

cerebellum, spinal cord, and brainstem.^[2] The unusual locations include leptomeninges and sellar-sphenoid sinus.^[4-6] Supratentorial location accounts for 4 to 13% of the cases.^[2] However, hemangioblastomas of the lateral ventricle are extremely rare and only 5 cases have been reported in the English literature till 2001.^[2] We report a unique case of a lateral ventricular hemangioblastoma.

A 30-year-old male presented with a progressive holocranial headache of six months duration. The neurological examination was normal. Magnetic resonance imaging (MRI) of the brain showed a 3 x 2 cm mass in the left lateral ventricle, which was hypointense on T1-weighted images, hyperintense on T2-weighted as well as fluid attenuated inversion recovery (FLAIR) images. The tumor showed brilliant contrast enhancement [Figures 1-3]. The patient had a left-sided parietal craniotomy and through a trans-sulcal approach through the superior parietal

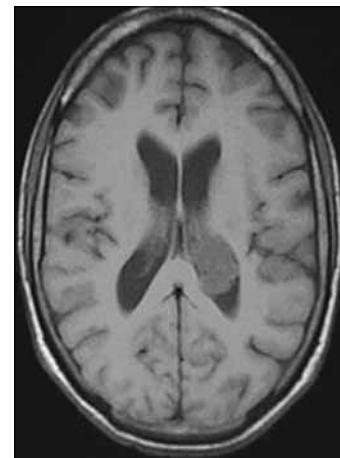


Figure 1: T1-weighted axial magnetic resonance imaging scan of the patient showing a mass in the body and atrium of the left lateral ventricle which is hypointense compared to the normal white matter

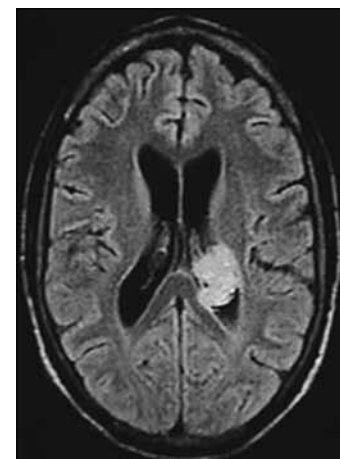


Figure 2: T2-weighted fluid attenuated inversion recovery axial scan of the patient showing a mass in the body and atrium of the left lateral ventricle which is hyperintense compared to the normal white matter

Hemangioblastoma of the lateral ventricle

Sir,

Hemangioblastomas are the most frequent vascular tumor of the central nervous system (CNS)^[1] and can be associated with von Hippel-Lindau (VHL) disease in 3 to 38% of the cases as a major manifestation.^[2] These tumors are predominantly found in the

lobule the lesion was excised. At operation the lesion was brownish red, soft to firm, highly vascular, and was adherent to the ventricular ependyma as well as the choroid plexus. An immediate post-operative computed tomography (CT) scan showed complete excision of lesion [Figure 4]. Post-operatively, the patient had transient sensory aphasia, which recovered fully in a 1 month. A histopathological examination showed tissue composed of a fine network of blood spaces separated by numerous polygonal stromal cells, with lightly stained cytoplasm [Figure 5]. Immunohistochemical staining showed a strong immunopositivity in several stromal cells for vimentin, epithelial membrane antigen (EMA), and neuron specific enolase (NSE) - typical of a hemangioblastoma. The patient was investigated for other features of von Hippel-Lindau syndrome but none were found.

Hemangioblastomas of the CNS are infrequent and they account for 2% of primary CNS tumors.^[1,3] The most common location is the posterior fossa, cerebellar hemisphere accounting for 83%.^[3] Supratentorial location is rare, sporadic hemangioblastomas 4% and VHL disease 13%.^[2] The lateral ventricular location is extremely unusual, only four symptomatic cases^[7-10] and one incidental postmortem finding^[8] have been described. All the four lateral ventricle hemangioblastomas reported till date had been in patients with VHL disease.^[7-10] In our patient we could not find any evidence for VHL disease. However, it is prudent to investigate patients with lateral ventricular hemangioblastoma for VHL disease. The size of the lateral ventricle hemangioblastomas in all the four documented cases has been greater than the size of the hemangioblastoma in other locations. This may partly be related to the fact that the volume of the lateral ventricle can accommodate a reasonable size mass lesion and any mass lesion in the lateral ventricle has to grow to a sufficient size to produce symptoms. In other locations cystic component of hemangioblastoma has been the contributing factor to the size of the tumor, whereas all the lateral ventricle hemangioblastomas including our case were solid.^[7-10]

Hemangioblastoma of the lateral ventricle is associated with a better prognosis than its counterpart in the third ventricle. Diehl and Symon were able to achieve complete resection of the tumor.^[11] Our experience was also similar. We were able to achieve good control of the feeding vessels from the choroids plexus and the wall of the lateral ventricle, thus enabling in toto resection of the tumor. Post-operative dysphasia/mutism is an infrequent and usually transient complication of hemangioblastoma surgery. In our case, it was probably related to the proximity of the

tumor to the posterolateral region of the thalamus and/or surgery-related edema and/or ischemia in

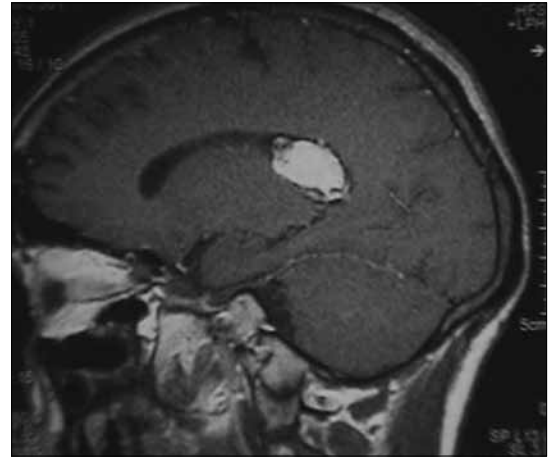


Figure 3: T1-weighted sagittal magnetic resonance imaging scan of the patient showing brilliant contrast enhancement of the lateral ventricular mass

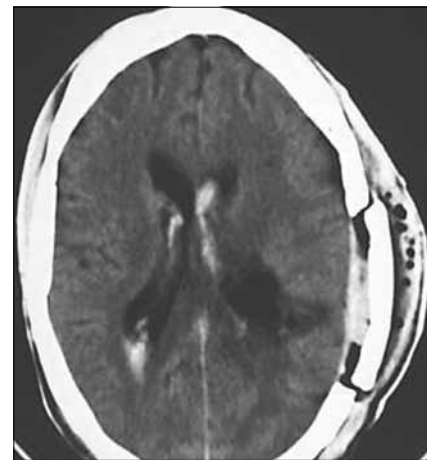


Figure 4: Post-operative axial head computed tomography scan of the patient showing complete excision of the left ventricular lesion

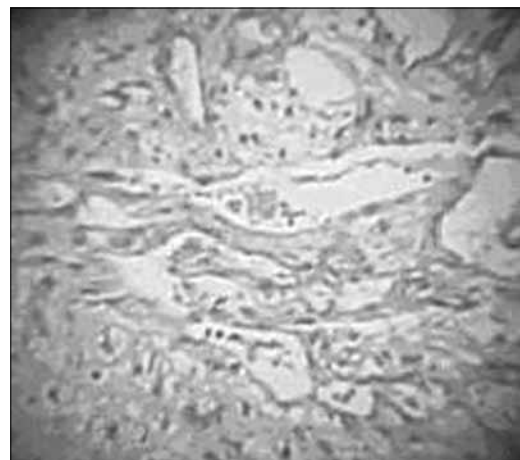


Figure 5: Histopathological photograph showing a tissue composed of a fine network of blood spaces separated by numerous polygonal stromal cells, with lightly stained cytoplasm

the parietal region. Its incidence can be reduced by delineating the speech area by functional MRI preoperatively^[12] or by using intraoperative cortical mapping.^[13]

**Ramandeep Singh Jaggi,
Ishwar Chandra Premsagar, Abhishek**

*Department of Neurosurgery, Dr. R.M.L. Hospital, Baba Kharak
Singh Marg, New Delhi - 110 001, India.
E-mail: rsjaggi2000@yahoo.com*

PMID: 19934579

DOI: 10.4103/0028-3886.57797

References

1. Strugar J, Criseuolo G. Primary intracranial vascular tumors. In: Vecht C, editor. Handbook of clinical neurology. Neuro-Oncology, 1997. Part II, pp 269-86.
2. Conway J, Chou D, Clatterbuck R, Brem H, Long DM, Rigamonti D. Hemangioblastomas of the central nervous system in von Hippel-Lindau syndrome and sporadic disease. *Neurosurgery* 2001;48:55-63.
3. Weil RJ, Vortmeyer AO, Zhuang Z, Pack SD, Theodore N, Erickson RK, *et al.* Clinical and molecular analysis of disseminated hemangioblastomatosis of the central nervous system in patients without von Hippel-Lindau disease. *J Neurosurg* 2002;96:775-87.
4. Reyns N, Assaker R, Louis E, Lejeune JP. Leptomeningeal hemangioblastomatosis in a case of von Hippel-Lindau disease: Case report. *Neurosurgery* 2003;52:1212-6.
5. Sajadi A, Tribollet N. Unusual locations of hemangioblastomas. *J Neurosurg* 2002;97:727.
6. Kachhara R, Nair S, Radhakrishnan VV. Sellar-sphenoid sinus hemangioblastoma: Case report. *Surg Neurol* 1998;50:461-4.
7. Prieto R, Roda JM. Hemangioblastoma of the lateral ventricle: Case report and review of the literature. *Neurocirugia (Astur)* 2005;16:58-62.
8. Vecchi B, Patrassi G. Angioreticuloma del plessi corioidei, con "aree di Gamma". *Schweiz Med Wochenschr* 1935;65:242-6.
9. Ho YS, Plets C, Goffin J, Dom R. Hemangio-blastoma of the lateral ventricle. *Surg Neurol* 1990;33:407-12.
10. Murakami H, Toya S, Otani M, Sato S, Ohiera T, Takenaka N. A case of concomitant posterior fossa and supratentorial hemangioblastomas. *No Shinkei Geka* 1985;13:175-9.
11. Diehl P, Symon L. Supratentorial intraventricular hemangioblastoma: Case report and review of literature. *Surg Neurol* 1981;15:435-43.
12. Hall WA, Kim P, Truwit CL. Functional magnetic resonance imaging-guided brain tumor resection. *Top Magn Reson Imaging* 2009;19:205-12.
13. Brell M, Conesa G, Acebes JJ. Intraoperative cortical mapping in the surgical resection of low-grade gliomas located in eloquent areas. *Neurocirugia (Astur)* 2003;14:491-503.

Accepted on 10-2-2009