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Case Report

Non-obstructive hydrocephalus associated with intracranial schwannomas: hyperproteinorrhachia as an etiopathological factor?

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Summary

Background. This series illustrates the association of communicating hydrocephalus with intracranial non-obstructive schwannomas. This association has commonly been observed, however it has only been reported once previously. Moreover, in all the patients we present, hyperproteinorrhachia was a common denominator. This finding may therefore be the underlying mechanism for hydrocephalus.

Method and findings. Seven patients presenting with intracranial schwannomas along with non-obstructive hydrocephalus and hyperproteinorrhachia are reported. Six had a vestibular schwannoma and presented with a unilateral deafness and various degrees of gait disturbance, urinary incontinence and neuropsychological impairment. Due to their advanced ages, these patients underwent a ventriculo-peritoneal shunt, and their symptoms related to hydrocephalus resolved.

One patient that suffered from hemifacial dysesthesia and memory deficits presented with a non-obstructive trigeminal schwannoma. In this case the tumour was removed and the hydrocephalus was consequently reversed, and the CSF protein content normalized.

Interpretation. The constant finding of hyperproteinorrhachia in all these patients suggests that a high CSF protein content may be the underlying cause of hydrocephalus through a speculative mechanism of decreased CSF resorption.

Keywords: CSF protein; hydrocephalus; trigeminal schwannoma; vestibular schwannoma.

Introduction

Intracranial schwannomas can be associated with obstructive hydrocephalus by compression of the 4th ventricle or the aqueduct of sylvius [1, 3, 4, 28]. In rarer instances non-obstructive hydrocephalus can also be observed with intracranial schwannomas.

In the past 20 years we have followed 6 patients with normal pressure hydrocephalus associated with small, non-obstructive vestibular schwannomas and one patient presenting with high-pressure hydrocephalus associated with a non-obstructive trigeminal schwannoma. Each of these patients had an elevated CSF protein level. This series of patients suggests that a raised CSF protein content can be the underlying pathophysiological mechanism of communicating hydrocephalus. This phenomenon has already been described in association with spinal cord tumours [8, 10, 20, 21], but, to our knowledge, it has only been reported once in 1977 by Kühne *et al*. [16] in association with intracranial tumours.

Patients and method

Patient data

There were two males and five females in whom age ranged from 23 to 79 years (median: 69 years). Duration of symptoms and signs prior to diagnosis varied from 2 months to 3 years (median: 10 months). The 6 oldest patients, with a vestibular schwannoma suffered from gait disturbances; 2 patients complained of unsteadiness, 3 had frequent falls and one was wheelchair-bound due to the severity of his gait difficulties. Five of the 7 patients had urinary complaints, four of them were incontinent. Two patients did not have any alteration in their cognitive functions, whereas 5 had mnesic alterations and concentration problems with some episodes of confusion. All patients with an acoustic schwannoma complained of unilateral deafness. The patient with the trigeminal schwannoma presented hemi-face dysesthesia (Table 1). Radiological investigations consisted of CT and MRI. These revealed the schwannomas and the hydrocephalus (Figs. 1–4), confirmed to be non-obstructive.

Table 1.

Case	Sex	Age	Duration	Presentation	CSF Pressure (cm H2O)	CSF proteins mg/100 ml	Side	Diameter	X-Ray	Operation
1 V	M	75	4 m	–unsteadiness, falls –incontinence –confusion –Lt. hypo-acousia	ventricular 13	ventricular 210	Lt.	1.5 cm	CT	VP shunt
2 V	F	77	3 m	–unsteadiness –incontinence –confusion –Lt. hypo-acousia	ventricular 11	ventricular 45	Lt.	1.5 cm		VP shunt
3 V	F	77	18 m	–unsteadiness, falls –urinary urgencies –Rt. hypo-acousia	ventricular 9	ventricular 39	Rt.	1.5 cm	CT MRI	VP shunt
4 V	M	77	4 m	–unsteadiness, falls –incontinence –memory deficits	ventricular 13	ventricular 172	Rt.	2 cm	CT MRI Fig 1 Fig 2	VP shunt
5 V	F	71	2 y	–unsteadiness –Lt. hypo-acousia	ventricular 12	ventricular 43	Lt.	2.3 cm	CT MRI	VP shunt
6 V	M	79	2 m	–unsteadiness, wheelchair –incontinence –confusion –Rt. hypo-acousia	ventricular 12	ventricular 110	Rt.	2.8 cm	CT MRI	VP shunt
7 T	F	23	9 m	–headache –memory deficits –Rt. central scotoma –Rt. face dysesthesia	lumbar 30	lumbar preop. 58 lumbar postop 37	Rt.	4 cm	CT MRI Fig 3	tumour resection

V Vestibule schwannoma, *T* Trigeminal schwannoma.

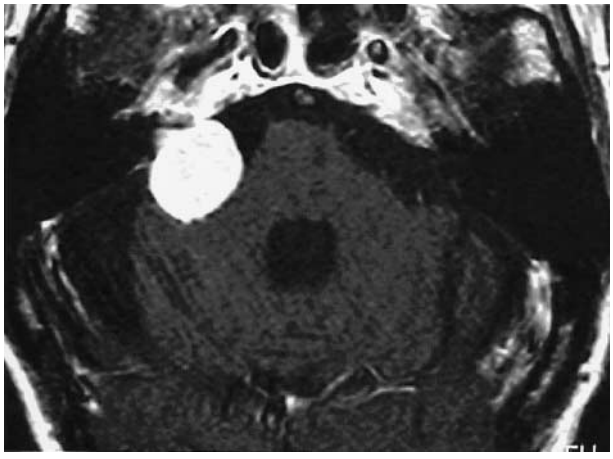

Fig. 1. Case 4. MRI, (T1 gadolinium) axial view. The fourth ventricle is not obstructed in the presence of a right cerebellopontine angle lesion which has the radiological characteristics of a vestibular schwannoma

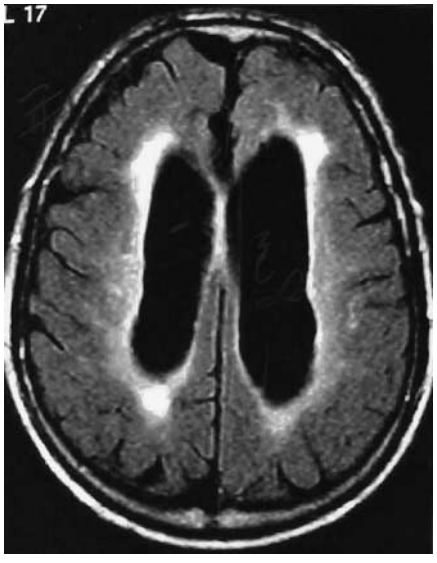

Fig. 2. Case 4. MRI, (T2 fluid suppressed sequences) axial view. Dilatation of the lateral ventricles with transependymal periventricular CSF resorption

Surgery

All patients with a vestibular schwannoma underwent a ventriculo-peritoneal CSF shunt to control the hydrocephalus. Low pressure Pudenz or Pudenz-Schulte pump device and low pressure peritoneal catheter were used. The lateral ventricle was tapped through a right parietal burr hole and the peritoneal catheter was introduced in the right lower quadrant of the abdomen. Ventricular opening pressure was recorded at the time of shunt placement and found to be normal, ranging from 9 to 13 cm H2O, the patient being under general anaesthesia with positive pressure ventilation. One patient presented with a subdural haematoma following the shunt insertion, requiring craniotomy for drainage. One patient required

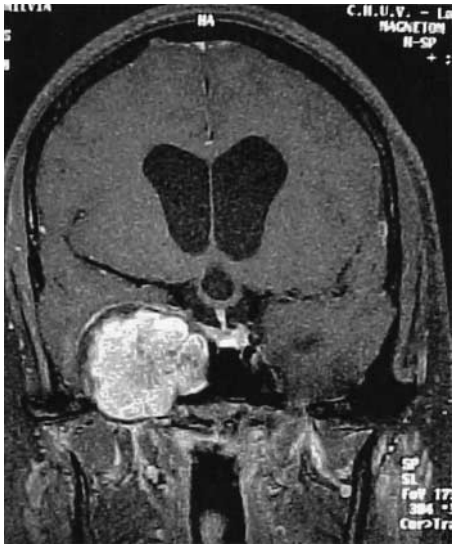

Fig. 3. Case 7. MRI, (T1 gadolinium) coronal view. Right non-obstructive trigeminal schwannoma with lateral and third ventricular dilatation

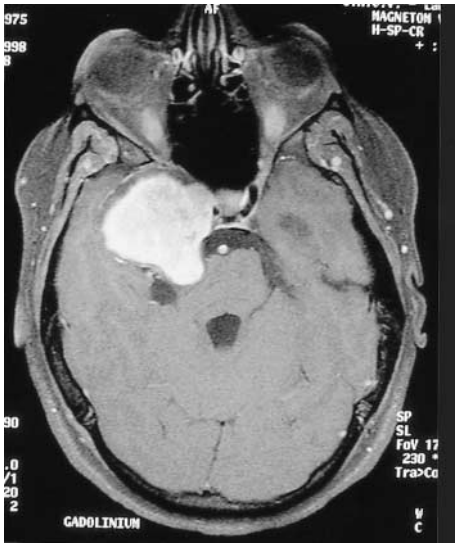

Fig. 4. Case 7. MRI, (T1 gadolinium) axial view. The trigeminal schwannoma is space occupying, however, the prepontine cisterns are still patient and the 4th ventricle is widely open

shunt revision for dysfunction. The patient with the trigeminal schwannoma underwent a right temporo-pterional craniotomy for total tumour removal. All patients were followed up clinically and radiologically.

CSF characteristics

In all the patients with vestibular schwannomas, ventricular CSF was obtained at time of shunt placement. Protein level ranged from 39 mg/100 ml to 210 mg/100 ml (median: 103 mg/100 ml). In the youngest patient with the trigeminal schwannoma, a lumbar puncture performed before tumour resection revealed an opening pressure of 30 cm H2O and a protein level of 58 mg/100 ml. In this patient, a lumbar puncture was repeated one month postoperatively and the CSF protein concentration had decreased to 37 mg/100 ml.

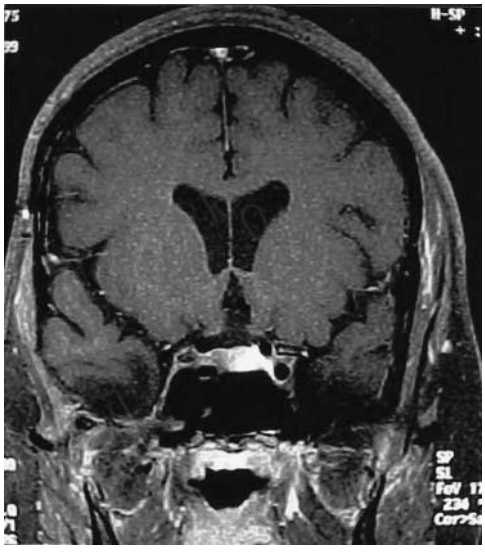

Fig. 5. Case 7. MRI, (T1 gadolinium) coronal view. Following the right trigeminal schwannoma resection, ventricular size is reduced

Follow-up

All patients with vestibular schwannomas improved in every aspect of their symptomatology within 2 weeks after placement of the VP shunt. One patient died post-operatively of unrelated causes. Following removal of the trigeminal schwannoma, the patient showed an improved neuropsychological examination with normalisation of her memory functions. A control MRI scan (Fig. 4) showed a reduced ventricular size.

Discussion

Seven patients presenting with non-obstructing hydrocephalus and hyperproteinorrhachia are reported. This group represents 3% of the collection of patients we follow-up for cranial nerve schwannoma.

Six patients in this series had a small-sized vestibular schwannoma and presented with deafness and a clinical picture of NPH (cognitive impairment, gait disturbances and urinary problems). The last patient, who complained essentially of hemi-face dysesthesia and memory deficits, had a non-obstructive trigeminal schwannoma, associated with a high CSF pressure.

Hydrocephalus associated with an intracranial schwannoma may be treated according to different approaches. In most instances, the hydrocephalus results from a large tumour obstructing the CSF flow by compression of the fourth ventricle [1, 3, 4, 28]. In such

instances, the treatment alternatives are to first proceed to a CSF shunt and later to the resection of the tumour, or to excise the tumour, which in most instances will be followed by a resolution of the hydrocephalus. In cases of small, non-obstructive cranial nerve schwannomas, the aetiology of the associated hydrocephalus has to be established. In the presence of an elevated CSF protein content and communicating hydrocephalus, one could proceed to the excision of the tumour and hope that CSF protein content will normalize and hydrocephalus resolve. A surgical alternative may consist in performing a CSF shunt procedure to control the clinical manifestations due to hydrocephalus. Considering the small size of the tumours, as well as the lack of specific symptoms secondary to the space occupying effect, and also because of the advanced age of our first 6 patients, we elected not to proceed to surgical resection. Rather, we decided to treat the hydrocephalus with a shunting procedure, and observed the evolution of the tumour clinically and radiologically, as previously advised by others for patients in this age group [19]. Ventricular pressures recorded at surgery were normal in all cases. Ventricular CSF protein was found to be elevated in all cases; markedly so in case 1 and 6, less so in the others.

All the patients with vestibular schwannomas had their CSF specimens obtained from the lateral ventricles at time of surgery. Protein concentration distributes in a gradient fashion in the CSF ventricular and subarachnoid compartments. CSF protein content is the highest in the lumbar subarachnoid space, and progressively lowers at more rostral levels. Normal values are 15 mg/100 ml in the ventricles, 25 mg/100 ml in the cisterna magna, and 45 mg/100 ml in the lumbar subarachnoid region [18]. Factors contributing to this gradient include greater permeability of the blood-brain-barrier to protein in the lumbar region [13], and decreased rate of protein removal in this region as compared to more rostral levels [14]. Thus, owing to the fact that CSF was obtained by ventricular tap, protein concentration values of 45, 39 and 43 mg/100 ml in patients 2, 3 and 5 represent a 2.5 to 3 fold increase as compared with physiological CSF protein content at this level. The last patient was 23 years old, her tumour was larger and symptomatic, we thus decided to remove it. CSF pressure measured pre-operatively during lumbar puncture was higher than normal (30 cm H2O). Lumbar protein content pre-operatively was 58 mg/100 ml, one month post-operatively, it had decreased to 37 mg/100 ml. The 2 follow-up CT scans performed 1 week and 2 months postoperatively showed a reduction in ventricular size (Fig. 4).

To our knowledge, the phenomenon of communicating hydrocephalus secondary to elevated CSF protein content has only been reported once in association with cranial nerve schwannomas [16]. However, this phenomenon was reported for the first time by Nonne and collaborators in association with 1–3% of spinal cord tumours [20]. A large series of 151 spinal cord tumours collected from 1901 to 1990 provided by Celli and co workers [8] even reported that hydrocephalus was associated with 65% of malignant gliomas of the spinal cord. In malignant spinal cord tumours, the pathophysiology of hydrocephalus can easily be understood, since atypical floating cells as well as repeated tumour bleedings are classically observed and may contribute to increase CSF cellularity and viscosity. This hyperviscosity is suspected to interfere with CSF absorption by blocking the Pacchionian granulations. Under these conditions, hydrocephalus does not necessarily resolve following tumour removal. In low grade gliomas of the spinal cord, floating cells in the subarachnoid spaces have rarely been described [9, 17], whereas hyperproteinorrhachia is a common finding that may contribute to communicating hydrocephalus. Hyperproteinorrhachia with hydrocephalus has also been described in association with a few cases of spinal schwannomas [10, 21], and even in about 4% of patients with Guillain-Barré syndrome [12, 23].

The association of hydrocephalus and an elevated CSF protein content is an interesting concept, although a study conducted by Brydon and co-workers [5] indicates that high protein concentrations do not greatly affect the viscosity of CSF. Hyperproteinorrhachia may rather interfere with CSF absorption at the level of the Pacchionian granulations. To understand better why the absorption is decreased at the level of Pacchionian granulations we have to consider the pathophysiology of these structures. Two main mechanisms are proposed to explain the passage of CSF into the venous system: 1) through transendothelial pores and 2) by pinocytosis across the endothelial cells. The first mechanism is passive and concerns small particles, and the second one is an energy-dependent process, more commonly used by the bigger particles. There is probably a saturation of one or both transport system in the presence of hyperproteinorrhachia, but no study has demonstrated it yet

[2]. The most recent hypothesis considering low grade gliomas of the spinal cord based on the observation of Cinafli and co workers, suggests the presence of an abnormal amount of fibrinogen in the CSF [9, 21]. This increased fibrinogen level could result either from a chronic inflammation and an abnormal blood-brain-barrier or from repeated subarachnoid bleeding from tumour vessels. The CSF fibrinogen will be converted into fibrin at the level of the Pacchionian granulations causing an increase in the CSF outflow resistance and secondary communicating hydrocephalus. In our cases the fibrinogen was not measured.

There are certainly many possible factors that participate in the occurrence of non-obstructive hydrocephaly in the presence of cranial nerve schwannomas. However, our series demonstrates that this phenomenon is very individual because it only occurs in a minority of patients (3%).

Even if these individual factors cannot always be identified, some predisposing factors are relevant to underline. For instance, we have observed in our group that aging plays an important role. In fact, the mean age of the patients we present is higher than the average age of the collection of patients followed in our clinics for a cranial nerve schwannomas, (69 years old compared to 55). Therefore, we hypothesize that older patients have less adaptation to CSF changes and are more prone to develop hydrocephalus.

In conclusion, the present study suggests that communicating hydrocephalus may be associated with hyperproteinorrhachia and non-obstructive cranial nerve schwannomas. This is an interesting observation that has to be considered carefully since documented cases of hydrocephalus associated with intraspinal tumours resolved following excision of the tumour. These observations cannot be generalized, because other cases may worsen despite normalization of CSF proteins, and because not all patients with spinal cord tumours or vestibular schwannomas and increased CSF protein develop hydrocephalus. Considering the variety of responses, hyperpoteinorrhachia is probably not the only factor to consider. It is obvious that each of the above theories does not apply in every single case. One has to individualise the situation and consider also, some "constitutional factors" that may predispose some individuals to develop hydrocephalus in association with CNS tumours. For instance, older individuals suffering from periventricular atherosclerotic leuco-encephalomalacia may be more susceptible to become symptomatic of communicating hydrocephalus secondary to hyperproteinorrhachia. These unidentified constitutional factors appear to play an important role.

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Comment

The association of hydrocephalus due to high CSF protein from tumors is commonly known, but not apparently well documented in the literature. This article beautifully confirms and presents it.

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