CASE REPORT



Hydrocephalus associated with spinal intramedullary pilocytic astrocytoma

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ABSTRACT

Hydrocephalus secondary to intraspinal tumors is a well-known but rare condition. We report a case of holocord intramedullary pilocytic astrocytoma associated with hydrocephalus in a 29-year-old male patient. He underwent ventriculoperitoneal shunt followed by subtotal resection of the tumor.

Key words: Hydrocephalus, intramedullary tumor, spinal cord tumor, ventriculoperitoneal shunt

Introduction

Hydrocephalus secondary to intraspinal tumors is a well-known but rare condition since only about 1% of patients with spinal cord tumors have various degrees of hydrocephalus at the time of initial presentation.^[1] Since the first report by Nonne in 1900,^[1] association of hydrocephalus with intraspinal tumors has motivated a large number of publications.^[2,3] We report a case of holocord intramedullary pilocytic astrocytoma associated with hydrocephalus.

Case Report

A 29-year-old male patient was admitted with back pain and band-like sensation over the thoracic region along with bladder bowel incontinence for the past 3 years. He also had a history of progressive numbness and spastic quadriparesis (4/5, Medical Research Council grading) for the past 1 year. Contrast-Enhanced Magnetic Resonance Imaging (CEMRI) revealed an intramedullary mass lesion from the cervicomedullary junction to the T10 level [Figure 1a-c].

While waiting for operation for spinal tumor, he developed headache along with an episode of generalized tonic

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Dr. Sachin A. Borkar, Room No. 720, Department of Neurosurgery, Neurosciences Center, All India Institute of Medical Sciences, New Delhi - 110 029, India. E-mail: sachin.aiims@gmail.com clonic seizure, for which a non-contrast computerized tomography (CT) scan was done. The CT scan demonstrated hydrocephalus and peri-ventricular ooze [Figure 2]. He underwent a low-pressure ventriculo-peritoneal (VP) shunt placement [Figure 3]. During the operation, a cerebrospinal fluid (CSF) sample was obtained, which revealed a CSF protein level of 24 mg/dl. After the operation, the patient's condition improved significantly.

One week later, a midline sub-occipital craniotomy and C1-7 laminoplasty were done to decompress the intramedullary lesion. Intra-operatively, there was a poor plane of cleavage between the tumor and the surrounding normal spinal cord. Therefore, subtotal resection of the tumor was done and dorsal extension of the tumor was left for second-stage surgery. Histopathological examination revealed pilocytic astrocytoma (WHO grade-1). Intra-operative electrophysiological monitoring was not used in this case.

Postoperatively, the patient developed quadriplegia, probably secondary to high cervical spinal cord injury. He received intravenous methylprednisolone infusion for 48 h. Power in the upper limb improved, so that he could move his upper limbs against gravity (3/5, Medical Research Council grading), whereas there was no improvement in power in the lower limbs. The patient had difficulty weaning off from the ventilator so he was tracheotomized. Gradual weaning off from the ventilator was done and the patient was discharged with a

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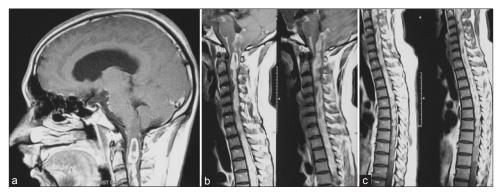


Figure 1: (a-c) CEMRI revealed an intramedullary lesion from the cervicomedullary junction to the T10 level

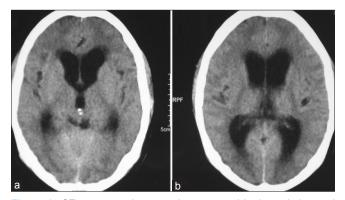


Figure 2: CT scan on admission demonstrated hydrocephalus and peri-ventricular ooze

plan to re-admit later for second surgery for the residual tumor. The patient was re-admitted in emergency with chest infection, with sepsis and large bed sores in the sacral and trochanteric area. He was managed with intravenous antibiotics and daily dressings. Second surgery was deferred in view of the poor general status of the patient.

Discussion

Hydrocephalus associated with intraspinal tumors is very rare.^[1] Among these tumors, the most commonly reported are neurinomas and gliomas.^[3]

Many authors have discussed the pathogenesis of hydrocephalus in association with intramedullary tumors, and the possible causes of hydrocephalus include elevation of CSF protein content, leptomeningeal infiltration by tumor cells, and obliteration of the cisterna magna due to rostral extension of the tumor.^[4] Finally, it is likely that the two patho-physiological mechanisms proposed by Bamford and Labadie^[5] and Maurice-Williams and Lucey^[6] could co-exist at different stages of the evolution of the disease. At an early stage, abnormal presence of fibrinogen in the CSF and its conversion to fibrin at the level of Pacchioni's granulations would cause an increase in CSF outflow resistance and communicating hydrocephalus.^[7,8] The stagnation of the

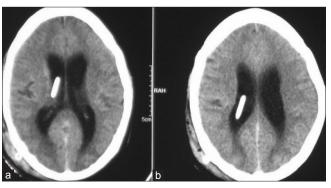


Figure 3: CT scan after right ventriculoperitoneal shunt demonstrated shunt tip *in situ* with decompressed ventricles

CSF that results causes further formation of fibrin nets in the subarachnoid spaces of the cerebral convexities and the cranial base, and their organization in fibrous tissues.^[9] At a later stage, in the cases of intramedullary tumors of the glial origin, formation of subarachnoid adhesions probably becomes a predisposing factor for further implantation of neoplastic cells and leptomeningeal dissemination after surgical trauma and shunting procedures, as already proposed by Russell and Rubinstein^[10] for leptomeningeal seeding in intracranial tumors. The neoplastic seeding favored by these factors would induce an irreversible and self-maintaining condition that would explain the rarity of curing hydrocephalus after tumor removal, the frequent late onset of hydrocephalus in the absence of local recurrence of spinal lesion, and the high mortality rate in this group of patients.

Conclusion

This rare case highlights the importance of suspecting hydrocephalus, and investigating and managing it accordingly, in cases of holocord spinal intramedullary tumors.

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Conflicts of interest

There are no conflicts of interest.

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