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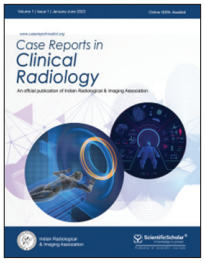

Case Report

Unusually large craniopharyngioma: A case report

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ABSTRACT

Background: Craniopharyngiomas are benign slow-growing tumors of the central nervous system originating from squamous remnants of incompletely involuted craniopharyngeal duct. Craniopharyngiomas are said to be "giant" when they attain sizes of 5–6 cm or more.

Case Presentation: Here, we present a case of unusually large craniopharyngioma in a child, which, to the best of our knowledge, is the largest craniopharyngioma reported in the literature. The provisional diagnosis of craniopharyngioma was established using computed tomography and magnetic resonance imaging.

Conclusion: Craniopharyngiomas are to be considered in the differentials for sellar and suprasellar region lesions causing mass effect. The provisional diagnosis is established by imaging but definitive diagnosis always requires histopathology.

Keywords: Craniopharyngeal duct, Adamantinomatous craniopharyngioma, Papillary craniopharyngioma

INTRODUCTION

Craniopharyngiomas are slow-growing benign tumors occurring in the sellar/suprasellar region. Occasionally they can attain huge sizes and extend far into surrounding regions, thereby posing diagnostic difficulty. Here we discuss such a case which turned out to be a large craniopharyngioma though it was hard to believe due to its size.

CASE REPORT

A 12-year-old boy presented to the pediatric emergency with severe headache and vomiting. He was confused and complained of blurred vision. His attendant said that he had been complaining of vision abnormalities for the past 6 months and his family members had also noticed changes in his personality lately. The initial workup included serum electrolyte and blood sugar levels, complete blood counts, and electrocardiogram, all of which came out to be normal. Computed tomography (CT) scan was done which demonstrated a large cystic hyperdense mass with rim calcification at the sellar and suprasellar region extending to the anterior and middle cranial fossa and encasing the Circle of Willis [Figure 1]. Magnetic resonance imaging (MRI) was ordered to better delineate the mass and its relationship to adjacent neurovascular structures [Figure 2].

MRI revealed a hyperintense mass lesion extending from the basifrontal region up to the craniovertebral junction in sagittal T1-weighted image (T1W1). In the coronal T2 fluid-attenuated inversion recovery (FLAIR) sequence, a large hyperintense mass was seen in the sellar, suprasellar region with bilateral parasellar extension. Axial T2 image further characterized the mass to have a lobulated surface in the frontal and basifrontal regions. The mass was surgically

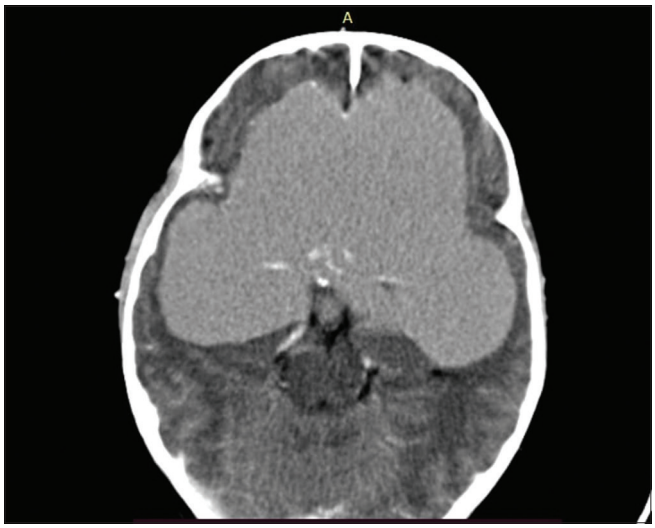

Figure 1: Axial contrast computed tomography showing mass lesion encasing the circle of Willis.

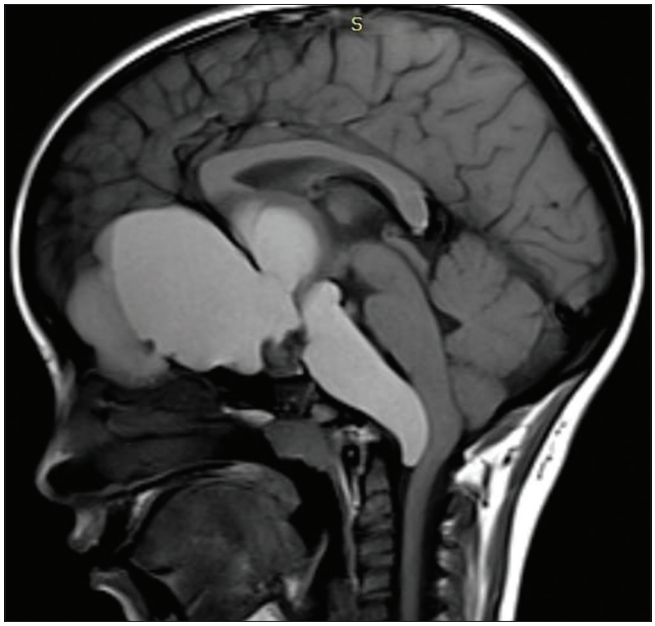

Figure 2: Sagittal T1 image showing hyperintense mass lesion extending from basifrontal region up to cervicovertebral junction.

excised and histopathologic examination revealed a typical adamantinomatous craniopharyngioma [Figure 3].

DISCUSSION

Craniopharyngioma is a slow-growing benign sellar/suprasellar region mass likely arising from squamous epithelial remnants of the Rathke's pouch. They represent 2–5% of all primary intracranial tumors with an incidence rate of 1–2 cases/million a year. About 30–40% of cases present during adolescence while around 25% are diagnosed in patients over age 25.[1] Craniopharyngioma is the most common non-glial neoplasm in children.

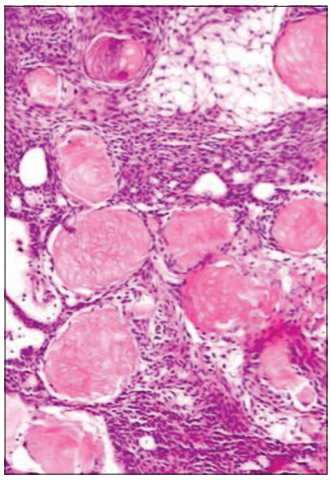

Figure 3: Histopathology showing adamantinomatous craniopharyngioma having peripheral layers of palisading stratified squamous epithelium surrounding nodules of wet keratin.

Two types of craniopharyngiomas are recognized – adamantinomatous type (90%) and papillary type (10%). Adamantinomatous craniopharyngiomas are infiltrative, multilobulated solid-cystic suprasellar masses containing viscous "machinery oil" fluid rich in cholesterol crystals. Papillary craniopharyngiomas are solid encapsulated cauliflower-like masses that do not adhere to adjacent structures.[2] The former is seen in children while the latter occurs in adults. Clinical symptoms include visual defects, pituitary and hypothalamic disorders, and symptoms of intracranial hypertension. The larger the mass is, worse is the prognosis. Seizures are commonly seen with larger masses and are thought to be due to mass effects exerted over the suprasellar and/or temporal areas, eliciting an epileptic discharge. Regardless of size, these tumors rarely undergo malignant degeneration.

Giant craniopharyngiomas, described as a tumor larger than 5–6 cm on their largest axis, are more common in children and extremely rare in adults. They are usually adamantinomatous craniopharyngiomas and can range from 5 cm to 11 cm.[3] Giant craniopharyngiomas may extend into both anterior and middle cranial fossae. Posteroinferior extension between clivus and pons down to foramen magnum is seen in exceptionally large lesions.

Microscopically, adamantinomatous craniopharyngiomas are characterized by cystic portions and stratified squamous epithelium surrounding calcified wet keratin with prominent peripheral palisading. Papillary craniopharyngioma has solid sheets of mature squamous epithelium.

CT and MRI are the diagnostic tools. In CT, solid cystic portions can be differentiated, calcifications delineated, and enhancement observed. MRI shows the relationship with surrounding neurovascular anatomy. Craniopharyngiomas have variable signals on T1W1 and are usually hyperintense on T2/FLAIR. Nodular or rim enhancement is seen in 90% of cases. MR spectroscopy shows a large lipid lactate peak.

Gross total resection is the best treatment option;[2] however, recurrence is a problem, especially in larger and incompletely excised lesions. Hypothalamic injury is the major risk of resection surgery. Long-time survivors are more likely to experience reduced quality of life, mostly due to morbid hypothalamic obesity. Most cases of craniopharyngioma malignant degeneration occur in patients with multiple recurrences and prior radiotherapy, it is thus recommended that patients undergo regular MRI follow-up after surgical resection to ensure that no relapse or malignant degeneration occurs.

DIFFERENTIAL DIAGNOSES OF GIANT CRANIOPHARYNGIOMA

1. Pituitary adenoma especially hemorrhage in macroadenoma.
2. Eosinophilic granuloma and other infectious or inflammatory processes in this region.
3. Vascular malformations like aneurysms.
4. Congenital anomalies like Rathke's cleft cyst.
5. Other sellar and suprasellar masses.

CONCLUSION

Craniopharyngiomas are to be considered in the differentials for sellar and suprasellar region lesions causing mass effect. The provisional diagnosis is established by imaging but definitive diagnosis always requires histopathology.

TEACHING POINTS

1. Craniopharyngiomas can attend huge sizes, so they must be considered in differential diagnoses of sellar/suprasellar region lesions causing symptoms of mass effect.
2. Magnetic resonance imaging and computed tomography are to be used for establishing provisional diagnoses, but definitive diagnoses are always through histopathology.

MCQs

1. Craniopharyngiomas are thought to arise from:
   a. Notochord
   b. Diencephalon
   c. Craniopharyngeal duct
   d. Mesencephalon

Answer Key: c

2. Adamantinomatous craniopharyngioma is characterized histologically by all except:
   a. Wet keratin nodules
   b. Palisading squamous epithelium.
   c. Cystic portions
   d. Solid sheets of epithelial cells

Answer Key: d

3. The structure most likely to be injured while operating a large craniopharyngioma
   a. Optic bulb
   b. Pons
   c. Tectum
   d. Hypothalamus

Answer Key: d

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Ethical approval

Institutional Review Board approval is not required/waived-off.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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