Imaging of Monstrous Craniopharyngioma: A Pictorial Essay

Craniopharyngioma is usually a mixed solid and cystic suprasellar tumor, which usually occurs in children and adults in the fourth and fifth decades of life. The tumors are histologically benign and slow growing, but focal invasion and peritumoral fibrosis leads to high tumoral recurrence after surgery.

We had 11 patients admitted with different symptoms such as headache, nausea, vomiting and visual loss in whom extensive craniopharyngioma was proved after surgery. In some patients, primary imaging studies were reviewed. In others who presented with new onset symptoms several years after surgery, brain CT scan and MRI studies were performed. On brain CT scan of most cases, a mixed cystic and solid suprasellar mass with calcifications in both solid and cystic parts was detected. In this article, we present some cases of large craniopharyngioma with interesting extensions and invasions to adjacent brain structures.

Keywords: Craniopharyngioma, Computed Tomography, Magnetic Resonance Imaging

Introduction

Craniopharyngioma is a histologically benign slow-growing tumor (WHO grade I), which accounts for approximately 3% of all intracranial tumors. It has a bimodal age distribution with the first peak in children and the second in adults at age 40-60 years. It is not predominant in any sex or race. It originates from the embryonic precursor of adenohypophysis which is called Rathke's pouch. The adenohypophysis and infundibulum migrate from the pharynx to the sella turcica during fetal life. This fact shows that craniopharyngiomas may occur anywhere along this migration tract.

Craniopharyngioma has three histological types: adamantinomatous, papillary and mixed. The adamantinomatous type predominantly occurs in children. They are classically cystic tumors with a solid component. The fluid in the cyst is usually similar to motor oil. It contains high protein materials, cholesterol crystals and blood products. The solid part contains calcification, squamous or columnar epithelium, fibrosis and inflammation. Complete resection of the tumor is difficult because of this fibrosis and dense adhesion to adjacent structures. This issue leads to frequent recurrence of tumor in patients treated with surgery. The papillary type predominantly occurs in adults. It has only a solid component typically without calcification. This type is more encapsulated and has less recurrence after surgical resection.

Because of the slow-growing nature of tumors, the symptoms develop insidiously and the common interval between onset of symptoms and diagnosis is 1-2 years. The most frequent reported symptoms are headache, nausea, vomiting and visual disturbance. Growth failure and hypogonadism are also encountered frequently. Bitemporal hemianopia is the most frequent disturbance of the visual
field. Other presentations include seizures, precocious puberty, inappropriate secretion of ADH, hearing loss and epistaxis.\textsuperscript{10}

In patients with raised intracranial pressure or rapid visual function deterioration, we must focus on surgical treatment of hydrocephalus or tumor cyst decompression. The main management of tumor is surgery with an attempt to gross total resection of tumor, but in cases with extensive tumors in which total resection is impossible, limited resection with tumor debulking is recommended to reduce the tumor compression of optic pathways and to help CSF drainage. Limited surgical resections are followed by radiotherapy.\textsuperscript{2}

One of the most interesting issues in craniopharyngioma imaging is its extension and invasion to multiple brain structures, which is best appreciated on MRI. These tumors have different patterns of extension such as superior extension from the suprasellar region with compression of the third or lateral ventricles, lateral extension to parasellar areas, cavernous sinuses and temporal lobes, anterior extension to frontal lobes with superior displacement of A1 segment of the anterior cerebral artery and anterior communicating artery, posterior extension to perimesencephalic cisterns, brain stem, compression of the basilar trunk and involvement of cerebellum and superior cerebellar cistern and finally inferior extension to sella, sphenoid and ethmoidal sinuses and nasopharynx. Identifying these patterns of extension can help us in the surgical management of craniopharyngiomas to decide whether total or partial resection is recommended and to differentiate these tumors from other extensive skull-base tumoral lesions.

We encountered 11 patients with surgically confirmed craniopharyngioma. These are extensive tumors with unusual extension to their neighboring brain structures. Their brain CT scan and MR images are reviewed.

**Case Presentation**

**Patient 1**

The first patient is a six-year-old boy who presented with nausea, vomiting and ataxia 2 years after surgical treatment of craniopharyngioma. Brain CT scan showed a tumoral lesion in the suprasellar region with extension to the middle and posterior cranial fossa. The tumor is predominantly cystic with solid components. Eggshell calcification is seen in the cystic part. The solid part is also calcified in both popcorn and solid lump appearances. Right temporal craniotomy and an intracystic catheter are noted as well.

On brain MRI study, the mass is seen with both solid and cystic parts. The cystic part is hypointense on T1W and hyperintense on T2W images. After contrast injection, homogeneous enhancement is seen in the solid part and rim enhancement is also seen in the cystic part. The tumor extends from the suprasellar region to sella and then from the perimesencephalic cisterns to the posterior fossa, superior cerebellar cistern and cerebellum. It also extended to the right lateral ventricle and third ventricle and produces biventricular hydrocephalus due to compression of foramen of Monro. Pathologic examination revealed an adamantinomatous craniopharyngioma (Fig. 1).

**Patient 2**

The second patient is an 11-year-old boy who presented with visual loss 1.5 years ago. Brain CT scan revealed a tumoral lesion with both solid and cystic parts in the suprasellar and sellar regions. Calcification is seen in the lesion, which has eggshell configuration in the cystic part and lumpy appearance is also seen in the solid part. The tumor extends to the posterior cranial fossa. It also extends to the third ventricle from its superior margin. On MRI study, a T1 isointense and T2 hyperintense cystic lesion is seen in the suprasellar region with extension to sella turcica and involvement of the left parasellar region with left carotid artery encasement and left cavernous sinus invasion. The tumor extended to the posterior fossa and compressed the brain stem at its posterior margin. It deviated the pons, midbrain and fourth ventricle to the right. In addition, it compressed the basilar trunk at its posterior margin. At the superior margin, the tumor extended to the third ventricle and produced biventricular hydrocephalus. After contrast injection, thin rim enhancement was seen in the cystic component and nodular enhancement was encountered in the solid part (Fig. 2).
Patient 3
We encountered a six-year-old girl with headache, nausea and vomiting from 6 months ago. CT scan of the brain showed a mixed solid and cystic lesion with heavy calcification in the solid and cystic parts. The cystic part was seen with hypodense cystic contents. The tumor extended to the right lateral and third ventricles and produces biventricular hydrocephalus. On the MR imaging study, a mixed cystic solid tumor with low signal cystic contents on T1W was seen in the suprasellar region with extension to the right thalamus and lateral ventricles. Thick rim enhancement was seen in the wall of the cystic part after contrast injection. The tumor was adamantinomatous in pathologic examination (Fig. 3).

Patient 4
The fourth patient is a 12-year-old girl who presented with left eye blindness from 5 months ago. On brain CT scan performed after stereotaxis and placing an intracystic catheter, a heavily calcified suprasellar mass with extension to the sella was seen. Erosion and destruction of the anterior and posterior clinoid processes were noted. The mass extended to the anterior and posterior cranial fossa. A large cystic compo-
nent was seen in the left frontal region with mass effect and midline shift to the right. The MRI study was performed one year after surgery, the tumoral remnant was seen in the left thalamus and deep temporal lobe which has low signal intensity on T1W and high signal intensity on T2W images. Nodular enhancement was seen in the periphery of the cystic part after contrast injection. Pathologic examination

Fig. 2. An 11-year-old boy with visual loss from 1.5 years ago.
A. Axial brain CT scan, suprasellar lesion with calcification of the solid and cystic parts and extension to the posterior fossa.
B. Axial brain CT scan, tumoral extension to the third ventricle and compression of the foramen of Monro.
C. Axial T1W MRI, tumoral extension to the left parasellar area and left temporal lobe. Invasion to the posterior fossa and brainstem is noted as well.
D. Coronal T2W MRI, tumoral extension to the left parasellar area and encasement of the left carotid artery.

Fig. 3. A six-year-old girl with headache, nausea and vomiting from 6 months ago.
A. Axial brain CT scan, heavily calcified tumor with extension to the third ventricle and biventricular hydrocephalus induction.
B. Axial brain CT scan, cystic and calcified tumor with compression of the right lateral ventricle and midline shift to the left.
C. Axial T1W MRI, low signal cystic lesion with involvement of the right thalamus.
D. Sagittal T1W MRI with contrast, thick rim enhancement of the lesion.
revealed adamantinomatous craniopharyngioma (Fig. 4).

Patient 5
A 17-year-old female was admitted with headache and epistaxis from one year ago. Seven years ago she was operated because of craniopharyngioma. Brain CT scan showed a heavily calcified mixed solid and cystic mass in the suprasellar area with extension to the right cerebellopontine angle and third ventricle. Brain MRI revealed a mixed low and high signal intensity mass on T2W images in the suprasellar area with extension of cystic part to left temporal lobe and right cerebellopontine angle and interpeduncular cisterns. It is extended to anterior part of midbrain and cerebral peduncles. Low signal areas are seen in tumoral mass due to calcification. After contrast injection, the solid tumoral portion was enhanced densely and homogeneously. Comprehension of the basilar trunk was seen in the posterior aspect of the tumor as well as superior displacement of A1 segment of anterior cerebral artery (ACA) (Fig. 5).

Patient 6
A 15-year-old boy was admitted to hospital with headache and bitemporal hemianopia from 14 months ago. Brain CT scan study was performed 3 years after surgery. A heterogeneous calcified cystic and solid mass was seen in the sellar and suprasellar regions with extension to the anterior and posterior cranial fossae and also to sphenoidal and posterior ethmoidal sinuses and nasopharynx. Anterior and posterior clinoid processes, sellar floor and sphenoidal and ethmoidal bones are eroded. At the superior margin, the mass extended to the interpeduncular cistern. A hypodense area is seen in the right frontal lobe in favor of post-surgical encephalomalacic changes. A drainage device is also seen in the cystic part of the mass. Brain MRI revealed an isointense mass with gray matter on T1W images with nodular and rim enhancement and low signal areas in favor of calcification. The mass extended to the nasopharynx, posterior ethmoidal, left maxillary and sphenoidal sinuses from its inferior margin. It also extended to the posterior fossa and compresses the brain stem and the basilar trunk. From the superior margin, it extended to the interpeduncular cistern. Deviation of both internal carotid arteries and invasion of both cavernous sinuses are noted as well. A porencephalic area in the right frontal lobe and the drainage catheter are seen on MRI and CT scan. Pathologic examination revealed an adamantinomatous craniopharyngioma (Fig. 6).

Patient 7
A 5-year-old girl presented with visual loss 3 years after surgical treatment of craniopharyngioma. A calcified mixed solid and cystic mass was seen in the suprasellar area on brain CT scan. The tumor extended to the third ventricle and posterior fossa in the right cerebellopontine angle. Biventricular hydrocephalus was also noted. On T1W brain MRI study, right temporoparietal craniotomy is noted. A mixed solid-cystic mass is seen in the suprasellar area with extension to the third ventricle and compression of lateral ventricles with hydrocephalus induction in the right lateral ventricle. Low signal cystic materials are noted on T1W images. The solid part is seen with high signal intensity in some parts on T1W images probably due to calcification. A multi-cystic mass with low signal areas of calcification is seen on T2W images as well. Encasement of the proximal segment of the middle cerebral artery (MCA), ACA, the distal part of the internal carotid artery (ICA) and the basilar trunk is noted. Invasion to the brain stem from the posterior tumoral margin is noted as well (Fig. 7).

Patient 8
A six-year-old girl was admitted with headache, nausea and vomiting 3 years after surgical treatment of craniopharyngioma. Physical exam revealed paralysis of lateral gaze in the left eye. Brain CT scan showed a mixed solid and cystic mass with calcification in the suprasellar region with extension to the left temporal lobe and anterior cisterns of the brain stem. On MRI study, a T1W and T2W high signal intensity cystic mass is seen in the suprasellar region with extension to the left parasellar area and encasement of the left ICA and invasion to the left cavernous sinus and both temporal lobes. Compression and right deviation of the third ventricle are noted as well. Midline shift to the right is seen as well as superior displacement at A1 segment of the ACA.
Patient 9

A 23-year-old woman presented with headache, visual loss, nausea and vomiting 6 years after surgical treatment of craniopharyngioma. Brain MRI study was performed. A multi cystic mass with low signal intensity on T1W and high signal intensity on T2W images is seen in the sellar and suprasellar regions. Thin rim enhancement of the cystic part and nodular enhancement of the solid part are also noted after contrast injection. The mass extends to the right frontal lobe and compressed the optic chiasm. Compression of the third ventricular floor is also noted. Previous craniotomy scar is seen in the left parietal region (Fig. 9).

Patient 10

A six-year-old boy was admitted with headache,
nausea, vomiting and ataxia from 1.5 years ago. Brain MRI study was performed. An extensive solid and cystic mass is seen in the anterior, middle and posterior cranial fossae. The mass had multi-cystic components with high and low signal contents on T1W images. The high signal cystic part extends to the anterior fossa and compresses both frontal lobes and the optic chiasm. T1W imaging shows a low signal cystic part extending to the third ventricle with compression of the foramen of Monro and lateral ventricular hydrocephalus induction. Reticular and non homogeneous enhancement is seen in the solid parts. Right and left parasellar extension is noted as well. Posterior extension to the prepontine and interpeduncu-
lar cisterns with basilar trunk compression is presented (Fig. 10).

**Patient 11**

A six-year-old boy presented with testicular enlargement from 4 years ago. Brain CT scan was performed and a hypodense cystic suprasellar mass with superior extension to the third ventricle is noted. Thin eggshell calcification is also seen in the walls of the cystic part. Biventricular hydrocephalus is detected. On MRI study, a T1W low signal cystic mass with iso signal solid parts is seen in the suprasellar region with superior extension to the third ventricle and inferior extension to sella turcica. Encasement of the left ICA bifurcation and superior displacement at M1 segment of the MCA is noted. Posterior extension to the midbrain and basilar trunk compression is seen as well. Biventricular hydrocephalus is evident on MRI and CT scan (Fig. 11).
Discussion

Craniopharyngioma is the most common pediatric intracranial tumor of non-glial origin. This tumor should be considered in the differential diagnosis of suprasellar, parasellar, prepontine and posterior fossa tumors. Rathke cleft cyst, suprasellar, arachnoid cyst, hypothalamic or chiasmatic astrocytoma, and even dermoid or epidermoid cysts may be mimicked by some kinds of extended craniopharyngiomas at imaging. Being familiar with unusual and interesting imaging presentations of these common pediatric tumors could lead radiologists to a better diagnosis.

Craniopharyngiomas are partly cystic, benign, slow growing, usually suprasellar tumors which account...
for 5-13% of all intracranial tumors. They derive from Rathke’s pouch, the embryonic infolding of endoderm that extends superiorly from the stomodeum and gives rise in the early fetal development to the adenohypophysis; with manifestation during childhood and fourth or fifth decades of adult life. Frequent symptoms of manifestation are visual symptoms or endocrine disturbances such as polyuria and polydipsia due to diabetes insipidus. The tumor has three histologic types: adamantinomatous (pediatric type), papillary (adult type) and mixed. The adamantinomatous type is classically a cystic tumor with solid components. The papillary type is predominantly solid, without calcification. After surgery and radiation, the 5-year survival rate is more than 80% in children. Permanent endocrinologic deficits are seen in many patients after treatment but visual disturbances are usually reversible. The recurrence rate of the tumor depends on the size of the tumor, the recurrence rate is approximately 20% for tumors smaller than 3 cm and 83% for tumors larger than 5 cm.

CT and MRI are complementary studies for detection of extent and characteristics of the tumor. On CT scan, the pediatric type is presented as a predominantly suprasellar and calcified cystic mass with solid components in 90% of cases. Extension to sella turcica is seen frequently. The adult type tumors are usually solid and isodense. Calcifications and intraventricular craniopharyngiomas are rare.

In the papillary subtype, after contrast injection, enhancement is seen in solid components and the wall of cystic parts in 90% of cases. Anterior and superior extension of the tumor leads to anterior dislocation of the optic chiasm and superior dislocation of the A1 segment of the anterior cerebral artery. On CT scan, the usual extension of the cystic part is to the anterior and lateral but the solid part extends to the posterior and lateral.

MRI is preferred for better determination of the tumor extent. The pediatric type appears as a predominantly cystic mass with solid components but the papillary type is predominantly solid. Calcifications are better seen in GRE images and may show susceptibility effects. Signal intensity of the cystic part is variable on T1W images. Blood products, high protein concentration or cholesterol crystals in the cyst leads to high signal characteristics of the cystic part on T1W images. Cysts are hyperintense on T2W and FLAIR images. Solid components appear isointense on T1W and heterogeneously iso to hypointense on T2W images.

A prominent lipid spectrum is seen in MR spectroscopy around 1PPM because of the cystic contents. Also variable signal intensities are seen in diffusion weighted images because of variable cystic contents. After contrast injection, patterns of enhancement in solid and cystic tumoral parts are similar to CT scan.

The location of the tumor is predominantly suprasellar in 75%, supra and infrasellar in 12%, and purely intrasellar in 4% of cases. Encasement or displacement of the vessels forming the circle of Willis is noted. Lateral displacement of ICAs and superior dislocation of ACA are also seen. Posterior dislocation and compression of the basilar trunk is noted as well.

We can categorize the tumoral extension to three specific subtypes. In type A, the tumors are entirely within the sella and vascular displacement is not seen. In type B, the tumor is extended anteriorly between the optic nerves and pushes the optic chiasm posteriorly. Elevation of the anterior communicating artery and the A1 segment of the anterior cerebral artery are noted as well. In type C, elevation of the A1 segment of the anterior cerebral artery and anterior communicating artery, with posterior compression of the basilar trunk is noted. Anterior dislocation of the optic chiasm with third ventricular compression and hydrocephalus are seen as well.

References