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CHRONIC PULMONARY DISEASES AND ASSOCIATED NEUROLOGIC DISTURBANCES*

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In recent years, renewed interest in the neurological manifestations and complications of chronic pulmonary diseases has been aroused. Cameron in reporting the case of a young coalminer with pulmonary emphysema, visual disturbances, and papilledema, appears to have been the first to emphasize the association between these seemingly disparate disorders.¹ More recently, Austen has suggested a clinical syndrome consisting of signs and symptoms of pulmonary and cardiac failure, headache, papilledema, involuntary movements of the extremities, and impairment in the level of consciousness.² Chronic pulmonary insufficiency with its resultant hypoxia and hypercapnia appears to be the direct or indirect influencing factor in the production of these symptoms.

The primary problem of pulmonary insufficiency is common, and the neurologic complications relatively uncommon. The importance of recognizing this association is imperative if unnecessary diagnostic or surgical procedures are to be avoided. Frequently, the neurologic symptoms are so predominant that they may obscure the underlying pulmonary disease. With specific regard to this point Conn reported two cases in which the diagnosis of brain tumor was mistakenly made and in which the degree of the associated pulmonary disease was unrecognized.³ The true nature of the syndrome was revealed by necropsy in both cases. No evidence of an intracranial lesion was found and only signs of increased intracranial pressure were observed.

Also important in the recognition of the above syndrome is the necessity of instituting corrective therapy and, more specifically, that the therapy be directed at the primary underlying disorder. In most instances, measures that have corrected the hypoxia, hypercapnia, cardiac failure, and polycythemia, have resulted in complete remission of neurologic symptoms.

The usual clinical picture observed is that of a middle-aged individual with a longstanding history of chronic asthma, bronchitis, and emphysema, and with progressive development of pulmonary and cardiac failure, headaches, visual disturbances, papilledema, tremor and twitching of the extremities, and impairment of consciousness.

Chronic pulmonary insufficiency is the underlying disorder, but the specific pulmonary disease frequently differs in each case. Chronic bronchitis and asthma are by far the most common etiologic disorders. In none of the reported cases has the subsequent correct diagnosis proved to be brain tumor, either clinically or with post-mortem examination, as in three instances cited by Conn.³ Nevertheless, the differential diagnosis remains difficult, especially in those cases in which signs of increased intracranial pressure persist despite adequate corrective therapy. In these situations, exhaustive efforts must be made to exclude the presence of an intracranial space-occupying lesion.

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Congestive heart failure is present in almost every case. Both sides of the heart may be affected, although right ventricular decompensation is most common and manifested variously by elevated venous pressure, hepatomegaly, peripheral edema, and usually, EKG findings of right axial deviation and right ventricular hypertrophy. Left heart failure is generally indicated by the presence of dyspnea, basal rales, and pulmonary congestion.

Headache is an early and common neurologic symptom occurring in approximately fifty percent of the cases.² It is quite variable in location and may be generalized. It is frequently severe and intense, persistent and steady. Often the headache is nocturnal or occurs in the early morning hours, sometimes awakening the patient from a sound sleep. Nausea and vomiting are at times related to the headaches but are not a frequent feature of the syndrome.

The exact mechanism of headaches in these cases is not clearly established but is probably related to the carbon dioxide retention and resultant cerebral vasodilatation and increased cerebral blood flow, rather than to increased intracranial pressure.^{2,3}

Papilledema is a distinctive feature of the syndrome but cannot be distinguished from other causes of this symptom. It may be unilateral or bilateral, and varies in severity from haziness or blurring of the disc margins, to markedly choking, venous congestion, and retinal hemorrhages. Visual acuity may be significantly impaired but usually is not altered. Papilledema has been reversible in nine cases reported, following improvement of the cardiopulmonary status. The relationship between the severity of the papilledema and elevated cerebrospinal fluid pressure is unpredictable.

Manifestations of cerebral dysfunction vary widely and may include drowsiness and hypersomnolence, or even stupor and coma. The patients are at times forgetful, inattentive, irritable, and easily confused. Often they respond sluggishly to verbal and noxious stimuli and may appear disoriented and incoherent. Evidence of specific cerebral deficits, such as motor and sensory paralysis, aphasia, and visual—spacial disorientation, is usually lacking.

Tremor and twitching of the extremities are conspicuous and characteristic findings in the more recently reported cases.² The tremor is typically of the action type, demonstrated in the outstretched fingers, persisting throughout voluntary movement, and aggravated slightly with effort. The coarse twitching movement of the extremities is reported identical to that seen in hepatic coma, and is arrhythmic and asynchronous. It is usually demonstrated with the arms outstretched and the fingers and wrist in dorsiflexion. Similar twitching of the legs, eyelids, and mouth, are seen following activation and contraction of the respective muscles.

The exact mechanism of the movement disorder is unknown, but hypercapnia, ammonia intoxication, and acidosis, have been suggested as possible factors in its production. Carbon dioxide retention as a possible causative factor has also been suggested because of the observation of myoclonic movements and coma occurring in some patients with pulmonary insufficiency receiving oxygen therapy.^{4,5} These

Pulmonary Diseases and Neurologic Disturbances

signs usually disappeared when the oxygen was discontinued. Furthermore, rhythmic movements of the extremities have been described in patients receiving carbon dioxide therapy.⁶

Blood ammonia determinations were performed by Austen because of the striking resemblance of the twitching movements to that seen in hepatic coma.² The results were variable, and ammonia intoxication was not definitely established as a cause for this sign. Liver function tests were generally negative, and ammonium chloride administration after the twitching subsided did not necessarily produce its recurrence.

It has been suggested that increased acidosis may account for the twitching. However, this possibility seems less likely since twitching occurs in some patients with pulmonary insufficiency in the absence of acidosis, and in hepatic coma in which alkalosis reportedly occurs. Furthermore, in diabetic acidosis twitching has not been observed.²

The EEG findings are non-specific and consist primarily of increased voltage and a diffuse slowing of the electrical activity in the delta and theta frequencies. However, these findings occur in all types of coma due to metabolic disturbances and are frequently reversible.

In general, the various neurologic manifestations are in some way related to the underlying cardiopulmonary insufficiency with its resultant hypoxia and hypercapnia. However, the mechanisms for the production of the individual disturbances is variable and uncertain.

The occurrence of papilledema has been reported in twenty-five cases of chronic pulmonary insufficiency. The probable factors contributing to the production of this important sign include, 1) increased venous pressure, 2) hypoxia and hypercapnia, and 3) polycythemia.

Elevated venous pressure is known to produce increased intracranial pressure as manifested by the performance of the Queckenstedt maneuver. However, papilledema and other symptoms and signs of increased intracranial pressure are not necessarily produced by increased venous pressure.^{7,8} Evidence to this effect is the absence of papilledema in cases of right heart failure, and in mediastinal tumors producing obstruction of the superior vena cava, despite high venous pressures exceeding four hundred millimeters of water. Furthermore, in the majority of cases of chronic pulmonary insufficiency and papilledema, the venous pressure has been found to be normal or mildly elevated.^{9,10}

Beaumont reported a case of papilledema associated with heart failure and attributed the papilledema to increased pressure in the cranial sinuses resulting in interference of filtration of the arachnoid villi, and a rise of cerebrospinal fluid pressure.¹¹

Secondary polycythemia may occur as a physiologic response to chronic anoxia in chronic pulmonary disease. An increase in the concentration of formed elements in the blood may occur, changing the dynamics of the intracranial venous pressure

and resulting in increased blood viscosity. This has been suggested as the possible mechanism for the production of papilledema by several investigators.^{12,13} However, it is unlikely that polycythemia is a major factor in the production of papilledema, a view supported by Simpson's observation of only one case of secondary polycythemia in eleven patients with pulmonary insufficiency and papilledema.^{9,10} Furthermore, uncomplicated primary polycythemia is rarely associated with papilledema.^{2,14} Nevertheless, the association of papilledema and polycythemia is an important one and should raise the possibility of an intracranial expanding lesion. An example of this is hemangioblastoma of the cerebellum associated with polycythemia.

Another contributing factor in the development of papilledema is congestive heart failure as proposed by Austen because of its occurrence in nearly all of the twenty-five cases reported.² The pulmonary congestion is thought to augment blood-gas abnormalities, resulting in papilledema.

The most likely cause for the production of papilledema seen in pulmonary insufficiency appears to be the presence of hypoxia and hypercapnia, which are known to produce edema and congestion of the brain. This has been suggested by the post-mortem findings of cerebral edema and congestion in thirty-six cases of pulmonary insufficiency reported by Simpson.¹⁰ Our examination of necropsies of cases with chronic pulmonary insufficiency has also confirmed the presence of marked congestion and edema of the brain manifested by flattening of the cerebral convolutions, decrease in the size and sometimes near-collapse of the ventricles, and occasional pressure cone due to herniation of the temporal lobes or cerebellum.

Both an elevated carbon dioxide content and a low oxygen saturation may result in cerebral vasodilatation, increasing the cerebral blood flow, producing elevation of the cerebrospinal fluid pressure¹⁵ and papilledema.^{4,10} The effect of carbon dioxide tension is thought to be greater than that of hypoxia in producing these changes.

One problem which remains unanswered, however, is the development of papilledema in some cases of cardiopulmonary insufficiency and not in others. To add further confusion, a case of papilledema and diffuse emphysema, in the absence of hypercapnia, has been recently reported by Leggat.¹⁶ This stresses the need for further evaluation of this problem which is made difficult because treatment is necessarily directed at all the symptoms simultaneously, not allowing individual investigation of each contributing factor.

The mental changes that accompany chronic pulmonary insufficiency have been attributed to elevated carbon dioxide tension. In this regard certain observations are of importance.

Carbon dioxide retention is known to cause increased intracranial pressure, presumably as a result of cerebral vasodilatation and subsequent increased cerebral blood flow.^{4,9,17} Furthermore, it has been suggested that a fall in arterial pH accompanying hypercapnia may produce a diminished cerebral oxygen consumption by interfering with its utilization.¹⁸

Pulmonary Diseases and Neurologic Disturbances

The explanation of McCann ascribed to the development of cerebral symptoms in chronic pulmonary insufficiency, is that of impaired circulation at the capillary level due to increased alveolar pressure, as well as diminished negativity of the intrapleural pressure.¹⁹ Further impairment of venous return and insufficient cardiac output may occur due to lack of compensatory rise in intra-abdominal pressure.

More confirmatory evidence suggesting the probable role of hypercapnia in the development of alterations of consciousness, has been the observation of mental changes occurring in anoxic patients following the administration of oxygen, especially in high concentration.^{19,5,10} These changes range from somnolence and mild confusion, to coma and even death. Paradoxically, alleviation of these symptoms followed almost immediately upon discontinuation of the oxygen. The explanation for this phenomenon is that during chronic elevation of the carbon dioxide tension, the respiratory center is no longer responsive to changes in blood carbon dioxide levels. Hypoxia then takes over as the major respiratory stimulus through its reflex action on the chemoreceptors of the carotid and aortic-bodies. When oxygen is administered, this effect is abolished resulting in hypoventilation, further carbon dioxide retention, and subsequently coma.^{3,20} Such situations may usually be prevented with the use of mechanical body-respirators producing hyperventilation and reversing the elevated carbon dioxide content without significantly altering respiration during oxygen therapy. In addition, gradually increased concentrations of oxygen have been recommended.²¹ It is important to recognize the actual cause of impaired consciousness in these cases, especially where oxygen is used, since the sudden progression of symptoms may be wrongly attributed to the disease, rather than to its therapy.

The purpose of the above presentation has been to stress the existence of a few of the more common neurologic features of chronic pulmonary disorders in the hope that early recognition will avoid misdirected therapy and unnecessary surgical procedures.

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Bartone

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