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Communicating hydrocephalus and papilloedema associated with intraspinal tumours : report of four cases and review of the mechanisms

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Abstract

Communicating hydrocephalus and/or papilloedema associated with intraspinal tumours is rare. Four such patients are presented in this article. In addition to previous theories put forward to explain this condition, we would like to propose alteration of craniospinal compliance distribution as a possible underlying mechanism. Consequently, we suggest not performing shunt placement immediately if the intraspinal tumour can be removed.

Key words : Hydrocephalus ; papilloedema ; cerebrospinal fluid ; intraspinal tumour.

Introduction

Common causes of communicating hydrocephalus are idiopathic, subarachnoid haemorrhages and meningitis. Communicating hydrocephalus may be associated with superior vena cava syndrome, Guillain-Barré syndrome, cryoglobulinemia, antiphospholipid antibody syndrome and metabolic disorders (8, 9, 13, 18) but such cases are rare. Hydrocephalus secondary to intraspinal tumours is a well known but rare condition since about 1% of patients with spinal cord tumours have various degrees of hydrocephalus at initial presentation (2, 21). Even if hydrocephalus is accompanied by signs of increased intracranial pressure, such as headaches or papilloedema, diagnosis may be difficult because the symptoms referable to the spinal lesion may be minimal or initially overlooked.

In this article, the authors report four patients in whom an intraspinal tumour was associated with hydrocephalus, papilloedema or both. The problematic aspects of this condition, with regard to clinical presentation and pathophysiology, are discussed.

Case Reports

CASE 1

A 6-year-old girl was admitted in June 1997 because of headaches and vomiting for 2 weeks. Brain computed tomography (CT) scan revealed a communicating hydrocephalus. Neurological examination revealed pyramidal syndrome without motor deficit in the lower limbs. Spinal magnetic resonance imaging (MRI) showed an intramedullary tumour extending from T7 to T9. Complete resection of the tumour was achieved, and histopathological examination demonstrated a fibrillary astrocytoma. There was no postoperative neurological deficit but headaches were still present. Brain CT scan then confirmed the persistence of hydrocephalus and she underwent ventriculoperitoneal (VP) shunt surgery. Ventricular CSF contained 0.10 g/l of protein and 5 cells/mm^3. In November 1997, she developed epilepsy. MRI of the whole neuraxis showed diffuse leptomeningeal enhancement without any spinal cord tumour recurrence. Lumbar CSF analysis showed a raised protein concentration (7.5 g/l) and 22 non-specific lymphocytes/mm^3. In July 1998, MRI showed striking widespread leptomeningeal enhancement and spinal tumour recurrence was suspected. Cytological examination of the CSF did not reveal any abnormal cells but chemotherapy was initiated given disease progression. In December 2000, brain MRI disclosed 3 supratentorial cortical lesions. Biopsy of a left temporal lesion demonstrated an anaplastic oligoastrocytoma (WHO grade III). Despite various chemotherapy protocols, she died in September 2001.

CASE 2

A 13-year-old girl was admitted in September 1989 because of sudden paraplegia and urinary retention. Myelography showed a complete blockage of flow from T6 to T8 vertebral levels. At surgery, an intramedullary spinal cord haematoma was discovered and removed, and incomplete resection of the tumour was performed. Histopathological examination revealed an oligodendroglioma (WHO grade II). Postoperative examination showed motor weakness and sensory deficit predominantly in the right leg, and urinary

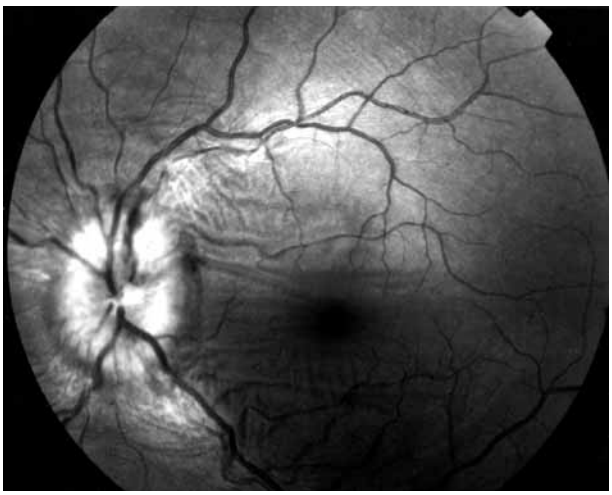

FIG. 1. — Funduscopic image of the left eye demonstrating papilloedema with peripapillary haemorrhages.

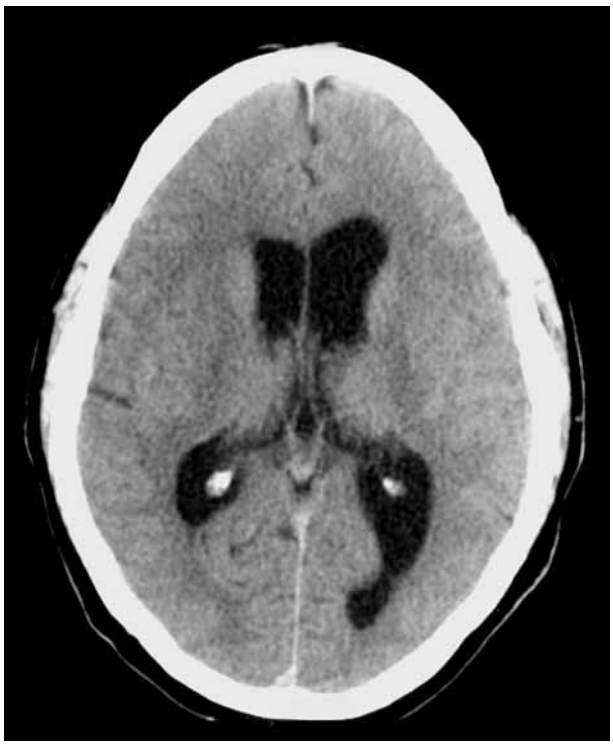

FIG. 2. — Computerized tomography scan showing a moderate hydrocephalus.

incontinence. For the next twelve years, her neurological status remained stable. Follow-up spinal MRI showed a stable image at the operation site which was consistent with tumour rest. In January 2002, she presented with a 3-month history of progressive headaches. Brain MRI showed mild enlargement of all ventricles. Examination revealed a right papilloedema without any visual disturbance. Spinal MRI disclosed a slight increase in the size of the remaining intramedullary image and tumoural progression was suspected. Lumbar CSF contained 2.23 g/l of protein with normal glucose and cell counts. The patient refused surgery, and she was treated with methyprednisolone and acetazolamide. The headaches were partially resolved but the papilloedema was seen to affect both eyes on funduscopic examination in May 2002. In August 2002, she underwent a VP shunt because of headaches, vomiting and diplopia. Ventricular CSF contained 0.61 g/l of protein and no cells. At that time, the CT scan showed significant hydrocephalus. The postoperative course was uneventful and symptoms were resolved in a few days.

CASE 3

A 49-year-old man was referred to the Ophthalmology department in August 2002 with a chief complaint of progressive bilateral visual disturbance for one month. His medical history included controlled hypertension. The ophthalmological evaluation revealed best-corrected visual acuity of 4/10 in both eyes. Funduscopic examination revealed bilateral papilloedema with surrounding peripapillary haemorrhages (Fig. 1). Brain MRI demonstrated moderate hydrocephalus with no intracranial mass lesions (Fig. 2). Neurological examination was normal but on further questioning, the patient gave a one-year history of intermittent low back pain. Spinal MRI disclosed a tissue mass occupying the entire sac at the L1-L3 vertebral levels (Fig. 3). He underwent a complete intradural excision and the postoperative course was uneventful. Histopathological examination revealed a myxopapillary ependymoma (WHO grade I). Within a month of surgery, the visual symptoms and papilloedema had been totally resolved. Brain CT scan performed 3 months later showed normal ventricles.

CASE 4

A 67-year-old woman was referred in September 2002 because of a 3-month history of neurological disturbances such as mental deterioration, gait disturbance and urinary incontinence. Brain CT scan revealed a communicating hydrocephalus (Fig. 4). She underwent a VP shunt and all symptoms were resolved in a few days. Two months later, she remained very satisfied with the operation but complained of having trouble doing her job : she was a music teacher and was unable to play the harp properly. Neurological examination demonstrated loss of dexterity in both hands and hyperreflexia in all limbs. Spinal MRI revealed an intradural mass at the C4-C6 level that was located anteriorly to the spinal cord (Fig. 5). Complete removal of a WHO grade II meningioma was performed together with VP shunt ligature. The postoperative course was uneventful and the patient was able to play the harp again. Three months later, brain MRI disclosed

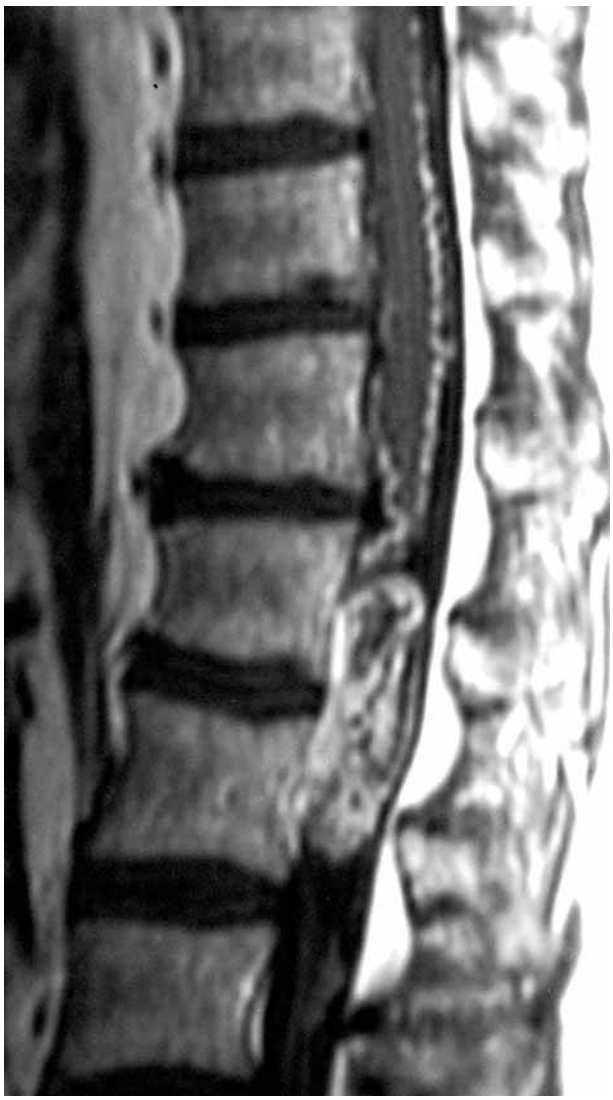

FIG. 3. — Contrast-enhanced sagittal T1-weighted MR image of the thoracolumbar junction of the spine showing a heterogeneous hyperintense lesion occupying the hole vertebral canal below the conus medullaris.

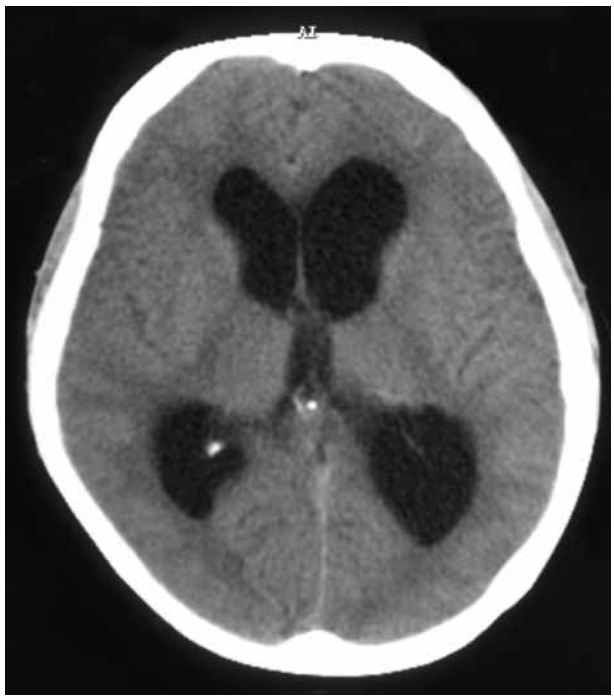

FIG. 4. — Computerized tomography scan showing a marked hydrocephalus.

normal ventricles (Fig. 6) and the VP shunt was removed. Neurological examination remained normal.

Discussion

Spinal cord tumours and contiguous nerve roots are a commonly encountered neurosurgical entity (1). Such patients classically present with a clinical history of progressive myelopathy and/or radiculopathy. Clinical symptoms and signs generally indicate dysfunction of the spinal cord and nerve roots. The association of increased intracranial pressure with an intraspinal cord tumour is an unusual phenomenon. The review of the literature shows over 200 cases with some common features : typically, a patient presenting with signs of increased intracranial pressure such as hydrocephalus, papilloedema or both (2, 21). These patients proved to be particularly confusing as they presented with complaints and findings which suggested an intracranial pathology. Therefore, initial investigations usually targeted the presumed intracranial problems and the true sites were only discovered indirectly. Consequently, a detailed history should be obtained for patients with an unexplained increase in intracranial pressure or a neuroimaging appearance of communicating hydrocephalus with headaches. Special attention should be given to previous surgery for a spinal cord tumour, back pain or lower limb weakness, and thorough neurological examination should be performed.

Several factors were considered responsible for increased intracranial pressure associated with spinal cord tumours. Raised intracranial pressure symptoms in patients with intraspinal tumours, located in the cervical region or cervicocranial junction, can be easily explained by mechanical obstruction of the cerebrospinal fluid (CSF) pathway by these tumours (16). Nevertheless, it is difficult to rationalise this association with more caudally located intraspinal tumours. Although almost one half of intraspinal tumours involve the thoracic spine (7), nearly two-thirds of those associated with raised intracranial pressure are either in the lumbar or thoracolumbar spine (16). The theory of an elevated protein content, which increases CSF viscosity and leads to mechanical clogging of the CSF absorption pathways or a rise in CSF outflow resistance, has been supported by several authors (2, 10, 20, 22, 23). Increased CSF protein content

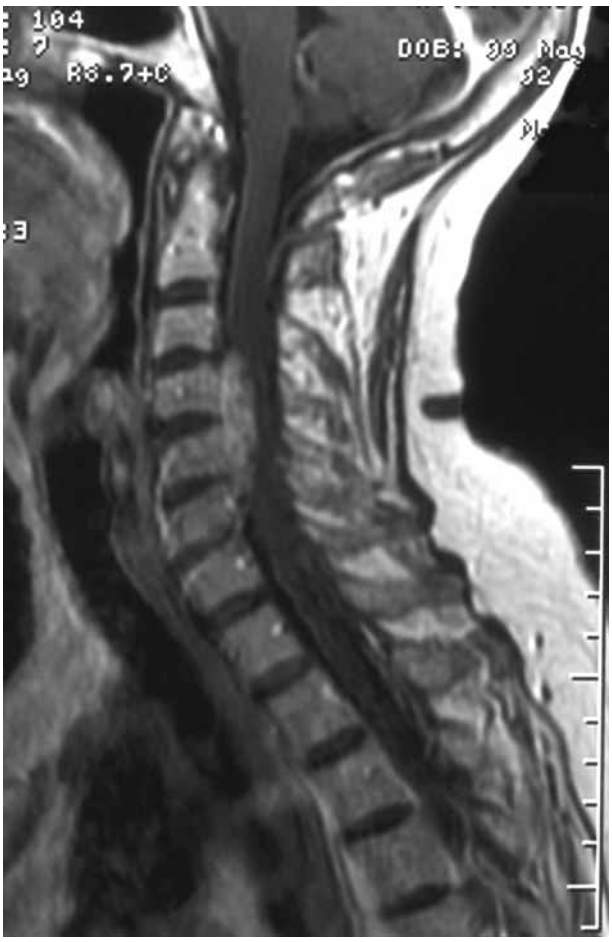

FIG. 5. — Contrast-enhanced sagittal T1-weighted MR image of the cervical spine showing a hyperintense lesion that is located in the vertebral canal at the C4-C6 level anteriorly to the spinal cord.

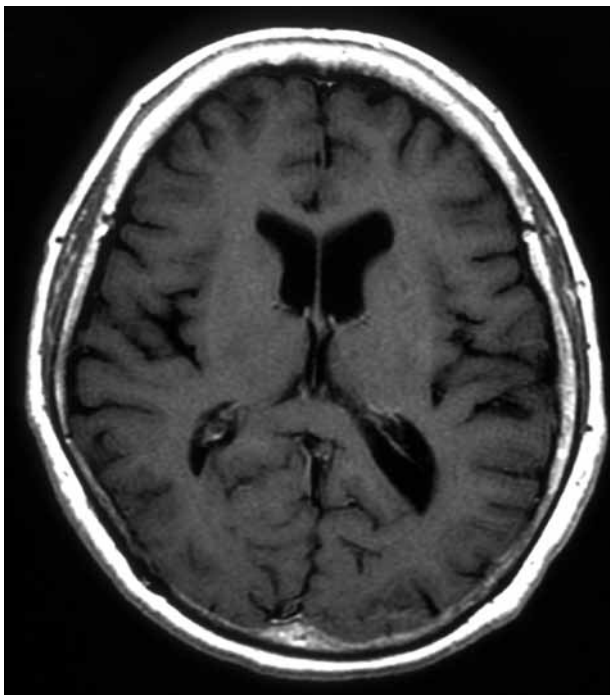

FIG. 6. — Axial T1-weighted MR image demonstrating disappearance of the hydrocephalus.

or the presence of products secreted by the tumour could also cause arachnoiditis of the posterior fossa, and this has been demonstrated by biopsy (2). However, some observations have proved that this sole hypothesis is unsatisfactory. Firstly, not all reports of hydrocephalus in patients with intraspinal tumours are associated with high protein levels (6, 14, 20, 24). Secondly, the protein concentration of the CSF rostral to the spinal tumour is usually 9 to 10 times less than in the CSF caudal to the tumour (24). Finally, it has been demonstrated that high protein concentrations do not greatly affect CSF viscosity, and that aetiology is also of little consequence (5). In cases of malignant intraspinal tumours (Case 1), pathophysiological explanations seem more likely as subarachnoid dissemination and widespread meningeal tumoural infiltration have been documented in the majority of cases (2, 15, 21). Similarly, the onset of intracranial hypertension symptoms in a patient previously operated on for a benign intramedullary spinal tumour should be considered as an early sign of recurrence (Case 2) or neoplastic intracranial seeding (6). Other mechanisms have been proposed based on impaired CSF absorption or circulation caused by tumoural subarachnoid haemorrhages (20), compression of the spinal venous plexus (11), and impaired spinal CSF reabsorption pathways around the nerve sheaths (24). Although each of these mechanisms alone is sufficient to induce communicating hydrocephalus, the rapid reversal of hydrocephalus and papilloedema after tumour removal in benign intraspinal tumours, observed by us (Cases 3 and 4) and by others (6, 11, 24), has led us to consider a dynamic theory in these cases.

During cardiac systole, there is a generalised expansion of intracranial volume due to transmission of cerebral arterial pulsations. According to the Monro-Kellie doctrine (17), intracranial content volume (brain, CSF, blood) remains constant at all times. Consequently, systolic increase in intracranial volume is dissipated by an outflow of CSF from the intracranial region, containing CSF spaces, down to the compliant spinal subarachnoid spaces, thereby producing a cranio-caudal systolic flow (3, 12, 19). Conversely, a cranio-caudal CSF flow is observed during the diastole phase (3, 12, 19). Spinal compartment compliance is based on increased elasticity of the spinal dura mater, a wide epidural space with an extended compressible epidural venous plexus, and sub-atmospheric epidural pressure (25). Cranial and spinal systems are instantaneously in equilibrium, and any variations in either compartment result in rapid compensatory changes in the other. We therefore assume that hydrocephalus in patients with benign intraspinal tumours could result from a reduction in

spinal compliance due to tumoural obstruction. In a less compliant spinal system, pressure changes will be greater for the same volume changes, and will lead to increased CSF pulse pressure. Furthermore, it has been suggested that high pulse pressure alone may be sufficient to create hydrocephalus due to a "water hammer effect" (4). The reduction in spinal compliance is partially compensated for by the vascular pool. However, if loss of spinal compliance is higher than the compensatory capacity of the vascular pool, there may be an increase in mean intracranial pressure. A flow-volume-pressure craniospinal model including the intracranial and spinal canal compartments was used to explain and quantify the relationship between arterial inflow, venous outflow and CSF flow between the cranium and the spinal canal in order to measure the intracranial compliance and pressure by a noninvasive MRI-based method (19). The intracranial pressure and compliance as well as the CSF volumetric flow rates into and out of the cranial compartment may now be estimated by using MRI-based methods including the velocity-encoded phase-contrast MRI technique (19). Finally, we would like to emphasise that increased CSF pulse pressure is not the only cause of hydrocephalus and papilloedema, and that other factors, such as CSF protein concentration, basal arachnoiditis or intracranial vascular compliance, probably combine to create a transmantle pressure gradient.

Conclusion

These four reports should serve as a reminder that communicating hydrocephalus and/or papilloedema with no intracranial explanation may be hiding an intraspinal tumour. The factors responsible for increased intracranial pressure remain unclear but are probably not univocal. In addition to previous theories proposed, we would like to add alteration of craniospinal compliance distribution. Future cases should be evaluated with a dynamic phase-contrast MRI technique to estimate intracranial and intraspinal CSF flow rates to provide a better understanding of mechanisms that lead to such a condition. A certain amount of faith should therefore be placed in the healing power of time and a shunt placement should not be planned immediately if the spinal obstruction can be removed.

REFERENCES

1. Anzil A. P. Spinal cord pathology. *Neurosurg. Clin. N. Am.*, 1994, **5** : 147-74.
2. Arseni C., Maretsis M. Tumours of the lower spinal cord associated with increased intracranial pressure and papilledema. *J. Neurosurg.*, 1967, **27** : 105-10.
3. Bergsneider M. Evolving concepts of cerebrospinal fluid pathology. *Neurosurg. Clin. N. Am.*, 2001, **36** : 631-7.
4. Bering E. A. Jr. Choroid plexus and arterial pulsations of cerebrospinal fluid : demonstration of the choroid plexus as a cerebrospinal fluid pump. *Arch. Neurol. Psychiatry*, 1955, **73** : 165-73.
5. Brydon H. L., Hayward R., Harkness W., Bayston R. Physical properties of cerebrospinal fluid of relevance to shunt function. 1 : The effect of protein upon CSF viscosity. *Br. J. Neurosurg.*, 1995, **9** : 639-44.
6. Cinalli G., Sainte-Rose C., Lellouch-Tubiana A., Sebag G., Renier D. *et al.* Hydrocephalus associated with intramedullary low-grade glioma. Illustrative cases and review of the literature. *J. Neurosurg.*, 1995, **83** : 480-5.
7. Cohen D. M., Dahlin D. C., MacCarty C. S. Apparently solitary tumors of the vertebral column. *Mayo Clin. Proc.*, 1964, **39** : 509-28.
8. Drucker D. J., Bookman A. A. Pseudotumor cerebri in association with polyarthritis, urticaria and cryoglobulinemia. *Can. Med. Assoc. J.,* 1985, **132** : 147-9.
9. Ersahin Y., Mutluer S., Yurtseven T. Hydrocephalus in Guillain-Barré syndrome. *Clin. Neurol. Neurosurg.,* 1995, **97** : 253-5.
10. Gardner W. J., Spitler D. K., Whitten C. Increased intracranial pressure caused by increased protein content in the cerebrospinal fluid : An explanation of papilledema in certain cases of small intracranial and intraspinal tumors, and in the Guillain-Barré syndrome. *N. Engl. J. Med.,* 1954, **250** : 932-6.
11. Glasauer F. E. Thoracic and lumbar intraspinal tumors associated with increased intracranial pressure. *J. Neurol. Neurosurg. Psychiatry,* 1964, **27** : 451-8.
12. Greitz D. Cerebrospinal fluid circulation and associated intracranial dynamics. A radiologic investigation using MR imaging and radionuclide cisternography. *Acta Radiol. (Suppl. 386),* **34** : 1-23.
13. Karmazyn B., Dagan O., Vidne B. A., Horev G., Kornreich L. Neuroimaging findings in neonates and infants from superior cava obstruction after cardiac operation. *Pediatr. Radiol.,* 2002, **32** : 806-10.
14. Koshu K., Tominaga T., Yasunobu F., Yoshimoto T. Quadraparesis after shunting procedure in a case of cervical spinal neurinoma associated with hydrocephalus. Case report. *Neurosurgery,* 1993, **32** : 669-71.
15. Maurice-Williams R. S., Lucey J. J. Raised intracranial pressure due to spinal tumours : 3 rare cases with a probable common mechanism. *Br. J. Surg.,* 1975, **62** : 92-5.
16. Mittal M. M., Gupta N. C., Sharma M. L. Spinal epidural meningioma associated with increased intracranial pressure. *Neurology,* 1970, **20** : 818-20.
17. Mokri B. The Monro-Kellie hypothesis. Applications in CSF volume depletion. *Neurology,* 2001, **56** : 1746-48.
18. Mortifee P. R., Bebb R. A., Stein H. Communicating hydrocephalus in systemic lupus erythematous with antiphospholipid antibody syndrome. *J. Rheumatol.,* 1992, **19** : 1299-302.
19. Raksin P. B., Alperin N., Sivaramakrishnan A., Surapaneni S., Lichtor T. Noninvasive intracranial compliance and pressure based on dynamic magnetic resonance imaging of blood flow and

cerebrospinal fluid flow : review of principles, implementation, and other noninvasive approaches. *Neurosurg. Focus*, 2003, **14** : 1-8.
20. Ridsdale L., Moseley I. Thoracolumbar intraspinal tumours presenting features of raised intracranial pressure. *J. Neurol. Neurosurg. Psychiatry,* 1978, **41** : 749-54.
21. Rifkinson-Mann S., Wisof J. H., Epstein F. The association of hydrocephalus with intramedullary spinal tumors : A series of 25 patients. *Neurosurgery,* 1990, **27** : 749-54.
22. Schaltenbrand G., Wordenhoff P. Ein einfaches Verfahren zur Bestimmung der Liquorproduktion und Liquorresorbtion in der Klinik. *Nervenarzt,* 1947, **18** : 458-63 (German).
23. Teng P., Wagner J. H., Buxbaum M. W. Giant ependymoma of the spinal cord associated with papilledema. Review of the literature and report of a case. *Arch. Neurol.,* 1960, **2** : 657-62.
24. Zavala L. M., Adler J. R., Greene C. S., Winston K. R. Hydrocephalus and intraspinal tumor. *Neurosurgery,* 1988, **22** : 751-4.
25. Zenker W., Bankoul S., Braun J. S. Morphological indications for considerable diffuse reabsorption of cerebrospinal fluid in spinal meninges particularly in the areas of meningeal funnels. An electromicroscopical study including tracing experiments in rats. *Anat. Embryol.,* 1994, **189** : 243-58.

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