

Intraventricular cystic meningioma

ABSTRACT

We report a case of a 45-year-old male patient with intraventricular cystic meningioma located in the left lateral ventricle. He presented with complaints of global headache, progressively increasing loss of memory, and frequent episodes of abnormal behavior, of 1 month duration. At the time of hospital admission, his general and neurological examination was normal. Neuroimaging studies showed a left lateral ventricular enhancing mass, composed of mixed solid and cystic areas. The tumor was completely excised via the anterior transcallosal approach. A histological examination revealed a meningothelial meningioma without any atypia. The aim of this report is to present the occurrence of an intraventricular cystic meningioma.

KEY WORDS: Cystic, intraventricular, meningioma

INTRODUCTION

Meningiomas are relatively common entities, accounting for approximately 15–18% of all intracranial tumors in adults.^[1-5] In contrast, meningiomas occurring primarily within the ventricular system are rare.^[2] Cases where these tumors have attained a substantial size at diagnosis often mimic a variety of other intraventricular neoplastic conditions. Most of these cases are managed by total excision of the tumor, and are associated with a favorable prognosis.

We report a rare case of an intraventricular meningioma in an adult male which was associated with a cystic morphology.

CASE REPORT

A 45-year-old male presented with complaints of global headache, progressively increasing loss of memory, and frequent episodes of abnormal behavior, of 1 month duration. There was no history of nausea or vomiting.

At the time of hospital admission, his general and neurological examination was normal. There was no evidence of raised intracranial pressure, motor or sensory deficits, or homonymous hemianopsia.

Routine laboratory investigations were within normal limits.

Neuroradiology

Neuroimaging studies showed a left lateral ventricular solid cystic tumor. Both solid and cyst wall components showed contrast enhancement.

The solid component and the wall of the cystic component were hypointense on T2-weighted images (T2WI) [Figure 1a and b] and hyperintense on FLAIR [Figure 1c], and showed enhancement on post-gadolinium T1WI [Figure 1d–f]. In contrast, the cystic component was hyperintense on T2WI, and slightly hyperintense to cerebrospinal fluid on FLAIR [Figure 1c] and T1-weighted images (T1WI) [Figure 1–f]. The left lateral ventricle was enlarged without any periventricular T2 hyperintensity. In addition, FLAIR axial showed dilated temporal horns (left more than right), without periventricular hyperintensity indicating compensated hydrocephalus.

Operative intervention and findings

Total excision of the tumor was performed by the anterior transcallosal approach. Intraoperatively, the tumor was seen occupying the body and antrum of the left lateral ventricle. It consisted of soft, grayish-white solid areas, with a multiseptate cystic component containing a yellow-colored fluid.

Histopathological examination

The tissue was fixed in 10% neutral buffered formalin, routinely processed, and paraffin embedded. Five-micron-thick sections were cut and stained by hematoxylin and eosin (H and E) stain. Subsequently, immunohistochemical staining was done by streptavidin-biotin immunoperoxidase technique (LSAB Kit, M/s Dakopatts, Denmark) using monoclonal antibodies to epithelial membrane antigen (EMA), vimentin, S-100 protein, glial fibrillary acidic protein (GFAP), and synaptophysin, while the proliferation index was evaluated using MIB-1 antibody staining (prediluted, M/s BioGenex).

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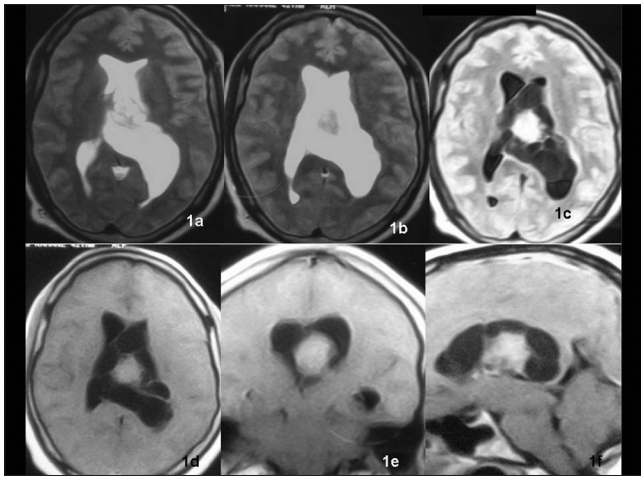


Figure 1: T2W axial (a and b), FLAIR axial (c), postcontrast T1W axial, coronal and saggital (d–f). Left lateral intraventricular mass, with a central solid and peripheral multicystic component, with an enhancing central solid component and enhancing walls of the cyst. The solid component is hypointense on T2W images and hyperintense on FLAIR. The cystic component is hyperintense on T2WI, and slightly hyperintense to CSF on FLAIR and T1W images. Left lateral ventricle is enlarged with no periventricular T2 hyperintensity

A microscopic examination revealed a meningothelial meningioma with large cystic spaces lined by meningothelial cells. Tumor cells were immunoreactive to EMA, vimentin, and S-100 protein, and immunonegative for GFAP and synaptophysin. There was no mitosis, necrosis, or vascular proliferation [Figure 2 a–c]. MIB-1 labeling index (MIB-LI) was < 1%.

Follow-up

A contrast-enhanced computed tomography (CECT) scan obtained on the second postoperative day showed a complete excision of the tumor. Postoperative recovery was uneventful. At the last review, after 6 months of surgery, he was asymptomatic, ambulant, and without any neurological deficit or any residual tumor on CECT scan or MRI.

DISCUSSION

Meningiomas are common intracranial tumors in adults, with only 0.7–2% of cases arising in an intraventricular location, with the majority (80%) being located in the lateral ventricles.^[2] Most of the intraventricular meningiomas reportedly occur in the 30- to 60-year age group (mean, 42 years), which is similar to the present case. Females are more likely to be affected by a 2:1 ratio. In the pediatric population, approximately one in five meningiomas occur in the ventricular system.^[4,5]

Patients with intraventricular meningiomas commonly present with features of raised intracranial tension (nausea, vomiting, headache), contralateral sensory or motor deficits, and homonymous hemianopsia,^[1-5] which are characteristically lacking in the index case.

It is believed that intraventricular meningiomas arise from the arachnoidal cap cells trapped within the choroid plexus, the tela choroidea, or the velum interpositum, owing to which, the trigone of the lateral ventricle is reported to be the most common site. Interestingly, intraventricular meningiomas constitute the most common trigonal mass in adults.^[2] The involvement of the third ventricle or the fourth ventricle is rare.^[1-5]

On imaging, intraventricular meningiomas are usually seen as a well-defined globular mass, displaying hyperattenuation compared with the adjacent brain parenchyma on a CT scan. On MRI, these are iso- to hypointense on T1W images and iso- to hyperintense on T2W images. These tumors show brilliant enhancement and are usually homogeneously solid. The degree of ventricular dilatation however depends on the extent of CSF obstruction. Imaging may also display calcification (50%), and periventricular edema, possibly due to the reversal of the transependymal CSF flow. Rare cases may also have associated subarachnoid or intraventricular hemorrhage.^[1-5] MR spectroscopy however appears to be more informative, and its pattern of decreased amounts of *N*-acetylaspartate and creatine; increased amounts of choline; and variable amounts

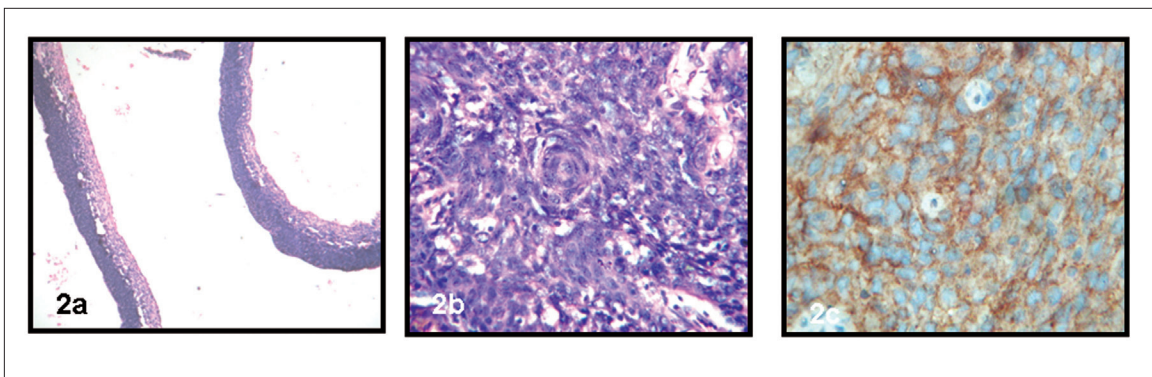


Figure 2: Photomicrograph of the cystic lesion (a: H and E, x40). Focally, the cyst wall contained aggregates of meningothelial cells forming whorls (b: H and E, x100). Tumor cells were strongly immunopositive for EMA (c: EMA, x200) confirming the diagnosis of the meningothelial meningioma

of lactate, lipids, and alanine is similar to that of meningiomas in other areas.^[6]

Cystic meningiomas or meningiomas with cystic morphology are also rare and account for only 0.7–11.7% of all meningiomas.^[7,8] Most of these are documented in the literature as isolated case reports only.^[6,7,9] The intraventricular location of cystic meningioma is exceptional, with very few cases reported till date.^[9,10] Contrary to the present case, cystic meningiomas tend to affect children and have a predilection for convexity and parasagittal sinus. Nauta *et al.*,^[11] has classified cystic meningiomas into types 1–4, based on the location of the cavity and the relationship between the tumor and the surrounding brain. Tumors with a centrally located intratumoral cyst are considered as type 1, while type 2 tumors have peripherally located intratumoral cysts. In contrast, type 3 has a peritumoral cyst with walls consisting of both adjacent parenchyma and the tumor, and type 4 cystic meningioma has a peritumoral cyst with walls formed by the arachnoid, while the cyst remains separated from the tumor by a distinct capsule. The present case represents a type 2 cystic meningioma.

The various theories proposed for cyst formation in a meningioma include ischemic necrosis, cystic degeneration, intratumoral hemorrhage, trapping of CSF, peritumoral edema into cyst, active secretion by tumor cells, and glial reaction and transudation.^[8]

An intraventricular cystic lesion, like in the index case, evokes multiple differential diagnoses on imaging studies, which include ependymoma, choroid plexus tumor, neurocytoma, metastatic tumor, glioma arising from the fornix, and subependymal giant cell astrocytoma.^[11] Histopathology, along with ancillary techniques of immunohistochemistry, is essential in such cases to reliably differentiate a meningioma from the above-mentioned neoplastic conditions.

Unlike pediatric cases, most of the intraventricular meningiomas in adults are of the menothelial type, displaying a benign histology with a low proliferation index,^[1-5] which respond well to total surgical excision. In one of the largest series of intraventricular meningiomas, Criscuolo and Simon^[12] noted recurrence in 2 out of 10 cases. One of the recurrent tumors was a highly malignant cystic meningioma which recurred within 10 weeks of surgery and proved fatal shortly thereafter.

The aim of this report was to highlight an extremely rare

manifestation of a common tumor. Owing to its atypical clinical presentation, compounded with a large tumor in an intraventricular location having cystic morphology, it had evoked possibilities of other neoplastic entities on imaging. However, histopathology proved vital in confirming the nature of the tumor, thus highlighting its role not only in diagnosis but also in prognostication, since these tumors are generally low grade; amenable for complete resection; and associated with a good prognostic outcome.

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