An Atypical Rare Case of Extracranial Craniopharyngioma

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Abstract We present a case of a purely extracranial, infrasellar craniopharyngioma that initially presented as a mass in nose and nasopharynx. Craniopharyngiomas are usually located within the sella. Purely infrasellar craniopharyngiomas have only rarely been reported in the literature. A 55-year-old man presented with 8-month history of progressive headache and epistaxis. Rhinoscopy revealed polypoidal mass in both the nasal cavities and nasopharynx. Pre-operative biopsy suggested Craniopharyngioma. A battery of tests necessary for the diagnosis of Craniopharyngioma was done which excluded other possibilities. Surgical resection was done and histopathology thereafter was confirmatory of Craniopharyngioma. Adjuvant radiotherapy was given to the patient. The patient is doing well. The Rathke's pouch arises from the roof of the primitive mouth and grows toward the brain at the fourth

week of gestation. Normally, it loses its attachment with the stomadeum completely by the eighth week of gestation. The craniopharyngeal canal (CPC) extends from the floor of the sella to the vomer and may rarely give rise to ectopic craniopharyngiomas. This case shows that such ectopic tumors may arise anywhere along the CPC. We are documenting this case as an atypical rare case of craniopharyngioma probably originating from tooth primordia.

Keywords Craniopharyngioma · CPC · Rathke's pouch · Sella · Parasella · Nasopharynx · Nasal cavity · Extracranial · Sphenoid · Primordia

Introduction

Craniopharyngioma is a benign, slow growing, extra-axial tumor of embryonic/metaplastic aetiology that almost always occur in a suprasellar location, making complete resection difficult and often necessitating radiotherapy. It may present with headache, endocrinal or visual disturbances. A man presenting with epistaxis and headache is being reported here.

Case Report

A 55-year-old male patient presented to the outpatient department of Otorhinolaryngology, Medical College and Hospital, Kolkata in February 2011 with history of insidious onset, gradually progressing epistaxis for 8 months and headache for 6 months and recent onset nasal obstruction and hyposmia for 2 months (Fig. 1). Anterior and posterior rhinoscopy revealed a pale, polypoidal mass involving both nasal cavities and the nasopharynx. There was no external

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Fig. 1 Preoperative photograph of the patient

nasal deformity or orbital involvement or palatoalveolar deformity.

Preoperative punch biopsy shows complex pattern of squamous epithelial growth showing dense and loose areas with peripheral palisading, micro cyst formation and keratinization suggestive of craniopharyngioma, but could not pinpoint subtypes (d/d ameloblastoma).

Other clinical examination reveal vision, fundus and field to be normal and investigations based on the histopathology and endocrinal check up showed no irregularity.

Radiologic studies with contrast enhanced CT scan of nose and para-nasal sinuses (PNS) (axial and coronal views) revealed a heterogeneous mass occupying the nasal cavities and nasopharynx with intact lamina papyracea and without any breach of skull base nor any involvement of sinus cavities except for sphenoid sinus (Fig. 2).

Surgery was envisioned for resection of the tumor from nose, nasopharynx and sphenoid with mid-facial degloving approach. The mass—smooth, lobulated and capsulated—was completely removed macroscopically and sent for histopathology.

Post-operative histopathological examination revealed anastomosing epithelial islands of squamous epithelium in cords, lobule and irregular trabeculae bordered by palisade columnar epithelium (picket-fence like epithelium) with islands of densely packed squamous cells merged with loosely cohesive aggregates of squamous cells known as stellate reticulum. Nodules of keratin, granulomatous inflammation associated with cholesterol cleft and debris, in a background of mixed inflammatory infiltrate are also seen in these sections suggestive of adamantinomatous craniopharyngioma (Fig. 3). Nature of the mass was confirmed with immunohistochemical (IHC) staining and it showed cytokeratin positivity confirming the diagnosis (Fig. 4).

Following surgical removal of tumor, patient had undergone External beam radiotherapy (55 Gy in 27

fractions over 1 month). The patient is currently under regular follow up and doing well.

Discussion

Craniopharyngioma is a benign, partly cystic epithelial tumor of the suprasellar region (most common) (with a minor intrasellar component) presumably derived from Rathke's pouch/duct epithelium (embryogenic/metaplastic theory) occurring histopathologically as adamantinomatous and papillary varieties. Adamantinomatous variety consists of reticular epithelial cells which have appearances reminiscent of the enamel pulp of developing teeth. Calcification is usually present (in ~90 %) [1]. The papillary subtype is formed of masses of metaplastic squamous cells [1]. Adamantinomatous (ordinary) craniopharyngioma occurs in children and tends to be less solid than papillary craniopharyngioma. Papillary craniopharyngioma occurs in adults and is a more solid tumor. It is a slow growing extra-axial tumor accounting for 1.2–4.6 % of all intracranial tumors. It constitutes the most common non-neuroepithelial intracerebral neoplasm in children (5–10 % of intracranial tumors). It has a bimodal age distribution in the 5–15 years and 45–60 years group [2]. There appears to be a similar incidence in both males and females. Craniopharyngioma with no intracranial extension is a very rare but benign tumor and unusual site involving nasopharynx and sphenoid have been cited in only 14 articles (among which only 3 nearly resemble present case) by Pubmed search [3–5]. The present case is unique in that the 55-year-old male patient had a craniopharyngioma mass limited only to sphenoid [6], nasopharynx and nasal cavity without sellar/suprasellar involvement.

Clinical presentation depends on Prechiasmal (optic atrophy: 62–84 % patients and more in adults), Chiasmal (headache and endocrinopathy: 52–87 % and more in children) or Retrochiasmal (raised intracranial pressure and hydrocephalus) involvement of tumor [7, 8]. No endocrinal deficits or perimetric field defects or cranial fossa invasion were found in our case but presented with headache, nasal obstruction and epistaxis in an adult.

Neuroimaging with CT scan of an adamantinomatous lesion generally depicts the sellar configuration with contrast enhancement of the solid portions and the capsule but CT scan of this patient depicts a heterogeneous mass without sellar involvement and confined to the aforementioned spaces.

Typically like a lobulated solid mass with a spongy feel due to cystic component with machinery oil like cholesterol rich greenish brown liquid, our patient had an adamantinomatous mass without extension into adjacent spaces, which is unusual. Histopathologically the

Fig. 2 CT scan of nose and PNS (axial and coronal views) of the patient

Fig. 3 HP microphotograph showing adamantinomatous craniopharyngioma with focal keratinization

Fig. 4 IHC reactivity for cytokeratin

adamantinomatous mass of our patient had the usual cord like squamous epithelial disposition, lobules and irregular trabeculae with border of palisade columnar epithelium, with loose and compact islands of stellate reticulum and densely packed cells, with nodules of wet keratin [9]. IHC study showed cytokeratin positivity, pointing towards adamantinomatous craniopharyngioma and decisively proving the diagnosis as against other differential diagnosis of

granulomatous inflammation like xanthogranuloma, tumors like epidermoid/dermoid cyst, infectious/inflammatory processes, vascular malformations and other congenital defects like a Rathke's cleft cyst [9].

Endoscopic endonasal approach provides an excellent route for the resection of these tumors. The treatment protocol for extensive lesion is surgical resection followed by Stereotactic radiotherapy. If hydrocephalus is present, a

shunt may be placed during surgery. The shunt will help remove excess cerebrospinal fluid from the brain and ease the pressure. In children younger than 3, radiation may be delayed by the use of surgery or hormone therapies. The most important factor associated with craniopharyngioma recurrence is the extent of surgical resection, with lesions greater than 5 cm in diameter carrying a markedly worse prognosis as also the incompleteness of surgical resection and especially so with an adamantinomatous type of craniopharyngioma [8]. Overall 60–93 % of patients have a 10-year recurrence-free survival and 64–96 % have a 10-year survival [10, 11]. Our patient was managed with total macroscopic surgical resection of gross pathology through a mid-facial degloving approach followed by irradiation that was started after 3 weeks of operation. The patient is currently under follow up and turning up every 3 months for the first 2 years and no recurrence is noted yet either clinically or on CT scan. Our patient is more fortunate as histological evidence for brain invasion, which is more frequently documented in the adamantinomatous variety, is absent.

Till date many studies have documented suprasellar or sellar craniopharyngioma as per PubMed search, but only four studies have documented involvement of nasopharynx in extracranial craniopharyngioma in review of literature [3–6]. The present case report is unique in the site of occurrence of craniopharyngioma in purely extracranial location of nasal cavity, nasopharynx and sphenoid, with no endocrinal deficits or perimetric field deficit or cranial fossa invasion and the clinical presentation is also unusual i.e. epistaxis, headache and nasal obstruction. The relative dearth of cases as well as the exclusivity has encouraged us to report this case.

Conclusion

Craniopharyngioma is an extra-axial, benign, slow-growing squamous epithelial tumor from Rathke's duct/pouch involving sellar and suprasellar in almost all cases excepting our case which originated probably from tooth primordia as per its unique location confirmed after CT

scan, surgery and histopathological examination [1, 9]. The present case is marked out by its extracranial involvement only of nasal cavity, nasopharynx and sphenoid without any intracranial continuity and presentation with primary nasal symptoms and lack of intracranial symptoms thus providing us a diagnostic dilemma.

The present case is under continuous surveillance for about 2 years till date without any recurrence. We are documenting this case as an atypical and rare case of purely extracranial, infrasellar craniopharyngioma.

Conflict of interest None.

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