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Pseudotumor syndrome in treated arachnoid cysts

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Abstract. We report three patients with arachnoid cysts treated by cyst-peritoneal shunting in whom intracranial hypertension occurred during episodes of shunt malfunction. In one case this was associated with re-expansion of the arachnoid cyst, whilst in the other two cases this did not occur. The similarities between these two cases and patients with pseudotumor cerebri suggest a common pathogenic mechanism – specifically, a disturbance of the cerebrospinal fluid circulation.

Key words: Arachnoid cyst – Pseudotumor cerebri – Slit ventricle syndrome

Although the precise causative mechanisms of intracranial arachnoid cysts awaits definition, they are clearly intracranial accumulations of cerebrospinal fluid-like fluid which may be associated with intracranial hypertension. In recent years there has been an increasing use of shunting (cyst-peritoneal) to treat these lesions, at the expense of older methods such as simple drainage or marsupialisation with or without the creation of a communication between the cyst and the subarachnoid space [4, 8–12, 15].

The report details three patients with intracranial arachnoid cysts treated by cyst-peritoneal shunting who presented with intracranial hypertension associated with shunt malfunction. In two cases this was not accompanied by reoccurrence of the cyst itself, whereas in the third case it was. This, it is thought, has relevance both to the mechanism of formation of arachnoid cysts and to the broader question of intracranial hypertension associated with expansion of intracranial fluid volume without visible enlargement of the normal fluid-containing spaces, i.e. pseudotumor cerebri and the slit ventricle syndrome.

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Case histories

Case 1

S.J., a 12-year-old boy, first presented aged 3 years with headache, vomiting, mild papilloedema and fullness of the left temporal region. A computed tomographic (CT) scan revealed a large left temporal arachnoid cyst causing midline displacement. Although this was initially drained via a left temporal burr hole (with >60 ml fluid released under pressure), subsequent fluid collection and wound breakdown necessitated insertion of a cyst-peritoneal shunt. A postoperative scan revealed resolution of the midline shift with reduction in cyst size.

The boy remained well until he was 11 years of age, when he presented with fever and tenderness in the left iliac fossa at the site of the previous shunt insertion. On the basis of collapsed cyst demonstrable on CT scan and the presumptive diagnosis of shunt infection, the shunt was removed. Over the following 5 days the child developed severe headache, a left sixth nerve palsy, left pupillary dilatation, bilateral papilloedema, bradycardia and hypertension. Intracranial pressure, monitored via a subarachnoid bolt in the right frontal region, revealed pressures in excess of 50 mmHg. Repeat CT scan showed only a small rim of residual cyst cavity and no hydrocephalus. A drain was inserted into the cyst space and fluid was drained over 24 h yielding an amount equivalent to the daily cerebrospinal fluid production. The cyst-peritoneal shunt was re-established but failed to function adequately. Due to persistent intracranial hypertension the cyst catheter was resited, after which the boy improved clinically. The monitoring bolt was removed. There was no evidence of infection in cultures. He was left with a residual left sixth nerve paresis.

Three months later he presented again with a 1-month history of intermittent bifrontal headaches associated with drowsiness, vomiting and photophobia. He had a left sixth nerve palsy and a left Babinski response. There was no papilloedema. CT and magnetic resonance imaging (MRI) showed small ventricles without any appreciable cyst cavity. A radionuclide patency study revealed a proximal shunt blockage [6]. Intracranial pressure monitored via a Camino system ranged from −5 to 15 mmHg (baseline) with pressure waves >50 mmHg associated with severe headache. On shunt revision sluggish proximal flow was noted with good distal flow and an opening pressure of 5 cm cerebrospinal fluid. The cyst catheter was replaced and improved flow established. The boy's postoperative course was uneventful and he has remained well.

Case 2

K.T. is a 12-year-old boy whose history dates back to infancy when he exhibited poor sleep patterns, behaviour difficulties and night

terrors. At 3 years of age he complained of frontal headaches which woke him in the morning. His neurological examination was normal; in particular, there were no signs of intracranial hypertension. A CT scan showed a right temporal arachnoid cyst causing midline shift with compression of the right lateral ventricle (Fig. 1). There was early separation of the cranial sutures on plain skull X-ray. Intracranial pressure monitored via a catheter into the cyst recorded pressure waves up to 50 mmHg. A cyst-peritoneal shunt (Hakim paediatric shunt, opening pressure 30–50 mmH_2O) was inserted. The postoperative course was marred by low-pressure headaches which settled spontaneously. Recurrence of these headaches necessitated changing of the valve, initially to a high-pressure (91–130 mmH_2O), later to a medium-pressure valve (56–90 mmH_2O) when the former was not tolerated. Prior to the last shunt revision there was a slight increase in cyst size on CT imaging. At both revisions the shunt was patent.

The patient presented 3 years later with temporal lobe epilepsy and headaches, both of which settled on cabemazepine therapy. At 7 years of age he again presented with a 1-week history of bifrontal headaches, diplopia, drowsiness and two episodes of night terrors. On examination there was bilateral papilloedema and bilateral sixth nerve palsies. A CT scan did not demonstrate any appreciable cyst cavity (Fig. 2). The ventricles were small. A radionuclide isotope shunt study was inconclusive. At shunt revision the proximal end was blocked and was replaced. The child's symptoms resolved but his papilloedema persisted; this settled after 3 months of acetazolamide therapy.

He presented aged 11 with recurrence of his headaches. Again these settled spontaneously but recurred 5 months later. A CT scan was unchanged. At operation the proximal end of the shunt and the valve were found to be blocked and were replaced, the latter with a Hakim paediatric valve (opening pressure 45 mmH_2O). There was minimal fluid in the cyst and it seemed to exert no pressure. A lumbar puncture performed at that time was also normal (pressure 8.5 cm cerebrospinal fluid).

The boy remained well until 2 months later when his headaches recurred. Intracranial pressure monitoring recorded pressure waves in excess of 40 mmHg. A proximal shunt block was suggested on a radionuclide isotope shunt study. An intraoperative constant rate infusion study was abnormal. A CT scan was performed and showed no change from the previous two scans. The proximal end of the shunt was noted to be nonfunctional and was replaced. The child has remained well postoperatively.

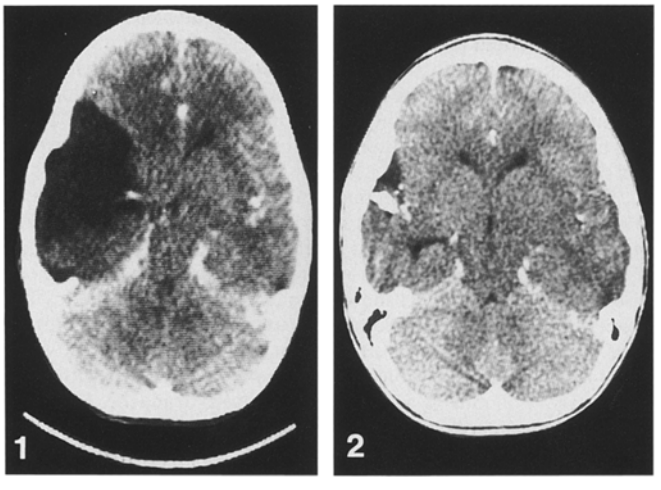

Fig. 1. Preoperative cerebral CT scan of case 2 showing a right temporal arachnoid cyst with compression of the frontal horn of the right ventricle

Fig. 2. Cerebral CT scan of the same patient as in Fig. 1 during an episode of shunt malfunction

Case 3

S.B., a boy now 3 years old, presented at 6 weeks of age with an abnormal rate of head growth, separation of the cranial sutures and a tense anterior fontanelle. Ultrasound and CT scan revealed the presence of a posterior fossa cyst causing compression of the IV ventricle and hydrocephalus. There was associated hypoplasia of the cerebellum. The dome of the cyst was excised and the cyst made to communicate with the subarachnoid space. Persistence of the cyst 3 weeks later led to the insertion of a cyst-pleural shunt (Hakim paediatric shunt, medium-pressure) which was later converted to a cyst-peritoneal shunt after development of a right pleural effusion. Post-operative CT scanning confirmed resolution of the cyst and of the hydrocephalus.

The infant next presented at 5 months of age with fullness of his fontanelle, a rapidly enlarging head and irritability. CT scanning showed recurrence of the cyst with obstructive hydrocephalus. The proximal end of the shunt was found at operation to be blocked and was replaced. A postoperative CT scan showed reduction in the ventricular and cyst size.

He was then well until 13 months of age when he presented with similar symptomatology. Recurrence of the cyst and hydrocephalus was again confirmed radiologically. The proximal end of the shunt was again malfunctioning and was replaced. A similar sequence of events occurred 4 months later. A proximal block was again causative of shunt malfunction with ventricular and cyst enlargement.

Apart from one episode irritability and drowsiness at 2.5 years of age associated with a mononucleocytosis of his cerebrospinal fluid, he has remained well.

Discussion

The clinical cases presented here raise two important issues. The first concerns the origin and potential means of disposal of the fluid contained in arachnoid cysts. The second focuses on the problem of how impairment of intracranial fluid drainage, either cerebrospinal fluid or cyst fluid – if indeed these are different – may cause intracranial hypertension without obvious radiological enlargement of the fluid-containing spaces.

The first detailed description of intracranial arachnoid cysts was probably that of Bright [2] in 1831, who proposed that these cysts represented loculations of fluid within the arachnoid membrane, which had a variable degree of communication with the subarachnoid space. Subsequently, Starkman et al. [18], developing the concept of Weed [19], proposed that during formation of the subarachnoid space, cerebrospinal fluid, impelled by choroid plexus pulsations, dissected the perimedullary mesh in an aberrant fashion to create pockets of cerebrospinal fluid entrapped within what would become the arachnoid mater. This view is, however, at variance with the more recent proposals by Osaka et al. [13] that the subarachnoid space develops prior to the appearance of the choroid plexus and indeed to the formation of the cerebrospinal fluid. Robinson [16] has proposed a further theory of arachnoid cyst formation, claiming that such fluid accumulations arise as an ex vacuo consequence of cerebral agenesis.

Whatever theory of formation of arachnoid cysts is espoused must be capable of accounting for the observation that these cysts may be under pressure and may progressively enlarge. One possibility, as outlined by

Smith et al. [17], is that fluid continues to enter the cyst from the subarachnoid space but its egress is prevented by a postulated valve effect. Alternatively, Go et al. [5] have proposed that arachnoid cysts may accumulate fluid by active secretion of fluid from the cyst wall, supporting their argument by the claim that there are, within the wall, microvilli, fenestrations and apical ATP-driven Na^+ and K^+ pumps. This would make the cyst a self-contained entity capable of enlargement by secretion of fluid within the cyst itself.

The cases here described clearly provide support for the view that a mechanism must exist for the continued accumulation of fluid within an existing arachnoid cyst. Of particular interest is that the presumed re-accumulation of fluid associated with drainage failures due to shunt malfunction is not necessarily accompanied by the re-establishment of an apparently loculated fluid collection, as was present prior to treatment. Thus, in the present report, a clear difference emerged between the two cases in which there was no re-accumulation of fluid within the cyst and the one case in which there was. In addition, the daily volume of drainage of fluid from the cyst in case 1 was equivalent to the daily cerebrospinal fluid production. We would suggest that the cyst fluid originates from cerebrospinal fluid in the subarachnoid space and then passes into the cyst cavity. In some circumstances, possibly related to cyst collapse with formation of adhesions after shunting, the cyst cannot re-expand (cases 1 and 2), whereas in other circumstances it may do so. If re-expansion is not possible, the cyst cannot again serve as a volume reservoir, the fluid must then be accommodated elsewhere, the suggestion here being that the same mechanism is operating as has been proposed for the pseudotumor syndrome [3, 7]. This concept raises the possibility that there may be an underlying disturbance of the cerebrospinal fluid circulation in arachnoid cysts in general and, similarly, that the original cyst formation may indeed be a consequence of such disturbance. This point might be clarified by studies on cerebrospinal fluid dynamics in patients with treated and untreated arachnoid cysts.

Turning to the second and related question of how, precisely, the increase in intracranial pressure occurs, it may be said that there is no obvious mechanism to account for the observed intracranial hypertension other than fluid excess. There is, indeed, absolutely no basis on which to inculpate the other two intracranial compartments, i.e. an increase in brain volume or an increase in blood volume. Thus there would appear to be a prima facie case for the first assumption: an increase in intracranial fluid volume, presumably from the same source as the fluid in the original cyst. On the issue of where this fluid is accommodated in patients in whom the cyst does not re-establish, it may be relevant, as alluded to above, to draw parallels with two other identifiable situations in which intracranial hypertension secondary to intracranial fluid accumulation may occur without visible enlargement of the fluid compartment, to wit, the pseudotumor syndrome and the so-called slit ventricle syndrome in treated hydrocephalus [3, 7]. Although both these conditions remain somewhat contentious in regard to mechanism, the argument has been advanced elsewhere, developing the concepts of Fishman [3] and Page [14] in particular, that the excess fluid consequent upon impaired cerebrospinal fluid absorption primarily or secondary to shunt malfunction passes from the cerebrospinal fluid compartment reversibly into the brain parenchyma. On the basis of the observations here reported and the patient described by Aoki et al. [1], it is proposed that a similar mechanism may be operational in these cases of arachnoid cysts.

In summary, three cases have been described in whom intracranial hypertension occurred following shunt obstruction in patients with previously shunted arachnoid cysts. In one patient this was associated with re-establishment of the cyst, while in the other two cases there was no such localised fluid re-accumulation. It is argued that in all cases the cause of the intracranial hypertension seems to be an increase in intracranial fluid volume, but that for reasons yet to be determined the fate of the fluid excess is different. Thus, the fluid may either re-accumulate within the previously established cyst or it may pass reversibly into brain parenchyma in a manner similar to that which may occur in pseudotumor cerebri and the slit ventricle syndrome. If this supposition is correct, such cases may, along with pseudotumor cerebri and its variants and the slit ventricle syndrome, be subsumed under the rubric of the "pseudotumor syndrome" (i.e. intracranial hypertension due to disorders of intracranial fluid circulation without enlargement of the fluid spaces). The challenge remains to identify and classify the precise mechanisms involved. In meeting the challenge it may be that important aspects of intracranial fluid dynamics, hitherto unrecognised, may come to light.

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