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## Syndrome produced by tumors at the foramen magnum

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THE SYNDROME PRODUCED BY TUMORS AT THE FORAMEN MAGNUM

BY THOMAS EMMETT MANGUM JR.

SENIOR THESIS: PRESENTED TO THE COLLEGE OF MEDICINE  
OF THE UNIVERSITY OF NEBRASKA IN THE YEAR 1940 A. D.

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## INTRODUCTION

The object of this paper is to report four cases of pathology at the foramen magnum which were furnished the author through the courtesy of Dr. J. J. Keegan. After the four detailed case reports a review of the literature is attempted with the idea of working out the possibilities which come into a syndrome when pathology occurs at the level of the foramen magnum.

Of the cases reviewed in the literature forty-eight are tumor, two are aneurysms of the vertebral artery, one a dislocation of the odontoid thought to be congenital and the other a fracture of the odontoid process similar to case two in the original part of this paper.

Cases reported in full in the first part of the paper are not included in the chart, which is made up of only cases reviewed in the literature. With each case, on the chart, is the bibliography of that case.

I wish to thank Dr. Keegan for allowing me to include the original cases in this paper. To Dr. R. Young and Dr. P. Tollman I am also grateful, for their help in the organization of the material reviewed from the literature and its presentation in this paper.

## THE SYNDROME PRODUCED BY TUMORS AT THE FORAMEN MAGNUM

Tumors at the level of the foramen magnum are rare, but are interesting in that they present a problem not only in diagnosis but also in proper surgical management. Following are four case reports which present most of the features of the syndrome.

Case I : (A-143-39) C. Y. a white married male farmer, age 34 entered the University Hospital 11/21/39 complaining of:

1. Numbness of the right side, especially the right arm, for the past one and one-half years.
2. Difficulty in swallowing for the past four months.
3. Vertigo for the past five months.
4. Stomach distress since the spring of 1938.

Present Illness: In June 1939 the patient started to have sensations of numbness in the right hand which progressed to involve the whole arm, right side and right leg down to the knee. Three weeks after the above onset the patient stated that he suffered difficulty in swallowing. This has become progressively worse to date. Objects began to whirl in June of 1939, and recently these attacks have become more frequent. During the past few months he has fallen several times but has not lost consciousness.

Family History, Marital History and Past History are all negative.

History by Systems negative except as in Present Illness. He had received treatment for gastric ulcer two years previous to hospital admission.

Physical Examination: Negative except as in Neurological. The eyes showed a horizontal and vertical rotary nystagmus. The tongue protrudes to the right. The neck showed some tenderness on passive motion and seemed rigid; some torticollis present.

Cranial Nerves: No papillo-edema. Third, fourth and sixth show a rotary nystagmus. Hypesthesia over the left lower fifth distribution. Slight weakness of upper right face. Ninth, tenth and eleventh--Gag reflex absent and palate deviated to the left. Twelfth--deviation of tongue to the right with right sided atrophy.

Trunk: hypesthesia in the left pectoral region. Arms and legs showed good motor power with indefinite sensory changes. Romberg was positive and Gait was to the left.

Laboratory:

Spinal Fluid examination showed a pressure of 18 mm. Hg., cell count of 2, protein of 11, Kline colloidal gold 0012110000.

Blood studies were normal.

Urine negative.

X-rays negative. (Lipiodol studies not made.)

Diagnosis: 1. On admission was multiple sclerosis.

2. Pre-operatively--Right Medullary tumor.

Operation: Left occipital trephine and needle into the lateral ventricle at the depth of 5cm. Free flow of considerable fluid under pressure. Midline suboccipital incision, lateral detachment of muscles and removal of bone and first cervical arch. Two trephines over cerebellum and wide area of bone removed with rongeur, including first cervical arch. The explored dura was quite tense, although needle in left ventricle was checked for free flow. Incision over low cisterna region with free flow of fluid, and the cord closely in contact with dura. Pressure still not much relieved over cerebellum with more bulging on the right side. Incision through dura over both cerebellar lobes extended radially with disturbing cerebellar herniation through these openings.

Dura finally opened widely across midline and down midline to the second cervical vertebrae. The right cerebellar lobe appeared larger than the left and retraction at the lateral inferior angle disclosed a rounded moderately firm tumor. An incision was made into this tumor for curettage but the tumor was too firm for this procedure. A dark cystic membrane was seen about 5 mm. below the surface which was perforated and considerable fluid escaped marked by an uncertain blood color. There still was disturbing herniation of cerebellum, making tumor exploration difficult. Dissection was attempted medially and the appearance was

of extension into the medulla. The arch of the second cervical vertebrae was removed, normal cord identified at this level and a fusiform enlargement of cord and medulla seen extending into tumor leading to diagnosis of intra-medullary tumor, not an external neurinoma. A small lateral portion of rather firm tumor tissue was removed by sharp dissection and curettage. Bleeding was satisfactorily stopped by silver clips, deep muscle and fascia. Closed with interrupted silk. Moderate tendency of cerebellar lobes to herniate indicating unexplored tumor beneath medulla. Ventricular needle removed and cotton and collodian dressing applied.

**Autopsy:**

On the right side of the medulla oblongata is a tumor approximately 3.5 X 2 X 1 cm. The upper border of the tumor lies at the edge of the Pons. The tumor is partly cystic in appearance but otherwise of very firm consistency. Diagnosis: Astrocytoma.

Microscopic sections of both the surgical specimen and the specimen obtained at autopsy showed only a dense fibrous tissue thruout which may be seen relatively few spindle shaped cells. Diagnosis: Neurinoma.



Case II. E. B. a colored housewife, age 38, entered the Creighton Memorial St. Joseph's hospital 7/5/38 complaining of stiffness and soreness over her entire body, numbness of the entire body, extreme weakness of the entire left side of her body (from neck down) all of three weeks duration. She also complained of a sense of heaviness on the shoulders of 6-7 months duration, aches and pains in the back of the head, neck and shoulders of 1 year duration, constipation and a sense of epigastric fullness whenever she eats any food of two weeks duration.

Patient states that 2-3 years ago she began to notice stiffness of the neck and different parts of her body when she arose from bed in the mornings. This stiffness and soreness has been increasing slowly until the last three weeks when it has been severe. In the last three years her entire left side has been weak, this has progressed until at the present time she is unable to lift the left arm or leg off the bed. Patient states that she was here in June, 1937 and had her tonsils removed in hopes that her condition would improve but the condition became progressively worse.

Past history:

Pleurisy 7 years ago, recurrences and exacerbations for 7 years. Scarlet fever when 10 years of age and measles when a child. On 2/13/35 the patient was admitted to the hospital complaining of pain in the neck radiating to the shoulders, of 2 days duration and stiff-

ness of the neck of 1 year duration. A diagnosis of stricture of the rectum was made, treatment was medical and patient was dismissed 3/9/35 improved. On 5/21/37 patient was again admitted to the hospital with complaints of stiffness of the neck and shoulders of 2 years duration. At this time the diagnosis was torticollis, fibromyositis due to focal infection. The tonsils were removed and the patient dismissed 6/7/37 improved.

Physical Examination: Blood pressure 140/60, eyes react to light and accommodation directly and consensually, ophthalmoscopic examination showed no evidence of papillo-edema or retinal sclerosis, ears negative; mouth showed marked dental caries and pyorrhea alveolaris; heart negative; lungs negative; abdomen moderately distended with fluid and liver 1-2 fingers below costal margin. Atrophy of muscles of entire left side of body, excluding head; reflexes--right and left were hyper-active; abdominals absent. Babinski (positive) on right and left. Unable to discriminate between hot and cold at all on right side of body and only vague discrimination on the left. Practically a complete flaccid paralysis on left side of body from neck down.

Laboratory:

Urine negative.

Blood: Hb. 61%, R. B. C. 3,810,000, W.B.C. 5,500.

Spinal Fluid: Xanthochromic, Queckenstedt positive,

Pressure 13 mm. Hg., Total Protein 154.6 mgm.%,

Wassermann negative.

X-ray: Negative.

Pre-operative Diagnosis: Extramedullary Tumor of Cervical Cord.

Operation: Cervical Laminectomy, Exploration of Cervical cord and partial removal of Tumor.

Under local novocain anesthesia a linear incision was made in the midline posteriorly over the second through the seventh spinous processes of the cervical vertebrae. The deep cervical muscles were separated from their attachments to spinous processes and laminae throughout the length of the incision. Spinous processes and laminae of the fourth, fifth and sixth vertebrae were removed thus exposing the dura. Palpation revealed that the dura and underlying cord were more firm and slightly larger in the upper two-thirds of the opening than in the lower. A small median posterior incision was made in the dura with no escape of spinal fluid. The incision was extended in the midline by blunt dissection superiorly to the upper angle of the opening. The cord thus uncovered, appeared somewhat fusiformly enlarged, and a small incision was made in the median dorsal raphe of the cord to the depth of 3 mm. without encountering any abnormal tissue. To increase the exposure and facilitate exploration, spinous process and laminae of the seventh cervical vertebrae were removed by rongeur, and the dura was opened in the midline. Exploration revealed a dark, grayish-red soft tumor mass lying anteriorly and

slightly to the left and extending from the level of the sixth cervical nerve roots cephalad to the limits of the exposed area. The sixth nerve roots were obviously distorted and displaced posteriorly by the mass. Delivery was accomplished with difficulty and the tumor was removed piece-meal. Were sacrifice of the nerve roots possible without their attendant residual loss, the procedure might have been accomplished more easily, and more completely. It was impossible to tell for certain that the tumor was extra-medullary at this level, but there was no evidence of communication with the cord substance. Spinous processes of the third and second vertebrae together with their laminae were next removed, and the dura underlying them was opened. Tumor was found in the same relative position extending upward through this level also, and piece-meal removal was again resorted to. Although it was felt likely that the tumor extended even higher and possibly even above the foramen magnum, it was considered inadvisable to do any more extensive surgery because of the considerable trauma already given to the cord, and because of the proximity to respiratory and cardiac centers. After repeated flushings with normal saline, and after satisfactory hemostasis was accomplished by means of packing and silver clips, the wound was closed in layers. The dura was left widely open, and muscles were approximated over it.

Number 2 chromic catgut was used in closing deep fascia and in suturing muscles. Interrupted silk suture was placed in the subcutaneous tissues, and skin was approximated with locked continuous silk suture. A simple dressing was applied. The post-operative diagnosis was the same as the preoperative.

**Autopsy:** Several hours post-operative.

There is a long cystic tumor extending from the foramen magnum down to about the fourth cervical segment on the anterior and left lateral surface of the cord pushing the cord posteriorly and to the right with moderate compression about the level of the fourth cervical segment. The tumor is intradurally and extramedullary. It extends posteriorly as far as the dorsal roots and it is not attached to them. It surrounds but apparently does not involve the left anterior roots. It is quite densely adherent to the pia anteriorly and also by a small pedicle to the dura in the midline at the level of the second cervical segment. The point of origin is not definitely determined. The tumor is soft, cystic and yellowish and contains bloody fluid and blood clot.

Microscopic sections of the cord tumor are composed of two types of tissue. The first type being that of palisaded neurinoma commonly seen in peripheral nerve tumors while the other type, which is more prominent is cellular reticular type of tissue referred to

by Antoni as Type B. The palisaded areas exhibit considerable whorling in places. There is no calcification however. In the type B areas there is some pleomorphism and an occasional large binucleated cell. With Mason's tri-chrome stain the reticular tissue stains bluish as so the fibers in the palisaded areas.

Diagnosis: Intradural, extramedullary differentiated neurinoma, arising from a spinal nerve in the cervical region.

Case III L. A. a white school boy age 10, entered Dr. Keegan's care September 11, 1935, with the following story. At the age of three or four a right internal strabismus was noted and glasses were fitted with some benefit. At about this time his mother noted that his mental capacity seemed to be slightly impaired, particularly in the quickness with which he remembered rhymes. At the age of five years he rolled out of the door of an automobile traveling about thirty-five miles an hour but showed no concussion symptoms and appeared to suffer only some scratches on the back of the head. About a year after this, which was three years before coming to Dr. Keegan, the father first noted right facial weakness although he had previously observed delay in development of the right teeth and maxillary processes. He consulted a M. D. who suggested the possibility of brain tumor and recommended further

study at the Mayo Clinic. Instead the father was reassured by some osteopathic advice and during the following year this treatment was given. His mental retardation in school continued and about two years ago some unsteadiness in his gait was observed. Potassium Iodide treatment was recommended by an optometrist who was keeping track of the eyes and there seemed to be some improvement. Recently the optometrist noted the appearance of some papillo-edema and sent the parents to Dr. Keegan for neurological studies.

**Neurological Findings:** Show a rather large boy with a noticeably large head, internal strabismus and right face weakness and with some unsteadiness in gait. The cranial nerves show an optic papillo-edema of three diopters in each eye, no visual field defect and very slight visual impairment in the right eye. There was bilateral external rectus weakness more marked on the right. Nystagmus was present and there was motor paralysis of the right fifth nerve, no sensory loss. Right seventh nerve showed a peripheral type of paralysis but with no loss of taste. The right eighth nerve showed slight reduction in hearing. There was evident paralysis of the right ninth and tenth nerves and the left twelfth. There was some ataxia in the upper extremities and a positive Romberg. Sensory, motor and reflex tests otherwise were essentially normal. At one time a questionable positive right plantar reflex was obtained.

Spinal pressure measured 18-20 mm. of Hg., the fluid serology was negative. X-rays of the skull showed mottling of chronic internal pressure and some separation of the suture lines, absorption of the clinoids but no other significant findings. Ventriculogram and Encephalogram did not seem warranted due to slight herniation symptoms following spinal puncture and because of seemingly positive localization in the posterior fossae as shown by cranial nerve loss.

**Operations (Done at Methodist Hospital)**

A satisfactory cerebellar exposure was obtained without troublesome bleeding or herniation. The right cerebellar lobe appeared slightly larger than the left, the midline was displaced slightly to the left and the inferior poles were herniated into the foramen magnum. A small area of smooth encapsulated rather firm tumor appeared at the lower border of the right cerebellar lobe. Blunt dissection easily separated the cerebellum from this tumor and disclosed a rather large, firm, nodular growth, estimated five by four centimeters. It was about four centimeters in depth as well. The greatest diameter was transverse and the medial portion extended well across the midline beneath the cerebellum and over the medulla which could be seen well outlined at the inferior border. At first it was thought that this portion of the tumor could be lifted free from the medulla, but it seemed firmly attached in this direction



on the under surface and further blunt dissection was not possible. Laterally the tumor extended well toward the angle and although free was so firmly attached and rounded that a view of its undersurface could not be obtained. After ligation of a few surrounding vessels a considerable portion of the tumor was removed with the electrosurgical loop. Laterally a nodule of the tumor could be seen extending between and below nerve root fibers, probably the eleventh nerve as shoulder contraction occurred from stimulation in this region. Medially the tumor base still remained firmly attached over the medulla and removal was discontinued when a suspected medullary shock reaction was noted. Bleeding was controlled with some difficulty, although the pulse and blood pressure remained fairly good. Respiration, however, had developed into more labored type and the patient did not rouse much during closure of the wound. A transfusion was given and the pulse and respiration were of fair quality on return to the room. However, the patient appeared to be rather deeply unconscious not attributable to remaining anesthetic. The respirations became poorer in quality, pulse and temperature rose and death came at 5:00 P. M. that day.

**Pathology report:**

Autopsy was not done but the microscopic sections of the tumor showed a polar spongioblastoma and was considered in the astrocytoma group.

Case IV.

V. L. a white male laborer, age 39 entered the University hospital for the fourth time Dec. 1932 being advised to do so by Dr. Keegan. A summary of his history is as follows:

In 1913 the patient fell from a house and struck on his rump. He discontinued work only for one-half hour and worked the remainder of the summer. In the fall of 1913, he noticed some pain in back which was not severe enough to prevent his working. These pains cleared up at times. In 1916 the patient suffered another fall following which he had occasional pain in the hips. This pain was relieved by a sacro-iliac belt. The patient enjoyed good health up until 1926 when pains in his back began to get more severe with recurrent pain in his hips. In 1927 during the month of February the patient was forced to give up work because his pain was aggravated by work. In the fall of 1927, pain of a severe nature began to occur in his neck which had persisted most of the time until the present entrance. (1932)

In April of 1928 the patient entered the University hospital for the first time complaining of stiffness and pain in the neck with less severe pain in his back and hips. Examination at this time was negative except for local pain and tenderness in the neck. X-ray was suggestive of Cervical spondylitis. The treatment insti-

tuted at this time was partial alveolectomy and use of a Minerva cast for several months. He was dismissed from the hospital June 1928.

In June 1929 the patient was readmitted complaining of pain in the right occipital region. Examination showed local pain and tenderness of the neck with a fluctuating mass of the right side of the neck. The neck muscles on the right were spastic. The mass proved to be a lipoma. X-ray and Laboratory revealed no further findings. The diagnosis of Cervical arthritis was made and the patient received physical therapy and head traction. He was dismissed from the hospital in July of 1929.

On July 31, 1929 the patient entered the University hospital for the third time with practically the same complaints as on previous admissions. X-ray at this time showed cervical dislocation and enlargement of the foramen magnum. Spinal fluid was negative. Tendon reflexes were hyperactive and ankle clonus was present both right and left. Hoffman positive on the right. Paresthesia of the inner left thigh. Pain and temperature loss in inner left thigh area. Vibration and touch unimpaired. Exploratory operation was advised and done. Cervical laminectomy and section of the second, third and fourth dorsal roots was done. The diagnosis preoperatively was cervical spondylitis with cord pressure involving the right second, third and

and fourth cervical roots. Post-operative diagnosis was the same except that the pathology was known to be anterior to the cord because at operation posterior bowing of the cord was seen. November 12, 1929 a second operation was done with removal of the posterior border of the foramen magnum and the arch of the first cervical vertebrae. The patient improved greatly and was dismissed March 7, 1930. Dribbling and rectal incontinence had been present since the first operation.

For the first year the patient improved greatly but from then on began to go down hill. This continued to be the case until the admission December 1932.

**Examination:**

No Cranial nerve involvement.

Head and neck rigid with marked torticollis to right.

Ataxia of the left arm.

Reflexes: Abdominal present and equal. Knee Jerks hyperactive. Bilateral ankle clonus. Babinski positive on the left. Sphincters spastic.

Diagnosis: Cervical spondylitis with sclerosis of the cord. Chronic pleuritis.

Dismissed from the University Hospital 12/17/32 to enter the Immanuel Hospital on the same day. Here he died 1/2/33.

Autopsy: Upon exposing the medulla and cord a bony process was found to have passed through the right mid-portion of the cord at the lower part of the decussation

of the pyramids. This bony process was identified as the odontoid process of the second cervical vertebrae which was in an almost transverse anterior posterior extending posterior to the margin of the foramen magnum. Death was caused by multibronchiectatic abscesses.

**Comment:**

The four above cases are interesting from the viewpoint of diagnosis and management. It is obvious that observation over quite a period of time is usually necessary before making a diagnosis of pathology in the region of the foramen magnum.

The fracture of the odontoid process of the second cervical vertebrae, Case IV, was reported in full because it somewhat parallels cases 13, 21, 43, and 51 which concern pathology of the odontoid process and may be found and studied on the chart in the following part of this paper. These cases are considered with others found on the chart in the following discussion.

## REVIEW OF THE LITERATURE

The chart on the following page is the method chosen for reviewing the literature because of the advantages in getting a clear picture of the various possibilities in the syndrome of tumor at the foramen magnum. Since no magnifying glass accompanies the greatly reduced chart, the following explanatory remarks may be in order.

The discussion in the remainder of this paper is taken directly from the chart on the following page. The first column represents the bibliography number of the various case studies. The second column is merely the number given to a particular case on the chart. Age, Sex, Occupation and Trauma are considered in the next four columns in that order. The next column is devoted to type of pathology with the use of the following key: M-Meningioma; N-Neurinoma; O-Osteoma; H-Hemangioma; D-Dermoid Cyst; E-Ependymoma; Astrocytoma and others written out in full. Relation to the cord is next considered: A-Anterior; AL-Anterior Lateral; P-Posterior; PL-Posterior Lateral; R and L, when used, refer either to right or left. The extent of tumor is charted both as to high and low level; F in this column refers to the foramen magnum. Duration of symptoms, Early symptoms, Late symptoms, Examination, Cerebrospinal fluid, X-ray studies plain and with lipiodol, Surgery and Remarks are considered in separate columns in the order above.



Year	Month	Day	Sex	Profession	Age	Onset	Duration	Course	Site	Signs	Diagnosis	Pathology	Prognosis	Outcome	Notes														
1931	14	M	G	Teacher	M	1931	1-1931	2-R	1931	Weakness of Tongue	Quadraplegia	Typhoid Vertigo Enuresis	VII R VI R & L VII R & L	Atrophy of Both hands	L R Partial	R T	3+ P 2 neg 3 neg	P neg L +	✓	Recovery Spine-Cranial									
1931	30	34	F	Housewife	M	1931	1-1931	2-R	1931	L. abdominal Pain	Quadraplegia	Numbness				R T	L + by Cistern	✓	Death from Stn. operation Cranio-Spinal										
1931	10	F	Factory Worker	M	RAL	C2	1931	1-1932	2-	Pain radiated Shoulder	L Paraplegia	Torticollis		Atrophy of L hand & Fore L	↑ R	↑ Left	L +	✓	Recovery Cranio-Spinal										
1932	30	34	F		M	RAL	C5	1932	1-1922	2-							✓	Recovery Cranio-Spinal											
1933					D.	P	C2	20m	1-R									✓	Recovery Cranio-Spinal										
1934					M	P	C9	16m		Pain in shoulder			VII Slight VIII					✓	Recovery Spine-Cranial										
1934					A	RAL	C5	F	1				V.R.M.S.					✓	Death Spine-Cranial										
1936					M	LPL	C2	10m							General +			✓	Recovery Spine-Cranial										
1937					M	AL	C1	↑F	1									✓	Death Autopsy Cranio-Spinal										
1938					M	AL	C3	↑F	1									✓	Recovery Cranio-Spinal										
1939	14	F	Schoolgirl	L	RAL	F	↑F	Max. Sept. 1927	1928	1	3R Hand	Unsteady Gait	Quadraplegia	Urine Retent. Speech diff.	VIII	Not Marked				✓	Death Autopsy Cranio-Spinal								
1939	40	34	F		L	RAL	↑F	Child 1927	1	3R Hand	Wasting	Vertigo	1929	Dysarthria	VIII IX X XI XII	Atrophy R Hand				✓	Death (Presumed) Autopsy Cranio-Spinal								
1941	12	29	F		H	PL	At Foremen	1934	1936	1	3R Hand	Wasting	Vertigo	Weakness	Walks to left						✓	Recovery New + re-linguam Spine-Cranial							
1942	25	50	F	Housewife	M	AL	C2	↑F	1928	1932	2	1	Pain c Straining	Quadraplegia			Weakness of L & atrophy	↑ vid	↑ Lc Lt	✓	Recovery after a Max. Cr. Cranio-Spinal								
1943			M		O	A.	At Foremen	1919	1920					Loss of Sphincter Control	Spastic Gait	Dyspnea Priapism	VIII L II Blurred Vise	Weakness of L neck mus.	↑ +	↑ Left R+		✓	Death Autopsy Osteoma of Olfact.						
1944	24	M		H	P.	↓F	↑F	1911	1913	1				Number of L. 3 Hand 2 Feet	Occurred from the start Nausea 2 Vom.	L. Early	2. Vertigo	Unsteady Gait	H VI VII VIII	Quadraplegia Atrophy L Hand	6 vid L	6 L	✓	1913 L Autopsy Cranio-Spinal					
1945	36	M	Truckman	H	P.	↓F	↑F	1920	1922	1							Early	Unsteady	H VIII	Ataxia	6 L	Phon+	✓	1913 L Recovery					
1946	48	M	Salesman	H	P.	↓F	↑F	1922	1923	2							L Early	Dropped Articles from L. Hand	Ataxia	J	L Tone +	6 L	↑ R & L E Clonus	✓	1923 Apr 1923 Cranio-Spinal				
1947	40	F		M	LAL	C1	↑F	1932	1933	1							1933 Numbness 2-L. Arm	1	Stiffness of Neck	Weakness L. Leg	Incontinence after onset	IX XI XII	L. Atrophy Twitching Tendon Dist.	6 R	+	✓	Death 182 p.o.		
1948	42	M	Clerk	M	LAL	C1	↑F	1928	1933	2							R. + L.	1	Twitching of fingers.	L. Leg Weak	Girdle Sens. of Chest	XII	1923	6 vid Late	6 R Lt	✓	Death Tumor found Treated for 4 mos. Cranio-Spinal		
1949	40	F	Housewife	E	PL	C3	↑F	1928	1931	3	Paired							Distum c			Torticollis			6 vid erg+	6 L	✓	Still in Hosp. L. too medullary		
1950	19	F		N	RAL	C1	10m	Max. Nov. 1931	1931	1											Intermittent	Weakness of Upper 9 Lower Lt	1 final Quadraplegia	XI	Atrophy S.C.M.	6 R & L	+	✓	Recovery Bone Graft P.O. Cranio-Spinal
1951	14	M	Schoolboy	O	A	At Foremen	3 Years														1-Weakness 2-Dysphagia	3-Dyspnea 4-Priapism	VIII XI	General Impairment	Not Definite	6 vid	6 R Lt	✓	Death Autopsy Osteoma of Olfact.
1952	42	F		M	RPL	C1	20m	3 Years	1-1929												Remittent	Weakness 1932			6 R & L	6 R	✓	Recovery Spine-Cranial	



## AGE.

Of the series of cases reviewed there were only forty in which the age of the patient was given. Of these forty the older of the cases were reported by: Muller (31) who reported one case which occurred in a female age sixty-two; Guttman (29) told of a case in a female age fifty-nine; and Symonds and Meadows(49) reported a tumor in a female age fifty-seven. All of these were meningiomas.

The younger cases reported were as follows: Abrahamson and Grossman (1), Bouttier Bertrand and Mathieu(7) and Misch (35) each reported cases in fourteen year old patients; they were, in the order above, osteoma, glioma and lipoma. Adson and Ghormely (3) reported a neurinoma in a young girl of nineteen. Abrahamson and Grossman (1) gave the account of a meningioma occurring in a twenty-four year old female.

The average age of the whole series of cases reviewed was thirty-eight years. This fact show that this type of pathology occurs during the productive period of life.

## SEX.

Of the forty-six cases where the age of the patient was reported thirty-one or sixty-seven per cent occurred in females. This fact was brought out in discussions of spinal cord tumors as a whole, Elsberg (21), but was not mentioned by any authors regarding pathology occurring in region of the foramen magnum.

## OCCUPATION.

This was not reported in enough cases to draw any significant conclusions. It is interesting to note, however, that both of the cases reported by Cushing(16) occurred in hairdressers.

## TYPE OF TUMOR.

In studying the incidence of the types of tumor occurring in this region it is interesting to note that ninety-two per cent were extramedullary while the other eight per cent were intramedullary.

Of the ninety-two percent which occurred extramedullary: sixty two percent were meningiomas, six percent were neurinomas, twelve percent were hemangiomas, six percent were osteomas, four percent were dermoids and four percent were lipomas; of the eight percent which occurred intramedullary six percent were gliomas and two percent ependymomas.

The chart may be referred to for a bibliography to the original case reports of these various tumors. On the chart they are abbreviated by their initials.

## TRAUMA.

This factor though insignificant was seen in cases reported by Muller (36), Symonds and Meadows (49), Abrahamson and Grossman (1) and Misch (35) There forty-eight cases in which no history of trauma was elicited.

#### RELATION TO THE CORD.

Of the fifty cases where the relationship of the tumor to the spinal cord was given twenty-three were in the anterior or anterior-lateral position, twenty-three were in the posterior or posterior-lateral position and four were intramedullary. Cushing (16) says that those which are anterior or anterior-lateral are more apt to have arisen from the posterior fossae and extended into the spinal canal, while those which lie posterior or posterior-lateral are more apt to have arisen from the spinal canal and extended into the posterior fossae; to the former he gives the name cranio-spinal and to the latter the name spino-cranial. Elsberg and Strauss (22) call these tumors either supra-foraminal or infra-foraminal depending upon the relationship of the point from which they arise to the foramen magnum.

#### EXTENT OF THE TUMOR.

In reporting the level at which tumors in this region lie most authors are rather indefinite as to the upper limits of the tumor while the lower level is usually definitely reported. This is probably due to ease with which the lower level may be compared to adjacent bony landmarks and to the lack of such discreet landmarks at the upper level as well as to the difficult exposure of the upper level in some cases.

Of the cases reviewed the lowest level reported was in a tumor reported by Abrahamson and Grossman (1) which extended down the canal to the level of the sixth cervical vertebrae. This compares with Case II reported in full in the first part of this paper.

The highest extent was reported by Cushing (16) as extending 3.5 cm. above the level of the foramen magnum. Schreiber (44) reported one case which extended to the cerebellar-pontine angle. (a variable landmark)

The level of the tumor is important from the standpoint of the surgical attack to be made as well as in determining somewhat the sequence of events before and after surgery.

#### EARLY SYMPTOMS.

Careful consideration of the early symptoms is necessary if the presence of pathology in this area is to be determined early.

The most common early symptom is that of suboccipital pain with or without headache. The pain is usually on the ipsilateral side as the tumor and may remain localized or radiate to the temporal or even to the frontal area. This was noticed and reported as the first symptom in cases of Cushing and Eisenhardt (16), Rhein (39), Guttman (29), Symonds and Meadows (49), Abrahamson and Grossman (1), Rubenstein (43), Muller (36), Schreiber (44), Elsberg (21) Misch (35), Cushing and Bailey (15) and Soltz and Jervis(45).

In twenty-four of the fifty-two cases reviewed this was the first symptom noted and was present in many other cases before there was any appreciable weakness.

The second most common symptom was the occurrence of paresthesias usually in the ipsilateral hand and arm or shoulder and side of the neck but occasionally on the contralateral side. This has been noted as the first symptom in ten of the fifty-two cases reviewed. The description of the paresthesia varies a great deal and was described by different patients to various authors as follows: Numbness in cases reported by Cushing and Eisenhardt (16), Abrahamson and Grossman(1), Globus and Strauss (27), Rubenstein (43), Cushing and Bailey (15) and Soltz and Jervis (45); Pinching pain in the ipsilateral hand by Favill, Faxson and Palmer (23); Tingling sensations reported by Symonds and Meadows (49) and Globus and Strauss (27); Sensations of cold reported by Symonds and Meadows (49); Prickly sensations in cases of Symonds and Meadows (49)and Abrahamson and Grossman (1).

The third most common symptom was that of Gastro-intestinal disturbances and was found by the following: Hemorrhage, reported by Cushing and Eisenhardt (16); Pulling sensations of the stomach, by Favill, Faxson and Palmer (23); Nausea and vomiting was reported in cases of Abrahamson and Grossman (1), Rubenstein (43) Heuer (30), David (18), Cushing and Bailey (15) and Soltz and Jervis (45).

Early loss of sphincter control though not a common early symptom, interestingly occurred as the first symptom in both cases of osteoma of the odontoid process of the second cervical vertebrae. One of these cases was reported by Abrahamson and Grossman (1); the other by Krauss and Silverman (31).

The inability to identify objects by touch was the first symptom in a case reported by Rubenstein (43). This was found objectively in several other cases. Those reporting such cases are as follows: Cushing and Eisenhardt (16); Symonds and Meadows (49); Abrahamson and Grossman (1); Globus and Strauss (27); Soltz and Jervis (45); Rubenstein (43); Bouttier, Bertrand and Mathieu (7); Roussy and Levy (42); Cushing and Bailey (15); and Freidman (25).

These early symptoms are interesting in that they usually occur on the ipsilateral side and are subject to remission as may be seen in cases reported by the following: Muller (36); Abrahamson and Grossman (1); Adson (2); and Martin and Bogaert (33). Increased intensity of various symptoms with straining is the rule and is found in cases of Symonds and Meadows (49); Rubenstein (43); Abrahamson and Grossman (1) and Others.

## LATE SYMPTOMS

The early symptom usually progress to weakness in the extremity which was first affected with the paresthesia or on the side where suboccipital pain was first noticed. If these were more or less bilateral the weakness is apt to be bilateral. The upper extremity is the most frequently involved, but sooner or later the ipsilateral leg becomes involved and difficulty in walking due to a hemiplegia is noted. Gait difficulties as a late symptom is noted in cases reported by Rubenstein (43), Symonds and Meadows (49), Favill Faxson and Palmer (23), Krauss and Silverman (31) and Cushing and Bailey (15).

Other late symptoms are the progression of the hemiplegia to a triplegia and in cases where diagnosis was not made or where surgery was not done this triplegia finally became a quadraplegia. Some form of paralysis was noted in every case reviewed.

Dyspnea was another quite frequent symptom this was proven to be caused by diaphragmatic paralysis which was in turn caused by pressure upon the phrenic roots or directly upon the respiratory center. Such cases were noted by Cushing and Eisenhardt (16), Borgorodinsky (8), David (18), Muller (36), Elsberg and Strauss (21) and Freidman (25). Paralysis was usually on one side only but was observed to be bilateral in one case of Symonds and Meadows (49).

EXAMINATION

CRANIAL NERVES

The following chart gives a summary of the cranial nerve finding in the whole series of cases.

Cranial Nerve	II	V	VI	VII	Nystagmus	IX	X	XI	XII
Case No.	2						3	3	3
						5	5	5	5
					7	7		7	7
						9			9
					12				
				14	14	14	14	14	
					15				
					16				
				18				18	
	19			19	19				
					21				
					22	22			22
						25			25
						26			26
	27								
				30		30		30	30
				34	34				
	35								
					38				
				39	39	39			39
					40	40			40
	43				43				
	44		44		44	44			44
	45				45				
	46								
						47		47	47
									48
								50	
					51			51	
Totals	<u>6</u>	<u>2</u>	<u>1</u>	<u>6</u>	<u>16</u>	<u>13</u>	<u>3</u>	<u>8</u>	<u>13</u>

Besides nystagmus, which was discussed with the cranial nerve findings, there were twenty-three cases out of the fifty-two cases reviewed which showed some



cranial nerve involvement.

In twelve cases a partial Horner's syndrome was found. That this occurred in only those cases in which the tumor lay anterior or anterior-lateral to the cord may be of some significance. In most of the cases this was found objectively.

#### MOTOR SYSTEM.

Outside of the findings which, because of the symptoms, were obvious to the examiner; that is hemiplegia, paraplegia etc.; the following are the most significant findings of involvement of the motor system.

Atrophy of the intrinsic muscles of the hand was found in twelve cases. This indicates lower motor neurone damage and was reported by Cushing and Eisenhardt (16); Rhein (39); Favill, Faxson and Palmer (23); Muller (36); Borgorodinsky (8); Symonds and Meadows (49); Abrahamson and Grossman (1); Elsberg (21); Misch (35); Oldberg (38); Cushing and Bailey (15); and Soltz and Jervis (45).

Atrophy of the shoulder girdle was found in a case reported by Abrahamson and Grossman (1). Weakness of the ipsilateral shoulder was a frequent finding.

#### SENSORY FINDINGS.

Tactile sensation when reported was not disturbed in but few cases. When disturbed it was usually only of the

dermatome corresponding to the lesion. Lack of tactile loss was prominent in the whole series of cases and is the chief reason for considering it here.

Pain and Temperature disturbances were found in most every case reviewed. The degree of involvement ranged from patchy, indefinite involvement to complete loss on the contralateral side below the lesion. The ipsilateral side, when involved, was not affected until late in the course of the disease.

Proprioceptive sensation was impaired to a great extent in a number of cases. It was usually impaired in the upper extremity on the ipsilateral side early and often progressed to involve the ipsilateral lower extremity at a later date. Cases in which proprioceptive sense was disturbed may be obtained from the chart on page 19.

Positive Romberg's were reported in cases of Cushing and Eizenhardt (16), Cushing and Bailey (15) and Soltz and Jervis (45).

#### REFLEXES.

The tendon reflexes, in most of the cases, were hyperactive but usually to a greater degree on the ipsilateral side. In thirty cases out of the fifty-two considered in this review tendon reflexes were increased. Clonus accompanied this increase in cases reported by:

Symonds and Meadows (49); Abrahamson and Grossman (1); David (18); Oldberg (38) and Cushing and Bailey (15).

Thirty-five cases showed a positive Babinski. When positive on one side only it was usually on the ipsilateral side and was reported by nearly every author who reported a case.

Absent or decreased abdominal reflexes were observed in twenty cases. Differences in the two sides were reported by Symonds and Meadows (49) in two cases.

#### SPHINCTER DISTURBANCE.

Urinary control was either lost or diminished in fourteen cases which were reported by; Cushing and Eisenhardt (16) reported two cases one of which showed loss post-operatively; Symonds and Meadows (49); Abrahamson and Grossman (1); Rubenstein (43); Muller (36); Elsberg (21); Misch (35); Soltz and Jervis (45); and Freidman (25). This loss, as a rule, did not occur until late in the disease.

Rectal incontinence was reported in one case by Favill, Faxson and Palmer (23); Disturbance in function such as constipation was seen more often in cases of Cushing and Eisenhardt (16), Abrahamson and Grossman (1), and Misch (35)

#### SPINE.

Upper cervical spine was involved in thirty-two of

the fifty-two cases reviewed. This involvement consisted of rigidity; which gave the patient a characteristic carriage or attitude; or of tenderness or both. These were among the findings reported in cases of: Cushing and Eisenhardt (16); Rhein (39); Symonds and Meadows (49); Abrahamson and Grossman (1); Globus and Strauss (27); David (18); Rubenstein (43); Roussy and Levy (42); Elsberg (21); Misch (35); Cushing and Bailey (15); Soltz and Jervis (45) Freidman (25 ); and Martin and Bogaert (33).

#### LABORATORY

Spinal puncture was done in twenty-six of the fifty-two cases. Increased protein was found in cases reported by Symonds and Meadows (49), Rubenstein (43), Elsberg (21) Misch (35) and Freidman (25). Pressure was increased in cases reported by Freidman (25), Adson (2), Soltz and Jervis (45) and Misch (35). A positive or at least suggestive Queckenstedt was reported in cases of Symonds and Meadows (49), Globus and Strauss (27), Rubenstein (43), Oldberg (38) and Soltz and Jervis (45).

Plain X-rays were taken in twenty cases and in only cases reported by Soltz and Jervis (45) and by Cushing and Bailey (15) were the findings suggestive.

X-ray studies with lipiodol injections were used in seven cases and in five cases findings were definite while

in the other two cases the results were misleading. Symonds and Meadows (49) injected with lipiodol and in one of their cases the results were misleading in that the oil stopped at a lower level than the location of the tumor. Of two cases reported by Globus and Strauss (27) one was definitely localized and the other "due to the feel of the cisternal needle upon injection of the lipiodol" was thought to be located in the region of the foramen magnum. Elsberg (21) reports the use of lipiodol with varying degrees of success.

Ventriculograms were resorted to in cases reported by Symonds and Meadows (49), Rubenstein (43) and by David (18). The negative results obtained by the above in each case was thought to be significant and beneficial in that intraventricular and parietal lobe pathology was ruled out.

#### DIAGNOSIS.

The diagnosis of tumor in the region of the foramen magnum is not readily made in the early stages of the disease. It must be differentiated from the following: Amyotrophic lateral sclerosis; dorsi-lateral sclerosis; multiple sclerosis; chronic adhesive arachnoiditis; aneurysms and varicosities of the spinal vessels, herniation produced by other intracranial neoplasms, and metastatic Ca from other points.

Amyotrophic lateral sclerosis simulates upper cervi-

cal cord neoplasm and was seen as the differential in a case reported by Soltz and Jervis (45). They state that the facts of moderate change in deep sensibility and subjective pain aided them in making the differentiation.

Freidman (25) reported in detail cases which only after months of study was he able to differentiate upper cervical tumors from dorsi-lateral sclerosis. This was also noted by Soltz and Jervis (45). Laboratory evidence of questionable or complete block, increased protein, xanthochromia, X-ray studies and cranial nerve findings make differentiation possible.

Cerebellar signs occur with pyramidal tract involvement and make differentiation of cervical cord tumors from multiple sclerosis necessary. This was reported in the chart by Soltz and Jervis (45). Many in the chart had the diagnosis of multiple sclerosis made. Case I of the original cases was so diagnosed. Differentiation may be possible by the type of onset and the progression of symptoms. Demonstration of some degree of block, increased protein in spinal fluid, X-ray studies will help in making the diagnosis.

Chronic adhesive arachnoiditis, varicosities and aneurysms of spinal vessels and high cervical cord tumors are difficult to distinguish from each other, since the symptoms in each case are due to the fact that they all produce their effects by compression. "The clinical signs of radicular involvement of sensory or motor com-

ponents seem to predominate in tumors" Soltz and Jervis (45). Laboratory tests may be resorted to with positive or at least suggestive differentiation.

Cerebellar herniation may produce the same symptoms. The onset is usually more rapid, signs of increased intracranial pressure are greater and ventriculograms may be used with some degree of success. Spinal puncture is dangerous in this type of case. Rhein (39)

It is interesting to note that in addition to the above; metastasis must be considered. Oldberg (38) reported a case which was diagnosed as possible metastatic Cancer from the breast. At surgery a meningioma was removed with recovery.

#### SURGERY

Out of the thirty-three patients going to surgery fourteen had tumors which were posterior or posterior-lateral to the cord. Of these fourteen cases there were eleven recoveries. These were reported by Cushing and Eisenhardt (16), Symonds and Meadows (49), Gardner (26), Elsberg (21), David (18), Soltz and Jervis (45), Cushing and Bailey (15) and Martin and Van Bogaert (33)

The other twenty cases had pathology anterior or anterior-lateral to the cord and of these there were ten recoveries reported as follows: Symonds and Meadows (49) Abrahamson and Grossman (1), Rubenstein (43), Schreiber (44), Elsberg (21), Oldberg (38) and Soltz and Jervis(45)

The average duration of symptoms before death or before surgery which in not a few cases was the same, was three and one-half years.

Following is a note on surgical technique which was written by Julian Taylor and was included in the article of Symonds and Meadows (49). "To expose the foramen magnum it is best to have patient lying on the belly with the neck flexed. The flexion of the neck may embarrass respiration where there is foraminal block and this degree of flexion must be determined in consultation with the anaesthetist before operation is begun, but the technical advantage afforded is so great that the flexed position cannot be abandoned on account of the danger. Scalp and Neck are incised from external occipital protuberance downward and first three cervical arches are removed. The dura is inspected and presence or absence of respiratory pulsation is noted. If it be clearly seen in the sub-occipital region it is certain that a tumor, if present is below the foramen, especially when complete block is previously been demonstrated. Respiratory pulsations of the normal theca in this region is of such amplitude that its observation leaves little opportunity for error, either in recognition or in consequent deductions regarding level of the possible tumor. If a tumor blocking the foramen be thought to be present of the occipital bone is then removed, a fairly extensive craniotomy being



advisable. The dura of the posterior fossae and the spinal theca are then opened above and below the foramen and the incisions joined. The advantage from the large craniectomy is then seen if, as the result of tumor growth in the foramen the normal venous sinuses of the dura be found greatly increased in capacity and number. On opening the dura and exposure of the arachnoid the earlier observation regarding the pulsations is checked. With inspiration and expiration the ebb and flow of fluid through the unobstructed foramen is so freely tidal that there is now no possibility of erroneous deduction, but if pulsation is absent something is unquestionably blocking the foramen and thus if no tumor be seen there must nevertheless be one situated either in the central nervous substance or anteriorly. A cerebellar tumor is at once obvious, but at this stage unless there is an evident lateral displacement of the medulla, it may be difficult to distinguish between a medullary tumor and one anteriorly placed. An anteriorly or anterior-lateral placed growth almost necessarily entails division of the 1st and perhaps the 2nd posterior cervical roots, a step that usually permits the operator to rotate the medulla far enough to gain a view of what lies in front. It will usually afford enough room to enable him to remove the tumor, but the anterior roots and accessory nerve may sometimes be in the way.

The decision to divide even the posterior roots may be a difficult one to make in the case of a patient whose diaphragm and intercostal muscles are paralyzed and whose breathing is precariously dependent upon intact accessory and 2nd cervical nerves.

When a neurofibroma is found it is obviously necessary to remove a part of the affected nerve root, and when an endothelioma is present there are usually adhesions to surrounding nerves, dissection of tumor from which must be carried out with the precision that the situation demands. The removal of an endothelioma may at first appear to present formidable difficulties, but a successful attempt to free the tumor from its dural attachment may render it unexpectedly docile, when removal may be effected without difficulty. It is obvious that the force of manipulation in this region is transmitted undiminished to the medulla and thus that sudden fluctuations in the pulse frequency, blood-pressure and respiration may be experienced. If the operator therefore finds this part of the task anything but effortless or if his attempted division of a dural attachment has failed, he must then proceed to remove pieces from the accessible part of the tumor, in order that the remaining deeper part may be more easily displaced. For this purpose the writer's preference is the use of the knife to that of the diathermic cautery owing to the proximity of the vital medullary centers.

These endotheliomas have not been highly vascularized

and there has been little bleeding on incising the tumors. The bared dura may however, bleed freely from opened veins, but the hemorrhage is easily stopped by a temporary application of muscle grafts. The occipital bone is sometimes invaded by the growth, in which case the operator must decide for himself whether in a life-saving operation of this sort he should undertake the technically difficult, but pathologically correct, procedure of removing affected bone from the anterior or lateral part of the foraminal edge. The bone here normally increased by endotheliomatous invasion. The tumor gone, there appears at once the respiratory ebb and flow of fluid from the posterior fossa; and even if the respiration and pulse have been seriously affected by the manipulations there soon returns a good pulse and regular breathing of satisfactory depth. The wound is then closed in the ordinary way by suture of muscles and skin. As in other procedures in the posterior fossae the operator must be prepared for reactionary depression of respiratory and cardiovascular functions during the hours following operation."

Symonds and Meadows (49) Surgical Note by Julian Taylor.

## CONCLUSIONS

1. Tumors in the region of the foramen magnum present a problem in diagnosis and management. The possibility of their existence should be kept in mind.

2. Diagnosis of such tumors is made possible by careful consideration of the early symptoms and thorough neurological examination with careful interpretation of the neurological findings.

3. Surgery is the only method of treatment and many times the only accurate means of diagnosing the presence of a tumor.

4. Prognosis is better in those tumors lying posterior or posterior-lateral to the spinal cord than in those in the anterior or anterior-lateral position.

FINIS

THE SYNDROME PRODUCED BY TUMORS AT THE FORAMEN MAGNUM

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