



Case Report:

Craniopharyngioma - Transnasal Endoscopic Approach

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Abstract:

Craniopharyngiomas are slow growing tumours arising from remnants of the craniopharyngeal duct and occupy the sellar region. The patients may remain asymptomatic for long duration or present with headache or visual disturbances. Surgery is the mainstay of the treatment. Traditionally these tumours have been removed by neurosurgeons through the cranial approach but the advent of nasal endoscopes has opened new avenues for ENT surgeons to treat such patients. We hereby present a case of craniopharyngioma who was successfully treated by Trans-nasal Hypophysectomy.

Key Words: Craniopharyngioma; Trans-nasal Hypophysectomy

Introduction:

Craniopharyngioma is a slow-growing, extra-axial, epithelial-squamous, calcified cystic tumor arising from remnants of the craniopharyngeal duct and/or Rathke cleft and occupying the sellar region.(1) Craniopharyngiomas are dysontogenic tumors with benign histology and malignant behavior, as they have a tendency to invade surrounding structures and recur after total resection.(2,3) Craniopharyngioma usually presents as a single large cyst or multiple cysts filled with a turbid, proteinaceous material of brownish-yellow color that shines because of a high content of floating cholesterol crystals. Because of its appearance, it has been compared to machinery oil. It most frequently arises in the pituitary stalk and projects into the hypothalamus. Incidence of craniopharyngiomas is 0.5-2 per 100,000 per year.(4) Craniopharyngioma accounts for 1-3% of intracranial tumors and 13% of suprasellar tumors.(5) Symptoms frequently develop insidiously and mostly become obvious only after the tumor attains a diameter of about 3 cm. We are presenting one case that had mainly presented with visual disturbances and was removed by Trans-nasal endoscopic Hypophysectomy.

Craniopharyngiomas are rare intracranial tumours (0.5%) and it is even more rare to find them in a 20 year old, as they are more common in 1st decade and then in 5th decade. Secondly we were able to find and document the typical features of craniopharyngioma in this case and the patient also recovered almost completely.

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Case Report:

A 20 year old male presented with complaints of progressive diminution of vision in the left eye for 3 months and occasional headaches. Ophthalmic examination revealed vision of 1/60 in the left eye and 6/24 in the right eye, with optic atrophy of left eye on fundus examination. Multiplanar MRI imaging of cranium was done. T1 and T2 weighted images were obtained in axial, coronal and sagittal planes. Post Gadolinium enhanced T1 weighted images were subsequently acquired in all 3 planes. MRI revealed well defined lobulated lesion of 2.9 cm x 2.6 cm in sellar and suprasellar region. This lesion was hypointense on T2 weighted images with an iso to hypo intense component within suggestive of mural nodule, whereas it was hyper intense on T1 scans. Post contrast scan showed well defined peripheral ring of enhancement. These findings were consistent with craniopharyngioma of sellar and suprasellar pituitary. MRI Skull revealed a soft cystic mass in infra sellar pituitary. (Fig. 1) Hormonal assay of the patient was normal. The mass was removed by Trans-nasal endoscopic approach in association with neurosurgeon. Oily coloured fluid suggestive of craniopharyngioma was aspirated from the cystic mass before incision over meninges. Vision of the right eye recovered completely 3 days postoperatively. A post operative CT scan done after 3 weeks following surgery, showed complete removal of the mass (Fig. 2). HPE of the excised mass showed features suggestive of craniopharyngioma.

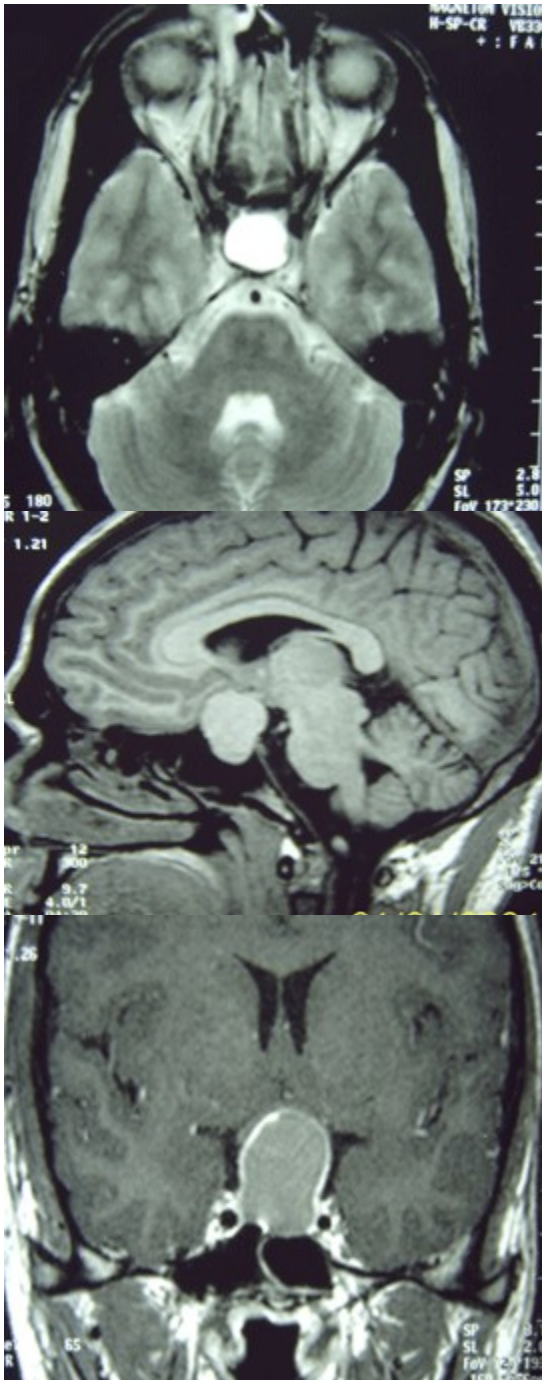


Fig 1: MRI Scan showing Craniopharyngioma



Fig 2: Post operative (after 3 weeks) CT Scan showing tumor clearance

Discussion:

Craniopharyngioma is a slow-growing, extra-axial, epithelial-squamous, calcified cystic tumor arising from remnants of the craniopharyngeal duct and/or Rathke cleft and occupying the sellar region. Two main hypotheses explain the origin of craniopharyngioma—embryogenetic and metaplastic. Embryogenetic theory relates to development of the adenohypophysis and transformation of the remnant ectoblastic cells of the craniopharyngeal duct and the involuted Rathke pouch, while metaplastic theory relates to the residual squamous epithelium (derived from stomodeum and normally part of the adenohypophysis), which may undergo metaplasia. (6) Another theory which explains the craniopharyngioma spectrum, attributing the adamantinous type to embryonic remnants and the adult type to metaplastic foci derived from mature cells of the anterior hypophysis is the dual theory. Craniopharyngiomas are dysontogenic tumors with benign histology and malignant behavior, as they have a tendency to invade surrounding structures and recur after what was thought to be total resection. Craniopharyngioma usually presents as a single large cyst or multiple cysts filled with a turbid, proteinaceous material of brownish-yellow color that shines because of a high content of floating cholesterol crystals.(2,5) Because of its appearance, it has been compared to machinery oil. It most frequently arises in the pituitary stalk and projects into the hypothalamus. Time interval between onset of symptoms and diagnosis ranges from 1-2 years. The most common presenting symptoms are headache (55-86%), endocrine dysfunction (66-90%), and visual disturbances (37-68%). The diagnostic evaluation for craniopharyngioma includes precontrast and postcontrast CT scans and MRI, Magnetic Resonance Angiography (MRA), complete endocrinologic and neuro-ophthalmologic evaluation with formal visual field documentation, as well as neuropsychological assessment.(3,4) Imaging studies strongly suggest the diagnosis. The radiologic hallmark of a craniopharyngioma is the appearance of a (supra)sellar calcified cyst. About 80-87% of craniopharyngiomas are calcified and 70-75% is cystic. Calcifications are more common in children (90%) than in adults (50%).CT scan is the most sensitive method to demonstrate calcifications as high-density areas and has replaced the plain radiograph. It is useful in defining both calcified and cystic parts. Cyst content usually has the same density as CSF; contrast administration better defines the enhancing cyst capsule.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.MRA is used for visualizing the major cerebral vessels and their relation to the tumor; it has largely replaced

the 4-vessel angiogram. The histologic spectrum of craniopharyngioma includes 3 main types—adamantinomas, papillary, and mixed. Adamantinomas consist of reticular epithelial masses, resembling the enamel pulp of developing teeth. This is seen predominantly in children. A distinctive feature is a palisading basal layer of small cells, which encloses a loose stellate reticular zone, as well as areas of compactly arranged squamous cells.⁽⁵⁾ They contain nodules of keratin ("wet" keratin), which are the hallmarks of this tumor subtype. Squamous papillary type is composed of islands of squamous metaplasia, embedded in a connective tissue stroma, with infrequent cystic degeneration and calcification. Essentially, 2 main management options are available for craniopharyngioma—(1) attempt at gross total resection or (2) planned limited surgery followed by radiotherapy.^(3,5) Gross total surgical removal is the treatment of choice. Recurrence rates can be as high as 20%. Some authors propose a plan of limited surgery, with postoperative radiotherapy as the management paradigm of choice for craniopharyngioma. Goals of this approach are (1) pathologic confirmation of the tumor and (2) surgical decompression of the optic chiasma. Surgery is followed by external beam radiation, at a dose of 5400-5500 cGy delivered at 180 cGy/fraction. Trans-nasal endoscopic procedures have significantly reduced the morbidity associated with this surgery. ENT surgeons can provide access to the pituitary without any scar and the morbidity associated with the trans cranial approach. Gamma Knife surgery has recently been used to treat craniopharyngiomas. These procedures don't actually involve the use of a "knife" or scalpel. In fact, no incisions are made at all. The skull never has to be opened up. Gamma Knife also differs from conventional radiation therapy. Because patients are injected with such low doses of radiation, they don't experience the side effects associated with traditional radiation therapy. In fact, several "shots" of therapy can be given during the same session, and treatment sessions can be repeated every few weeks if necessary.

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