PSEUDOTUMOR CEREBRI (OTITIC HYDROCEPHALUS)*

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Pseudotumor cerebri is a syndrome characterized by increased intracranial pressure, although the ventricular system and the composition of the cerebrospinal fluid are normal. The symptoms are similar to those of a tumor, but the disease cannot be diagnosed by ventriculography, electroencephalography, arteriography or surgical exploration.

Although not mentioned very often in the literature, it is not a rare condition, and is often seen in mild and subclinical forms.[1] Because the etiology and pathogenesis of this syndrome are not well known, it has been given a variety of names which do not describe it accurately. Pseudotumor cerebri, otitic hydrocephalus, toxic hydrocephalus, hypertensive meningeal hydrops, serous meningitis, and benign intracranial tension are all widely used.

The first report of pseudotumor cerebri was made by Quincke in 1896 under the name of serous meningitis.[2,3] The term pseudotumor cerebri was first used by Nonne. In 1931 Symonds reported three cases of otitic hydrocephalus. In 1936 Davidof reported several more. In 1937 McAlpine published reports concerning five cases under the name of toxic hydrocephalus and in the same year Dandy reported 22 cases of increased pressure within the skull without tumor. In 1939 Sahs reported five cases, and in 1947 Yaskin presented reports on 12 patients. Two hundred cases have been reported in recent years, including four to five annually from the Mayo Clinic.[3] We are presenting here a case diagnosed as otitic hydrocephalus

Case Report

A seven year old girl was brought to the hospita with complaints of pain in her right ear. Six days prior to admission, she had been treated with antibiotics by the first doctor attending her. A myringotomy was performed on her right ear, which gave temporary relief. Two days later she began to complain again of pain in her right ear and there was a profuse aural discharge. She also complained of double vision and dizziness. She was again admitted to the hospital. This patient had had otorrhea from her right ear, without pain, the previous year and two years before.

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Physical examination: Eyes: Pupil reflexes were normal, and there was a 3 plus diopter papilledema on both eyes.

Ears: There was a profuse, yellowish - white discharge from the right ear but no mastoid tenderness. The left ear was normal.

Neurologic examination: Tendon reflexes were normal, there was no parasthesia or nuchal rigidity, and Babinski was negative. The right lateral gaze was prominent. A sixth nerve palsy was more pronounced on right vertical gaze. Cerebrospinal fluid pressure was 250 mm H_2O. An electroencephalogram was normal.

Bacteriologic examination: B hemolytic streptococcus, cultured from fluid taken from the ear, was sensitive to all antibiotics. Hemoculture was negative, WBC was 9, 400, and liquid tests gave normal results.

Radiologic examination: There was no definitive pathologic change.

Treatment consisted of 100,000 U procaine penicillin and 1 Gm Chloromycetin administered daily. On the third day after admission, a mastoidectomy was performed, granulation in the mastoid cells removed, and the lateral sinus explored. No blood could be obtained with a needle placed in the lateral sinus. An incision was made in the wall of the sinus and lack of blood in the lumen indicated the presence of a thrombosis in the upper section. Drainage of the ear ceased after ten days but paralysis of the sixth cranial nerve continued and pain increased.

Etiology and Pathogenesis

The etiology of this disease is unknown, although many possibilities have been suggested. Predisposing factors such as infections, head injury, hormonal disturbances, and allergy may play a part in the etiology. Smith, [3] at Johns Hopkins Hospital, suggested three classifications according to the pre-existing symptoms:

1. Otitic group: Otitis media, upper respiratory tract infections, or post-trauma.
2. Metabolic obesity group: Cases seen in fat young women having menstrual troubles.
3. Nonotitic or idiopathic group.

Our case falls within the first group, discussed below.

Although the exact mechanism of the increased cerebrospinal fluid pressure is not clearly known, it is assumed to be caused by increased production or decreased reabsorption of cerebrospinal fluid secondary to a previous mild and nonpurulent or localized meningeal inflammation. Many authors suggest that dural sinus thrombosis is the main etiological factor in otitic

hydrocephalus. Symonds considers the disease may be caused by lateral sinus thrombosis in conjunction with otitis media on the same side. He thinks that in patients with this condition the opposite lateral sinus may also be congenitally defective.[5] Belloni[6] believes that otitic hydrocephalus is a focal disease caused by an aseptic inflammatory process of the brain and choriomeninges through an allergic mechanism. He suggests that purulent otitis media may cause septic intracranial complications such as thrombophlebitis, purulent meningitis, cerebral abcess, and phlegmons, by diffusion of germs, and it may also cause some aseptic complications of a focal allergic nature such as otitic hydrocephalus.

A thrombosis that irritates the vascular endothelium might provoke hydrocephalus, whereas a thrombosis that obstructs the passage itself causes cerebromeningeal edema. For example, edema of the leg in cases of femoral vena phlebitis is caused by a reflex phenomenon of the vasomotor system rather than a mechanical obstruction in the femoral vein. The edema disappears following resection of the thrombosed section of the vein. A sinus thrombosis could activate this reflex phenomenon, resulting in hydrocephalus.[6]

Besides otitic cases, Dees and McKay[7] in 1959 reported three cases in which the patients acquired pseudotumor cerebri after treatment with adrenal steroids for asthma. Gass[3] reported two cases contracted following attacks of poliomyelitis.

Dunn, Baker and Wagener,[8] because some patients responded satisfactorily to a diet without salt and water, connect the disease to an edema caused by a hormonal syndrome.

In some cases, spinal fluid is found along with small cystic masses within the arachnoid membrane in the subarachnoid spaces.[2]

Jefferson noted that this syndrome may accompany Addison's disease. Yaskin, Groff and Shenkin point out that pseudotumor cerebri may develop as a result of diffused inflammation of the central nervous system.[5]

Dandy describes this syndrome as increased skull pressure caused by secondary vasomotor disturbances related to changes in blood volume of the brain.

Millichap[4] has analyzed 224 cases which he found in the literature. He reported that 84 (37 per cent) were children and 75 (90 per cent) of these were in the 5 to 15 age group. According to this author, the syndrome appeared following an otitis media in 65 of the patients, and following mastoiditis, head trauma or other than otitis media in 66 of the patients. Ninety-three patients had no previous history of infection or trauma. Those who

contract the disease after otitis media are mostly children (66 per cent). The frequency of the syndrome in children is quite high and most of the cases among children are otitic.

Symptoms and Clinical Observations

The symptoms are the result of increased intracranial pressure. The most frequently reported complaint is headache, which is usually more mild than in the case of brain tumor.[5] Nausea and vomiting are also characteristic. Blurred vision or temporary loss of vision are common.[9] Blindness may occur much sooner than in brain tumors because of the sudden increase of pressure in the skull. Dizziness may also be reported. A primary symptom is diplopia caused by paralysis of the abducens nerve. There is usually no fever and the patient feels good and mentally alert. Papilledema is usually found on both sides, and in rare instances, there is also a paralysis of the fifth and seventh cranial nerves.[2] Foley reported internal rectus paralysis in addition to abducens paralysis in one case, and Smith reported a superior rectus paralysis in one case and inferior rectus paralysis in another.[3] However, generally no focal neurologic abnormalities can be found other than abducens paralysis. Ataxia and convulsions have been reported, and Maisel[10] reported retinal hemorrhage in three cases.[3-7]

Generally, cerebrospinal fluid pressure increases, but there are no chemical or cytologic changes in the structure of the fluid itself. Cerebrospinal pressure can be as high as 600 mm H_2O, but usually it is around 100 mm H_2O.

No asymmetry or shifting is seen in the ventricles, which are often smaller than normal.

Diagnosis

Otitic hydrocephalus should definitely be considered in cases where the patient develops headache, 5 to 6 plus diopter papilledema, with or without sixth nerve paralysis, following otitis media. Lumbar puncture is a useful diagnostic tool, as high cerebrospinal fluid pressures are characteristic of the disease. Electroencephalography, pneumoencephalography, arteriography, and other examinations give normal results. Although roentgen ray examination of the skull reveals nothing abnormal, roentgenograms of the mastoid may show some haziness and fulness. A patient with persistent otitis media should be examined with an ophthalmoscope. However, establishment of the diagnosis of pseudotumor cerebri with its benign prognosis should not signal the end of diagnostic studies.

Differential Diagnosis

Tumoral lesions, cerebral abcesses or hematomas within the skull should also be considered and eliminated as possible causes of the symptoms. Early formation of papilledema is an important factor in a differential diagnosis.

Prognosis

The prospects for complete recovery are good. About 90 per cent of the patients whose cases are reported recovered completely, in the course of time.[10] Loss of vision is the most important complication, but permanent loss of vision occurs in only five per cent of the cases. Complete recovery may take weeks, even years.

Treatment

Treatment is symptomatic. The aim is to prevent optical atrophy and to keep the cerebrospinal fluid pressure at a low level so that the nerve will not be permanently damaged. Lumbar punctures performed as needed will keep the spinal pressure within normal limits (200 mm H_2O). The pressure should not be decreased by more than half at any one time. The usefulness of a lumbar puncture varies with the stage of the disease, the virulence of the pathologic organisms, and the personal tolerance of the patient. This procedure is therefore not always successful in bringing the disease under control. If the patient's spinal pressure remains at 300 mm H_2O following three punctures, subtemporal decompression is necessary. The mastoid should be cleared of any infection, if present. If a thrombosis is suspected, anticoagulators (Heparin, Dicumerol) should be administered and the thrombosis removed if possible by mastoidectomy. Antibiotics, although they may conceal the disease in some cases, should be given because they usually slow down the progress of the disease. Some doctors recommend the use of anti-allergic drugs, thinking that the disease may be of allergic origin.

Some doctors are against surgical intervention , believing that the condition can be cured or controlled successfully with drugs such as ammonium chloride, acetozolamide (Diamex) and a low water and salt diet to encourage dehydration.

Summary

Pseudotumor cerebri (otitic hydrocephalus) is decribed and the author presents the case of a seven year old girl with the disease. Although considered relatively rare, reports reveal that it may be more common than ori-

ginally believed. It is characterized by increased intracranial pressure, although the ventricular system and the cerebrospinal fluid composition remain normal. The etiology and pathogenesis of the condition are not generally known. Treatment is symptomatic and the prognosis is good in the majority of cases.

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