symptoms which is a good thing otherwise there would be very many more spondylitic cripples than there are; the patient being quite unaware that he has escaped serious spinal trouble. A chance radiographic examination at a later period may reveal for the first time ankylosis of the sacro-iliac joints with or without calcification of spinal ligaments. A primary onset may be imitated as the process may abort early in life only to 'flare up' again later, giving rise to the erroneous impression that ankylosing spondylitis can develop after middle-age. (8)

The clinical and radiological findings having been presented it is now time to formulate our premise upon which an earlier diagnosis of ankylosing spondylitis may be made. It is apparent that an attempt was made to demonstrate an association of these recurrent attacks of wandering rheumatic pains in the young adult with some definite radiographic signs in the sacro-iliac joints as pathognomonic of ankylosing spondylitis and by this means a diagnosis could be made much earlier than had been made previously.. Our premise would find the patient in the teen age group or in the very early twenties with

no spinal symptoms. There would be pain in the knees, thighs, hips, or even girdle pains with perhaps a previous history of recurrent attacks of pain in the other joints. Upon physical examination there would be few if any physical signs, perhaps a slight spasticity of the muscles of the involved region. Radiographically there would be definite changes in the sacro-iliac joints, either of one of the three stages previously described or even partial ankylosis.

DATA SUPPORTING PREMISE

In seeking evidence to uphold our contentions established above, six cases of known diagnosed Marie-Strumpell disease were found in our source of records that I had access to. Two of the cases were particularly interesting in that they presented some of the variations in the life history of the spondylitic noted by Scott. One was a 40 year old male who developed a stiff neck 10 years ago with increasing back-ache. He also complained of girdle pain in the abdominal region. Upon physical examination it was found that the patient walked with a protective gait with the trunk in 30 degree forward flexion. Examination of the spine showed a moderate dorsal rounding with flattening of the lumbar spine. X-ray findings were that of the typical bamboo-spine of Marie-Strumpell disease. The other case was of a female, age 48, who complained of pain in back of both hips which radiated down the back of the legs for two years. She also complained of pain in the pelvis radiating into the lower abdomen and down the inner aspect of both thighs for nine or ten months as well as soreness in the lower abdomen for the same length of time. This pain was aggravated by stooping and nothing seemed to relieve it. Radiological findings were a straightening of the anterior margins of the bodies of the lumbar vertebrae most marked in L-4 and L-5. There were hypertrophic changes

present in the pedicles of the fourth and fifth lumbar vertebrae and a considerable increase in density in the region of the facets of the fourth and fifth lumbar vertebrae. The right sacro-iliac joint was partially obliterated by sclerosis and ossification. The comment by X-ray was that this was an early spondylitis rhizomelica.

Here are two cases which apparently have started rather late in life but which according to Scott would merely represent flare ups of earlier unrecognized cases of spondylitis. It was interesting to note that in the second case, it was thought variously to be a gynecological and neurological problem before the final diagnosis was made. These two cases can serve as examples that not only may the disease give rise to symptoms late in life, but also although it is rare it can occur in the ratio of about 14 to 1, in women, so when a diagnostic problem presents itself of a similar nature, keep ankylosing spondylitis in mind. (18)

The third case was that of a 31 year old male whose story was one of suffering from a fall upon his back with continuous pain for four weeks. His spine upon examination showed moderate kyphosis in the dorsal region with the lumbar lordosis flattened out. There was scoliosis to the left in the dorsal region. Radiologically there

was extreme calcification of the paravertebral ligaments with bilateral fusion of the sacro-iliac joints. Involving all of the mid-dorsal vertebra there was an old advanced kyphosis of about ninety degrees. At the level of the second to third lumbar bodies there is a fracture through the calcified paravertebral ligaments with a fracture through the roots of the posterior articulating processes of the second lumbar segment. Their comment was spondylitis rhizomelica with fracture dislocation between second and third lumbar segments with no gross displacement.

The important point in this man's history was that ten years ago at the age of twenty-one he complained of rheumatic pains across both hips and shoulders with the shoulders gradually drawing down. These rheumatic pains came on following a pyuria. There has been no change in his spine the last two or three years.

Case number 4 concerned a 33 year old male laborer who complained of increasing stiffness of back with thoracic kyphosis for the past ten years. Ten years ago patient had severe stabbing pain in the lumbosacral area with no radiation of this pain. Heavy lifting and stooping made the pain more severe. During the past four years the pain has become less severe with more and more spinal stiffness and kyphosis. There was

thoracic kyphosis and right lateral scoliosis upon skeletal inspection. The lumbar skeletal curve is flattened when bending forward. The chest is flattened and has diminished expansion. The typical findings of spondylitis rhizomelica was fusion of spinal segments and sacro-iliac joints were seen on radiological examination. But the diagnostic feature that I would like to bring out in this case was that there was a history of rheumatism in the right hip and knee when the patient was eighteen years old.

However, the task of finding radiological evidence turned out to be one of a difficult nature. No radiographs could be found that might have been taken in the early pre-spondylitic stages of the disease but rather only those that turned out to be good illustrations of advanced spondylitis who for the most part could be diagnosed from the clinical symptoms and findings. However in my case file I was able to find a series of radiographs which showed in one patient the development of the pathological process in the sacro-iliac joints that lead up to ankylosis recording changes in the sacro-iliac joints that had occurred several years before the onset of spinal symptoms. Plates 2,3,4

The last case in our series was one in which the patient was shuffled around with various labels attached

to him. It was thought to be: a muscle strain in the back, then a foci of infection was thought to have been found in the teeth and so two of them were removed, a course in vitamin treatment was instigated and finally he was thought to be suffering from nephrolithiasis before a final diagnosis of spondylitis rhizomelica was made.

In the above series of cases the last five will be found in the files of the University of Nebraska, College of Medicine and the first one will be found in the files of Dr. Johnson. While it was not possible to question personally these patients, nevertheless, these early attacks of pain indicating a much earlier stage in the disease were found in three of the cases. A serial radiographic record was obtained in one established case of ankylosing spondylitis which covered several years and clearly demonstrated the existence of a progressive active process in the sacro-iliac joints followed finally by complete ankylosis of the joints.

I believe that sufficient data has been given to support the contention of this thesis that an earlier diagnosis of Marie-Strumpell disease can be made. Some men might scoff at the presentation of so few a number of cases as supporting evidence or that such few cases were found therefore it must be rare so why worry about

it. The answer to their scoffs is that not all of the patients with rheumatism are found in the hospitals, many of them are going unrecognized and when they are diagnosed they are in the late stages of the disease when most of the damage has been done. I have found evidence to support my premise which was built upon the cumulative evidence of other men who had evidence to support their contentions. It is no laughing matter to one who is suffering from the disease to be shuffled around under various but not definitely established diagnoses and no effective treatment given to relieve the pain. One of Scott's cases might be quoted to further emphasize this point. "A young adult complaining of general rheumatism for some years came to me for a radiographic examination of the right hip-joint which was giving him trouble at this stage. Suspecting an early spondylitis with pre-spondylitic symptoms, the sacro-iliac joints were examined. The radiographs undoubtedly showed the characteristic changes associated with sacroilitis, but in a very early stage. I consequently reported that the evidence was in favor of an early spondylitis and that the outlook was grave if nothing was done.

Unfortunately for the patient, he was reradiographed by another radiologist the following week. My colleague

was experienced, but not in spondylitis, and failed to confirm my opinion which he considered unnecessarily pessimistic; so nothing was done. This same young fellow presented himself at the Charterhouse Rheumatism Clinic for treatment a few years later, a complete cripple, being now in an advanced stage of spondylitis with ankylosed sacro-iliac joints and a bamboo spine, a condition that could have been prevented if my warning had been heeded."

Perhaps another reason that not much interest was given to this disease is the fact that the treatment of spondylitis has not been too hopeful. However, the outlook is more favorable now for not only has there been advancement made in the realm of therapy for ankylosing spondylitis but it has also been shown that the diagnosis can be made several years before any crippling occurs.

Treatment is a broad field in itself and also one in which there are many differences of opinion. It is not within the scope of this thesis to delve into the matter of therapy; suffice it to say that treatment for ankylosing spondylitis has, in the main, resolved itself around three lines of thought: orthopedic, (19) wide-field roentgen-ray therapy, (8) (20) (21) (22) and a combination of those two. (23) However most of the

workers were in accord in their opinion of one thing and that was; early diagnosis is imperative for the best results of treatment. In fact Philip Lewin states that the remarkable results obtained by Scott so far in 300 cases of spondylitis in all stages extending over a period of eight years with wide field radiotherapy justifies his conclusions that it is possible to obtain in a large majority of, if not in all, cases complete and permanent arrest of this disease provided that they were treated in the early stages of the disease.(18)

The problem that this thesis undertook to solve has been successfully fulfilled by demonstrating that there was an earlier involvement occuring in ankylosing spondylitis than was heretofore known. This early involvement concerned the sacro-iliac joints and could be recognized radiographically, attention attracted to the sacro-iliac findings by certain clinical findings in the patient's history and by these means an earlier diagnosis could be made in Marie-Strumpell disease. It was further pointed out that the establishment of an earlier diagnosis of this disease brought about more favorable results in its treatment.

It is hoped that the seriousness of this disease has been forcibly brought out so that future readers of this work will think of Marie-Strumpell disease when

they come across patients who give a history of recurrent attacks of rheumatism over a period of years or who might actually be undergoing treatment for aches and pains which could not be satisfactorily accounted for and thus free future patients of pain, crippling, and anxiety.

Plate No. I

Normal appearing sacro-iliac joints showing the anterior and posterior fissures enclosing the "pear shaped" cartilaginous portion and the "stalk" formed by the fibrous section.

Plate No. II

Early sacro-iliac involvement as evidenced by a loss of definition of the anterior and posterior fissures with partial obliteration and a slight mottling of the cartilaginous section.

Plate No. III

Case No. II four years later: spinal symptoms first noticed. Partial ankylosis and sclerotic changes in right sacro-iliac joint are seen now.

Plate No. IV

Case No. II six years later: now typical ankylosing spondylitis with ankylosed sacro-iliac joints.

Plate No. V

Spinal changes of ankylosing spondylitis evidenced by a squaring of the vertebrae, a feature pointed out to me by Dr. Hunt.

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