CHOKED DISCS ASSOCIATED WITH OTITIS MEDIA

An Explanation of the Mechanism in Certain Cases

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Otologists are occasionally confronted with the problem in which the patient, making an apparently uncomplicated convalescence from an attack of otitis media, is found to have a choking of the optic discs. Because other symptoms of increased intracranial pressure are lacking, the condition may be called an optic neuritis. Spinal puncture in these cases, however, will usually disclose a high degree of intracranial pressure with no increase in the cell count of the spinal fluid.

In 1896, Quincke[1] described a syndrome in which the signs of increased intracranial pressure were associated with the findings of clear cerebrospinal fluid on lumbar puncture. He called attention to its occurrence chiefly in young women and its association with ear infections, head injuries, and other factors. He believed that, in these cases, there was an excess of cerebrospinal fluid as a result of hypersecretion of the choroid plexus. He referred to the condition as serous meningitis.

Numerous terms have since been used to describe and label these cases. In the group in which an otitic infection apparently initiated the syndrome, different authors have supplied the name cerebral pseudo-abscess, meningismus, serous meningitis, serous ependymitis, arachnoiditis, hypertensive meningeal hydrops, toxic hydrocephalus, and otitic hydrocephalus.

During the past eight years, 23 patients have come to the Clinic presenting a high degree of intracranial pressure, the cause of which could not be adequately explained. Of these 23 cases an otitic infection initiated the syndrome in 10 instances. This high incidence of otitic infection naturally suggests that these cases constitute a disease entity with a common etiological factor related to the otitis.

In 1931, Symonds[2] published an article in which he applied the term otitic hydrocephalus to the syndrome of increased intracranial pressure following an ear infection. He believed that the condition was due to an increased amount of cerebrospinal fluid. In a more recent article[3], he concludes that in this syndrome infection of the lateral sinus is the most important etiological factor, but that *obstruction* of the sinus is not an essential factor. He suggests that mural thrombophlebitis of the superior longitudinal sinus, by retrograde extension from the lateral sinus, may, by putting out of action a sufficiently large proportion of the arachnoid villi, so interfere with cerebrospinal fluid absorption as to cause otitic hydrocephalus.

A study of the ten cases we have observed also indicates that the

syndrome is due to sinus thrombosis. However, encephalograms which were made in ten cases will not permit of a diagnosis of hydrocephalus. It would appear rather that the increased intracranial pressure is due to venous congestion occasioned by interference with the venous outflow from the cranial cavity. The case which suggested sinus thrombosis as the cause of the increased intracranial pressure in this group was observed in 1934.

Case 1: This patient was a girl of nine years whose history stated that two years previously a discharging left ear had suddenly developed without previous pain. The discharge ceased after a few days and there appeared to be no sequelae. However, about one month later, while on a visit to another city, it was found that the vision was seriously impaired due to a marked choking of the optic discs. A right subtemporal decompression was performed, the choking of the optic discs subsided, and the vision returned. The patient had been in good health since that time, but there was a constant bulging at the site of the decompression, although this hernia was not tense.

Three days prior to her entry on January 20, 1934, a second attack of left otitis media developed. At the time of admission her temperature was 105° F. and she appeared quite ill. During the next few days there was a marked diurnal fluctuation in the temperature, and on the seventh day of her illness a left mastoidectomy was performed, disclosing a large amount of pus in the mastoid. A perisinus abscess was present and it was noted that the sinus was white in color instead of the usual deep blue. Culture revealed a type III pneumococcus. The temperature continued to fluctuate and on the twenty-first day of the illness the left internal jugular vein was ligated. The temperature subsided somewhat after this, but the hernia became quite tense and choking of the optic discs began to appear. The spinal fluid pressure was found to be 400 mm. of water. The fluid contained 110 cells, 60 per cent of which were lymphocytes and 40 per cent polymorphonuclears. After this, a definite purulent meningitis developed from which a type III pneumococcus was cultured and the patient died on the thirty-fourth day.

At postmortem examination the left lateral sinus was found to be represented merely by a fibrous cord, indicating that there had apparently occurred a thrombosis of this sinus with the attack of symptomless otitis two years previously, and that this thrombosis, which had healed without suppuration, had been the cause of the increased intracranial pressure at that time. There was an acute thrombophlebitis of the left superior petrosal sinus and of the sagittal sinus, together with an acute, diffuse fibrinopurulent leptomeningitis.

COMMENT: This, then, is a case of "Otitic Hydrocephalus" with recovery following the first attack and with necropsy findings after the second attack. This led to the belief that the condition which Symonds had described in 1931 under this title might be due to thrombosis of the intracranial sinuses which, by partially obstructing the venous outflow from the cranial cavity, causes an increased intracranial venous pressure, cerebral venous engorgement, and a consequent rise in intracranial pressure. Otologists have long been aware that papilledema occurs in a considerable percentage of cases of sinus thrombosis com-

plicating otitis media but Symonds, in 1937[3], was the first to offer convincing proof that the syndrome under discussion is due to sinus thrombosis.

Sinus thrombosis may be asymptomatic and may occur in the course of a very mild otitis media—a fact which is not universally recognized. Also, if secondary infection does not occur the thrombus will heal without suppuration and usually with recanalization. As recently stated by Sutherland[4], "A thrombus is essentially a protective measure and is not, as the minority believe, infected from the beginning. It is nature's method of blocking the blood channel in an effort to prevent further spreading of an infection. It is the secondary infection of the clot and not the thrombus that constitutes the danger."

In addition to this case, nine other cases have been observed. In seven of the ten cases the ages were between seven and twelve years. Sinus thrombosis was proved at autopsy or at mastoidectomy in five cases and its presence was indicated by the Toby-Ayer test in three additional cases. In one case a perisinus abscess was found but the lateral sinus bled when incised. In the remaining case there was no definite proof of sinus thrombosis. Encephalograms in six cases showed no evidence of an expanding lesion or of hydrocephalus. The total volume of the cerebrospinal fluid was increased in only one instance. All patients were treated by repeated spinal punctures with the exception of one case in which the pressure was only 200 mm. of water and the process was subsiding spontaneously. In four cases subtemporal decompression was performed. The intracranial pressure returned to normal in from one to eleven months in seven cases. In one case the pressure is still present three and one-half months after the onset. Two cases which came to necropsy showed extensive thrombosis of the dural sinuses, including the longitudinal sinus. In seven instances the disease process was on the right side and in three on the left side.

In Symonds' experience also this syndrome was found more frequently after a right otitis media. The explanation of this is found in the anatomy of the dural sinuses. When one of the lateral sinuses receives most or all of the blood from the superior longitudinal sinus, it is more liable to be the right than the left. Therefore, a thrombus of the right lateral sinus is more apt to seriously impair venous return from the cranial cavity and is also more apt to extend into the longitudinal sinus than is a thrombus in the left lateral sinus. On purely anatomical grounds, Symonds estimated that the chances of a right as opposed to a left lateral sinus thrombosis extending into the superior longitudinal sinus are as about 6 to 4 or 7 to 3. Of 26 cases of this disease, including his own, the right ear was affected in 17 and the left ear in 9. Adding

my cases there are now 24 cases following right otitis media and 12 after left otitis media.

Discussion

I should like to call attention to some false interpretations of certain observations which have been made in these cases. First of all, the fact that one can obtain a large amount of cerebrospinal fluid at lumbar puncture before the pressure is reduced to normal does not mean that an increased amount of cerebrospinal fluid is present. Due to the obstruction of the venous outflow in these cases, as spinal fluid is removed, it is immediately replaced by an increased amount of venous blood within the already engorged cerebral veins. This cerebral venous engorgement forces a larger quantity of cerebrospinal fluid out of the lumbar puncture needle than one would be able to obtain if the venous obstruction were not present. Everyone is familiar with the fact that a larger quantity of fluid can be obtained at spinal puncture if one applies jugular compression to increase the pressure and the rate of flow. A similar factor is operative in these cases. In other words, the effects of sinus thrombosis may be compared to the effects of continued jugular compression.

Another frequent observation in this type of case is that when a trephine is performed, the arachnoid membrane is found to be distended with fluid. This observation has led the surgeon to assume that an external hydrocephalus is present. This, however, is not the case, at least not in the majority of instances. Since the encephalographic films in these cases give essentially negative findings, it seems fair to assume that the distention of the subarachnoid space as disclosed at trephine is due to the local accumulation of fluid in the subarachnoid space at the site of operation due to the release of pressure at this area. In other words, in these cases there is a normal amount of cerebrospinal fluid which circulates freely so that when the increased pressure is released at a localized area, as in a decompression, the fluid rapidly flows to that point, causing a local distention of the subarachnoid spaces.

Still a third observation in these cases which has led to the belief that hydrocephalus is present is the fact that occasionally when a cannula is introduced into the lateral ventricle, there occurs a copious flow of fluid. This has been interpreted as meaning that the ventricles are dilated. That this assumption is incorrect is shown by the fact that in these cases the ventricles are normal in size or even smaller than normal, as demonstrated by encephalography. When one can obtain a large quantity of fluid from the ventricle in a case of this sort it simply means that, due to the absence of obstruction in the cerebrospinal fluid system, the expanding intracranial veins squeeze all the fluid toward the

point of exit. Therefore, toward the end of the drainage the fluid that is escaping from the cannula is really subarachnoid fluid which has been forced into the ventricle as a result of the drainage. In these cases, one cannot estimate the size of the ventricles from the amount of fluid obtained by ventricular puncture.

In conclusion, therefore, I am in entire agreement with Symonds that the syndrome of increased intracranial pressure without localizing signs occurring after an attack of otitis media is usually due to intracranial sinus thrombosis; that when this picture of increased intracranial pressure is associated with localizing cerebral signs and recovery occurs *without suppuration, it means that the thrombus (probably sterile)* within the intracranial sinuses has extended into the cerebral veins. I am not convinced, however, that the thrombus must necessarily extend into the sagittal sinus in order to produce increased intracranial pressure. If, as is not infrequently the case, one of the lateral sinuses constitutes the chief emissary for the intracranial circulation, then I believe that it is possible for a thrombosis of this sinus alone to produce the picture of increased intracranial pressure. Since the total volume of the intracranial fluid is usually not increased and since the encephalograms in these cases gave essentially normal findings, there obviously is no hydrocephalus. I am inclined to attribute the increased intracranial pressure simply to an engorgement of the intracranial veins plus, in some cases at least, the occurrence of a sterile subdural effusion as the result of external pachymeningitis. Recovery from this state of increased intracranial pressure occurs when, by means of recanalization or the development of collateral supply, the venous return from the cranial cavity is once more adequate. The chief value of a subtemporal decompression in these cases is not that it causes a satisfactory reduction of the intracranial pressure but that the degree of tension in the hernia serves as a guide as to when a therapeutic lumbar puncture should be done. An adequate name for this syndrome is "sinus thrombosis causing intracranial pressure."

References

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