Pediatric Intraventricular Meningioma: Radiopathological Study and Review of the Literature

Lama Al-Mudaimiegh

Department of Medical Imaging, King Fahad Medical City, Riyadh, Saudi Arabia

Corresponding Author: Lama Al-Mudaimiegh
L-almudaimeegh@hotmail.com

ABSTRACT

Pediatrics intraventricular meningiomas are rare poor prognosis tumors with reported incidence of 0.5-5% of all meningiomas. The presentation is in the form of raised ICP; therefore the definite diagnosis is based on neuroimaging studies and histopathology. These tumors pose a huge surgical challenge for neurosurgeons due to its deep location and risk of intraoperative massive blood loss. Here we describe the case of a 5-year-old boy presented with 2 months of progressive headaches associated with nausea and unsteadiness. Fundoscopy showed bilateral papilloedema. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed large solid intraventricular mass within the left lateral ventricle. On histology and immunohistochemistry, a diagnosis of intraventricular meningioma was made. Postoperatively the patient did not receive chemotherapy or radiotherapy, and he was doing well after 18 months follow-up and disease free on clinical and radiological examination.

Keywords: Pediatrics, Neoplasm, Neurosurgery, Neuroimaging, Pathology

1. INTRODUCTION

Primary intraventricular meningiomas are rare tumors comprise approximately 2% of all intracranial meningiomas primarily present between the 4th and 6th decades (1,2). Pediatric intraventricular meningiomas are even rarer with reported incidence of 0.5-5% of all meningiomas (3). These tumors pose a huge surgical challenge for neurosurgeons due to their deep location and risk of massive blood loss during surgery. Our case reviews the presentation of giant primary intraventricular meningioma, along with the imaging workup and pathological correlation in a pediatric patient. Literature of primary intraventricular meningioma is reviewed.

2. CASE REPORT

The tumor was detected in a 5-year-old boy was admitted with a 2 months history of progressive headaches associated with nausea and unsteadiness. The patient's antenatal and perinatal period was uneventful, and he had achieved normal developmental milestones.

On physical examination, the patient was stable, communicating, oriented and not in pain. Neurological examination revealed no neurological deficits. Fundoscopy revealed bilateral papilloedema.

Radiological studies as CT of brain and MRI brain were performed. A CT scan of the brain (Figure 1A&B) showed a well-defined hyperdense lesion lying within the body of the left lateral ventricle that enhanced intensely after contrast administration.
Figure 1(A, B): Axial noncontrast computed tomography scan of the brain showing well defined hyperdense lesion in the body of the left lateral ventricle with perilesional edema.

Magnetic resonance imaging of the brain (MRI) with gadolinium enhancement [(Figure 2A-D)] revealed large well defined lobulated mass in the left lateral ventricle which was hypointense on T1 weighted images, iso/hyperintense on T2 weighted images, hyperintense on the Fluid-attenuated inversion recovery (FLAIR) sequence. It enhanced intensely after administration of contrast material. The mass cause a significant dilatation of left lateral ventricle and mass effect on adjacent brain parenchyma. An abundance of edema was present in the surrounding white matter.

Figure 2(A-D): Pre-operative brain MRI

A) axial T2-weighted, B) coronal T2-weighted, and C) axial FLAIR images, D) sagittal T1-weighted.

Based on radiology, a differential diagnosis of choroid plexus papilloma and intraventricular meningioma was considered. The patient underwent total excision of the tumor using the parietooccipital (trigonal) approach. Intraoperative findings revealed the tumor was hard in consistency. Staging biopsies were performed, and samples were sent to his to pathology. The patient had a smooth operative period with no complication. Histopathological examination revealed fibroblastic type meningioma (WHO grade I). According to microscopy, the mass was moderately cellular meningioma composed of predominant whorling pattern and several psammoma bodies with a moderate amount of eosinophilic cytoplasm with mild nuclear atypia. There was no necrosis nor invasion of neighboring neural parenchyma, and the mitotic activity was found to be 0-1/10 BBA. The Ki-67 proliferation index was 3%. Fibrotic content 40by using Masson Trichrome stain. (Figure 3).

**Figure 3: Microscopic image of Hard consistency of our case as assessed by Masson’s trichrome stain.**

The patient showed improvement and was discharged after a week without complications. Post operation, the patient, did not receive chemotherapy or radiotherapy, he is currently asymptomatic and without signs of disease relapse after 18 months of follow-up.

3. **DISCUSSION**

Meningiomas arise from arachnoid cap cells. In a similar fashion, primary intraventricular meningiomas thought to arise from meningothelial inclusion bodies located in the tela choroidea and/or mesenchymal stroma of the choroid plexus[4]. Most commonly they are located in the lateral ventricle trigon (77.8%), followed by third ventricle(15.6%) and rarely in the fourth ventricle (6.6%) [5]. Pediatric Meningiomas are slow-growing tumors that become large at presentation. Raised intracranial pressure was the predominant symptom at presentation in most patients with lateral ventricular meningiomas followed by visual field deficits and seizures are rare. Meningiomas of the third and fourth ventricle present with obstructive hydrocephalus [3]. Neuroimaging is necessary for the diagnosis of intraventricular meningiomas. Their signal and attenuation characteristics on CT scans and MRI are the same as other meningiomas, demonstrating isodensity essentially and intensity to gray matter precontrast and vivid, usually homogeneous enhancement following administration of contrast. Areas of calcifications, necrosis and cystic change may be present [6].

The management of these tumors is challenging because of their large size at presentation, peculiar location, and aggressive behavior [7]. Surgical resection is the gold standard for treatment. However, management of these tumors requires careful planning cause surgery carries a huge risk and high mortality rate. According to Dash et al. Surgeons should watch out for massive blood loss during surgery, especially via the parietooccipital transcortical approach. Radiosurgery is a viable option in patients who are not fit for general anesthesia or who refuse consent for surgery. Radiation therapy is indicated for residual lesions, high-grade tumors, or recurrent tumors. Histopathological features of intraventricular meningiomas tumors were similar to those seen in dural meningiomas. These tumors can be any of the histopathological tumor types (predominantly fibrous, fibroblastic, meningothelial, or psammomatous) defined by the World Health Organization classification of meningiomas [8]. Angiomatic tumors were the most common type seen in our series, followed by meningothelial, fibroblastic, and psammomatous/osteoblastic types [9]. Prognosis in pediatrics population is poor as compared to adults. The outcome is also poor in tumors with higher grade, lesions that demonstrate cortical invasion and in patients with neurofibromatosis [7].

4. **CONCLUSION**

Pediatrics primary intraventricular meningiomas are extremely rare. It has a non-specific clinical presentation. Therefore, neuroimaging studies and
Histopathology are essential tools for definite diagnosis. Surgery remains the cornerstone of intraventricular meningiomas treatment. Although prognosis is poor, the good news that if it diagnosed in an early stage, the treatment will be effective and improve prognosis.

REFERENCES