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A RARE CASE OF INTRANASAL CRANIOPHARYNGIOMA

Abstract:
Craniopharyngioma is a benign, slow growing tumour with infrasellar location being very rare. Two histologic variants have been described in literature - papillary and adamantinomatous type. We present here a rare case of infrasellar adamantinomatous type of intranasal craniopharyngioma.

Key words: Craniopharyngioma, Infrasellar.

INTRODUCTION:

Craniopharyngioma is a benign tumour which constitutes 5 per cent of all intracranial neoplasms.[1] It is a slow growing tumour which arises from Rathke's pouch epithelium, and confined mainly to the sellar and suprasellar regions. Two variants have been documented: an adamantinomatous type, which commonly presents in childhood and has worse prognosis, and a papillary type, which occurs in adulthood and has better prognosis.[2] Infrasellar craniopharyngioma is very rare. In this study we have reported a case of infrasellar craniopharyngioma with no sellar involvement which is extremely rare.

CASE REPORT:

A 45 years female presented with right sided nasal obstruction and right nasal bleeding for two and half years. No other symptoms suggestive of pituitary dysfunction was present. Anterior rhinoscopy revealed a single, pinkish polypoidal mass in the right nasal cavity, arising medial to the middle turbinate. Contrast enhanced CT scan revealed a moderately enhancing mass arising from lateral wall of right nasal cavity in the region of middle turbinate, with areas of hyperostosis within the lesion. The mass caused deviation of nasal septum to left side. Punch biopsy was taken under local anaesthesia and histopathology revealed squamous epithelium arranged in lobules and irregular trabeculae, bordered by palisading columnar epithelium, keratinized areas and cystic cavities were noted within the lobule characteristic of craniopharyngioma. Patient underwent endoscopic excision of the mass along with endoscopic septoplasty under general anaesthesia. Per-operatively, 4x3cm yellowish firm mass along with areas of calcification was removed from right posterior ethmoids, which was going to the nasopharynx. Nasal pack was removed on the second post-operative day. Post-operative histopathology revealed the adamantinomatous type of craniopharyngioma.

DISCUSSION:

Craniopharyngioma is a tumour with benign histology, yet showing locally aggressive behavior. It has a tendency to recur after complete surgical excision. Ninety percent of these lesions occur in the sellar or suprasellar region, but rare cases of infrasellar craniopharyngioma have been documented(5%).[3] Erdheim postulated that craniopharyngioma develops from squamous remnants of an incompletely involuted craniopharyngeal duct representing the route taken by Rathke's pouch from the pharynx to the floor of the third ventricle. Lesions can arise at any point along this embryological path, explaining their occurrence in extracranial sites.[4] They can extend to the anterior, middle, or posterior cranial fossa. Rarely, craniopharyngioma arise primarily in unusual locations, such as the nasopharynx, sphenoid bone, third ventricle, pineal gland, sylvian fissure, and cerebellopontine angle. It has a bimodal age distribution in the 5–15 years and 45–60 years group. There appears to be a similar incidence in both males and females.[5] The clinical presentation depends on location and size of the mass. Patients with suprasellar lesions commonly present with visual or endocrine disorders. In contrast, infrasellar craniopharyngioma presents with symptoms of epistaxis and nasal obstruction, as our case.

Two histologic variants are found. Adamantinomatous variety consists of reticular epithelial cells which have

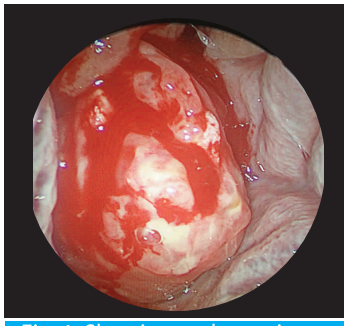

Fig. 1: Showing endoscopic appearence of mass

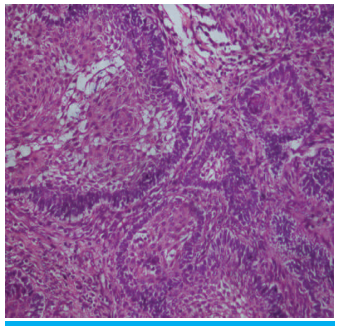

Fig. 2: Showing HPE

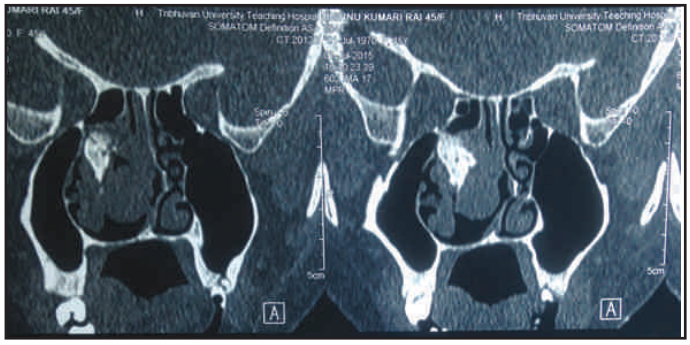

Fig. 3: Showing CECT of mass

appearances reminiscent of the enamel pulp of developing teeth. Calcification is present in nearly 90%. The papillary subtype is formed of masses of metaplastic squamous cells.6 Adamantinomatous craniopharyngioma occurs in children and tends to be less solid than papillary craniopharyngioma. Papillary craniopharyngioma occurs in adults and is a more solid tumor.

Computed tomography in craniopharyngioma usually reveals heterogeneous density of the tumor with its solid and cystic components, calcification and irregular enhancement. Calcifications are more common in children than in adults. MRI, with its multiplanar capability, is essential for defining the local anatomy and is the most important imaging modality used to plan the surgical approach.

Treatment is surgery with radiotherapy for incomplete resection. Surgical approach depends on the size and location of tumour. Local recurrence significantly depends on histology: papillary has a much lower recurrence rate than adamantinomatous variety. Gamma knife sugery is also being used as a treatment option. Transnasal endoscopic approach provides an excellent route for tumour limited to nasal cavity, nasopharynx, ethmoids or the sphenoid as in our case.

CONCLUSION:

Infrasellar craniopharyngioma, though rare, should be kept in differential diagnosis of a unilateral nasal polyp. Although benign, it has aggressive nature, therefore complete excision is essential. Adjuvant radiotherapy is recommended in incomplete excision.

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