CASE REPORTS

Papilloedema and hydrocephalus in spinal cord ependymoma

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*Figure 1 Goldmann
perimetry performed with
III/4 and V/4 isoptre before
the shunt procedure. Note the
absolute loss of nasal fields
and the strong depression of
temporal fields, especially in
the right eye.*

The association of spinal cord tumours with papilloedema is well known but occurs uncommonly. In most such cases the spinal lesion is intradural and in more than half of the cases it is an ependymoma.¹⁻³ We present a case of spinal cord ependymoma in which the initial manifestation was a lowering of vision, with the finding of secondary optic atrophy and non-obstructive hydrocephalus, and in which the spinal cord tumour was detected only 1 month after ventriculo-peritoneal shunt.

Case report

A 29-year-old man was admitted to the neuro-surgical department of our hospital because of progressive visual loss and blackouts. Five years earlier he had complained of low back pain, especially on effort, but computed tomography (CT) of the spine was normal and in recent years there had been no pain or any other complaints until the appearance of the ophthalmic symptomatology. On examination visual acuity was 6/30 in the right eye and 6/9 in the left eye and there was a Marcus-Gunn pupil. The visual fields, tested on a Goldmann perimeter with III/4 and V4 isoptres, showed on the right eye an almost absolute nasal field loss, a very strong contraction of the temporal field, and enlargement of the blind spot. In the visual field of the left eye there was also an almost total loss of the nasal field and a peripherally depressed temporal

Figure 2 Computed tomography of brain, showing hydrocephalus.

field with inferior arcuate scotoma (Fig 1). The fundus examination disclosed the beginnings of optic atrophy in both eyes, secondary to severe prolonged papilloedema. CT of the brain showed a non-obstructive hydrocephalus (Fig 2). The neurological examination was within normal limits. On lumbar puncture the opening pressure was 70 mm H₂O, the cerebrospinal fluid was xanthochromic, its protein level was 465 mg/100 ml, and glucose was normal.

Figure 3 Computed tomography of lumbar spine, showing ependymoma.

Figure 4 Goldmann perimetry performed with I/4 and V/4 isoptre after the shunt and after the spinal cord ependymoma excision. Note that this time the patient saw the I/4 isoptre and that visual fields enlarged.

The Millipore examination for cytology was negative. The diagnosis was non-obstructive hydrocephalus; it may present with variations from high to normal cerebrospinal pressure and this normal cerebrospinal pressure was considered to be a momentary variation. A supplementary examination of cerebrospinal fluid puncture was not done and the grossly elevated protein result was overlooked.

A ventriculo-peritoneal shunt was immediately instituted. One month later the patient complained of lower back pain and urinary incontinence and examination revealed a sensory level at D₁₀ and bilateral clonus. CT and myelography demonstrated an intradural block between L₂ to D₁₂ (Fig 3). At operation an intradural and intramedullary mass was found at the conus covering the filum terminalis. The histological examination showed myxopapillary ependymoma.

The postoperative course was uneventful and he received local irradiation. Subsequently, the visual acuity improved to 6/7 in both eyes and there was some enlargement of visual fields (Fig 4). Ophthalmoscopy showed bilateral secondary optic atrophy and a massive depletion of the nerve fibre layer.

Comment

According to the literature¹⁻⁴ more than 50% of the spinal cord tumours producing increased intracranial pressure are ependymomas, mainly located in the thoracic and lumbar region. The common denominator in all the reported cases of papilloedema with spinal cord tumours was the resolution of the papilloedema following surgical removal of the lesion.

In our patient the lack of symptomatology pointing to the presence of a spinal cord tumour led to a delay in its diagnosis, the spinal cord investigation having been made only 1 month after the diagnosis of hydrocephalus and secondary optic atrophy. Retrospectively we can state that the high level of protein in the cerebrospinal fluid should have served as a warning sign to continue the investigation. In fact, the present-

ing sign of the spinal tumour in our case was the isolated visual loss due to secondary optic atrophy. We were able to find only six similar cases in the literature.⁵

Several authors⁶⁻⁹ have offered different theories in the attempt to explain the development of increased intracranial pressure and papilloedema in certain cases of spinal cord tumours. Gardner *et al*⁶ postulated that large protein molecules may mechanically block the arachnoid pores and prevent the absorption of cerebrospinal fluid, thus producing a rise in intracranial pressure. Davson *et al*⁷ demonstrated experimentally that infusion of plasma serum into cerebrospinal fluid slowed its absorption. It is of note, however, that in Guillain-Barré syndrome there have been cases with a very high level of protein in the cerebrospinal fluid without development of increased intracranial pressure and papilloedema, and on the other hand there have been cases in which papilloedema developed although the protein level was only moderately elevated or even normal.^{10,11}

The hyperproteinorrhachia may also play an irritative role, producing an aseptic arachnoiditis, and the extension of the arachnoiditis to the chiasmatic cisterns probably explains the existence of severe papilloedema with haemorrhages and exudates in otherwise asymptomatic patients.¹² On the other hand, the hyperproteinorrhachia and the alteration in the cerebrospinal fluid pathway secondary to it, is the main factor responsible for the development of hydrocephalus associated with spinal tumours. Arseni *et al*⁸ suggested that under certain conditions low spinal cord tumours may release products of protein disintegration, increasing the viscosity of the cerebrospinal fluid, and that this hyperviscosity below and above the tumour upsets the balance between secretion and absorption of cerebrospinal fluid, with a slowing down of its circulation from the cranial region to the spinal spaces.

Describing a case very similar to ours Matzkin and Slamovits⁵ implicated repeated episodes of clinical and subclinical subarachnoid haemorrhage from the tumour as a possible mechanism of the increased intracranial pressure in spinal cord tumours, particularly ependymoma. Martin *et al*⁹ pointed out that the spinal cord acts as an elastic reservoir for cerebrospinal fluid, the flow of the cerebrospinal fluid into and out of the spinal canal being considered of importance in the maintenance of a constant intracranial volume. Spinal cord tumours may in fact reduce the capacity of this 'reservoir' by mechanical blockage and thereby cause papilloedema.^{5,9}

The many explanations offered for the aetiology of the increased intracranial pressure in these cases indicates that the search for an unequivocal answer has yet to be continued. It is stressed that the possibility of a spinal cord tumour should be kept in mind in cases of unexplained newly developed papilloedema.

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Metastatic prostatic adenocarcinoma presenting as complete ophthalmoplegia from pituitary apoplexy

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Pituitary apoplexy is a rare presentation of pituitary disease. A case resulting in sudden bilateral complete ophthalmoplegia due to a prostatic metastasis is described for the first time. The pathogenesis of this condition and relevant literature are discussed.

Case report

An 81-year-old West Indian man presented with a 4 day history of severe headache and sudden visual loss on the left. He reported polydipsia but no nausea or vomiting. His ophthalmic history included advanced open angle glaucoma blinding his right eye, previous left cataract extraction complicated by removal of the implant, and right branch retinal vein occlusion. Thirty two years earlier he had undergone a bilateral orchidectomy for prostatic carcinoma.

On examination he was afebrile, oriented but inattentive and had complete bilateral ophthalmoplegia and complete ptosis. Best visual acuities were light perception right eye and 3/24 left eye. Corneal sensation was absent but there were no other cranial nerve or long tract signs. He was normotensive with a pulse of 54 and there were no other signs of endocrine disease. Endocrine function tests confirmed panhypopituitarism and cranial diabetes insipidus (cDI).

A bone scan revealed widespread metastases and high resolution contrast computed tomography scan of the head demonstrated destruction of the pituitary fossa by a solitary enhancing mass eroding inferiorly into the sphenoid sinus and posteriorly through the dorsum sella to reach the pons. There was both supra and parasellar extension (Fig 1A and B).

A provisional diagnosis of pituitary apoplexy

Fig 1A

Fig 1B

Figure 1 Metastatic deposit demonstrated by contrast enhanced high resolution computed tomography. Horizontal (A), and sagittal sections with vertical reconstruction (B), demonstrating a solitary enhancing deposit filling the fossa and expanding superiorly, laterally, and posteriorly.

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