Intra-Fourth Ventricular Schwannoma With Obstructive Hydrocephalus – A Rare Case Report

A Babbu, R Katheerayson

Citation

Abstract
A rare case of intra-fourth-ventricular schwannoma with obstructive hydrocephalus in a 20-year-old male is described. CT and MRI showed schwannoma having cystic and solid components inside the fourth ventricle causing supra-tentorial obstructive hydrocephalus. Emergency right ventriculo-peritoneal shunt followed later by successful surgical resection of the tumor was done. Histo-pathological examination revealed a benign schwannoma. The clinical, radiographic, surgical, histopathological features and the aetiology of this tumor are elaborated. This rare intra-fourth-ventricular schwannoma is the first case reported in Hospital Pulau Pinang, Malaysia and also would be one of the very few cases reported in the literature.

CASE SUMMARY
A 20-year-old gentleman presented in a drowsy state (one week) with features of increased intra-cranial tension, ophthalmoparesis, brainstem & cerebellar compressions. There were no neuro-cutaneous markers and no family history of neurofibromatosis. CT-Brain & MRI-Brain showed a huge intensely contrast enhancing intra-fourth ventricular tumor compressing the brainstem and cerebellum causing obstructive hydrocephalus.

Figure 1
Fig. 1 CT-Brain (Plain & Contrast)

Figure 2
Fig. 2 MRI – Brain T1W & T2W
A pre-operative diagnosis of Choroid Plexus Papilloma / Ependymoma was made and an emergency right ventriculo-peritoneal shunt done. Shunt tube is found to be in-situ.

This was followed by posterior fossa craniotomy for surgical resection of the tumor. The gross appearance of the tumor was greyish, fleshy, firm and vascular with cystic components. Vermian split done to facilitate the excision of the whole tumor. Upon complete excision of the tumor CSF was flowing within the fourth ventricle.

Postoperatively there was an improvement in the conscious level and swallowing of the patient as well his headache symptom. There was no improvement in cerebellar signs. Patient could walk with support.

Histo-pathology of the tumor specimen under microscopy
showed areas of Antoni A (hypercellularity), Antoni B (hypocellularity). Focal areas of cystic changes seen. No necrosis and rare mitosis seen. In immuno-histochemistry, the lesional cells expressed S-100 protein (neural marker). The final impression was benign schwannoma.

**Figure 7**
Fig. 7 Histopathological slide – under low power

**Figure 8**
Fig. 8 Histopathological slide – under high power

**DISCUSSION**
Intracranial schwannomas comprise approximately 8% of all intracranial tumours. The incidence of intraventricular schwannoma is very rare. There are few case reports in the literature: (1) spinal accessory schwannoma mimicking a tumor of the fourth ventricle, (2) midline cerebellar cystic schwannoma, (3) a schwannoma arising from the dorsum of the pontomedullary junction and presenting as an exophytic mass in the fourth ventricle, (4) two cases of cystic schwannoma of the fourth ventricle presenting with hemifacial spasm. A variety of hypotheses have been proposed regarding the possible mode of origin of schwannoma unrelated to cranial nerves. In our case we propose the probable origin could be from the schwann cells in perivascular plexus.

Intra-ventricular schwannomas are rare tumors that are amenable to complete surgical removal having a good prognosis without the need of adjuvant therapy. In our case we could completely excise the tumor and he has not been on any adjuvant therapy. His follow-up scans showed no residual or recurrent tumor. We would be from Hospital Pulau Pinang, one of the few, to add to this rare intra-fourth ventricular schwannoma to the literature.

**ACKNOWLEDGEMENT**

1. Dr. Lee Suk Kam

Specialist Pathologist, Department of Pathology,

Hospital Pulau Pinang

**References**

8. Spinal accessory schwannoma mimicking a tumor of the fourth ventricle: case report. Neurosurgery 2004 Feb; 54(2):510-4; discussion 514 Kurokawa R, Tabuse M, Yoshida K, Kawase T Department of Neurological Surgery, Keio University School of Medicine, Japan
9. Howard L, Weiner1 , David Zagzag2, Ramesh Babu1, Herman J. Weinreb3 and joseph Ransohoff1 Schwannoma of the fourth ventricle presenting with hemifacial spasm Journal of Neuro-oncology 0167-594X (Print) 1573-7373 Volume 15, Number 1 / January, 1993
Author Information

A.Daniel Rajesh Babbu
Associate professor, AIMST University

Ravindran Katheerayson
Consultant Neurosurgeon & HOD of Neurosurgery, Hospital Pulau Pinang