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CRANIOPHARYNGIOMA IN 22 YEARS OLD MALE – A RARE CASE REPORT

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ABSTRACT

Craniopharyngiomas are rare intracranial tumors (0.5%) and it is rarer to find them in a 22 years old male patient. In this case, patient was presented with complain of diminished vision, headache which are the most common symptoms. Laboratory and imagistic investigations are helpful for the diagnosis, but histopathology is the gold standard diagnostic tool.

INTRODUCTION

Craniopharyngioma is a rare tumor [1]. It 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 [2]. Incidence of craniopharyngiomas is 0.5-2 per 100,000 per year [3]. It accounts for 1-3% of intracranial tumors and 13% of suprasellar tumors [4]. They can extend to the anterior (2-5% of cases), middle (2%), or posterior (1-4%) cranial fossa, and infrasellar extension is found in about 5% of cases [5]. There is a bimodal peak age distribution with the initial peak being in childhood and the second peak in the fourth and fifth decade of life [6]. The patients may remain asymptomatic for long duration or present with headache or visual disturbances. The microscopic appearance of most craniopharyngiomas shows an external layer of high columnar epithelium, a variable amount of polygonal cell, and acentral network of epithelial cells. Three clinicopathologic varieties have been delineated microscopically: the papillary squamous type, which is seen almost exclusively in adults and carries a better prognosis; and the classical adamantinomatous variant, which develops mainly in children and has a worse overall outcome [7]. The third variety is mixed type. As craniopharyngiomas are rare intracranial tumors (0.5%) and it is even rarer to find them in a 22 years old male here we are presenting a case where we are able to find and document the typical features of craniopharyngioma.

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Case report

A 22 years old male patient was apparently asymptomatic before 9 months then he started to have diminished vision that progressively increased. He had also complaint of headache and dizziness associated with vomiting for 8 months. Both general and neural examinations were normal. Hormonal assay of the patient showed raised cortisol level (>50ug/dl). X-ray skull finding showed that there was thinning of posterior wall of sellaturcica with possible erosion of sphenoid bone. CT scan of the sellar region revealed a cystic and focally calcified suprasellar mass. (Photo–1). The preoperative diagnosis was craniopharyngioma. The patient underwent craniotomy andoily brown colored fluid was aspirated

from the cystic mass before incision over meninges. The fluid was sent for cytological examination and microscopic examination revealed that smears were containing cholesterol crystals along with chronic inflammatory infiltrate. (Photo – 2). The diagnosis was in favor of craniopharyngioma. A post operative MRI done after 2 weeks following surgery showed complete removal of the mass. (Photo - 3). Histopathological examination of the excised mass showedepithelium resembles long bone tumors containing three distinct layers: Basal layer of small basophilic cells, Internal layer of stellate cells in loose connective matrix, known as "stellate reticulum" and the upper most layer of palisading keratinized squamous cells that shed into the cyst cavity forming "wet keratin". The overall features suggestive of Adamantinomas craniopharyngioma. (Photo - 4, Photo – 5)

Photo – 1. Preoperative CT scan shows a heterogenous mass arising from sellaturcica

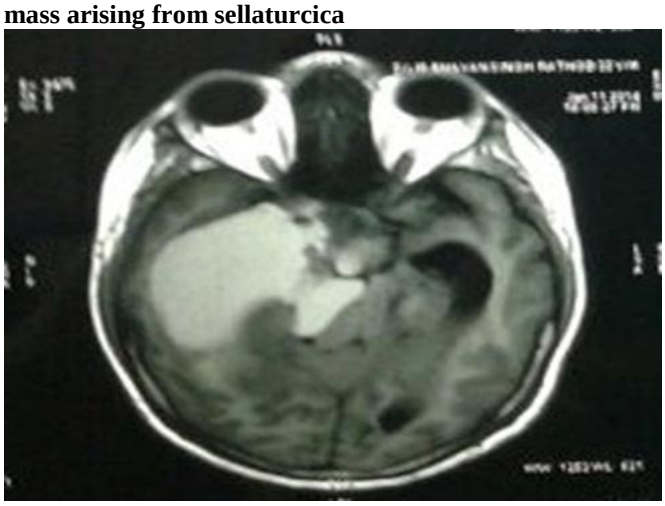

Photo – 2. Post operative MRI shows tumor clearance

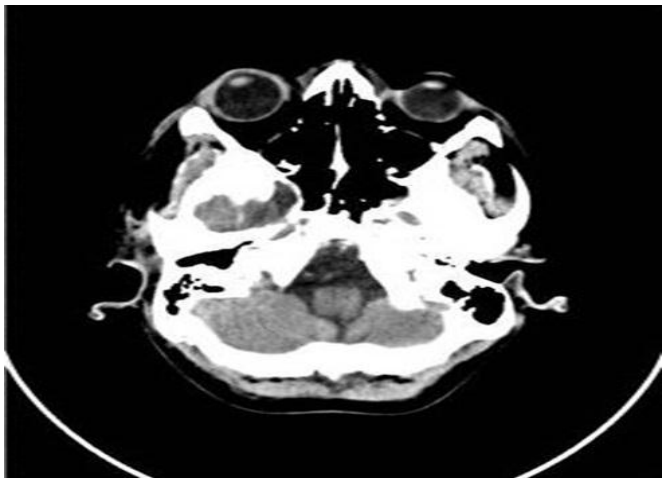

Photo – 3. Smears shows cholesterol crystals in the cystic fluid.

Photo – 4. Upper layer of palisading keratinized squamous cells that shed into the cyst cavity forming "wet keratin. [H & E, 20 X]

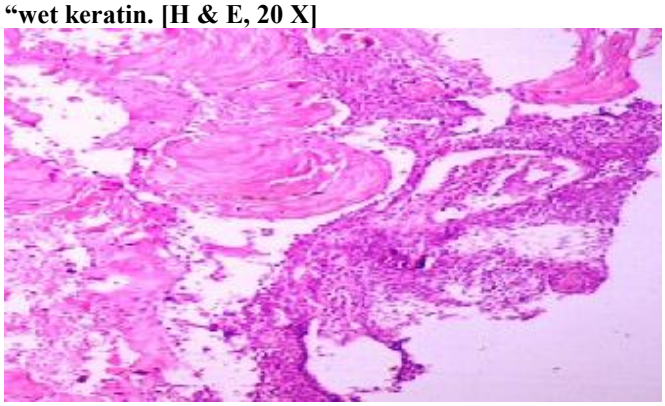

Photo – 5. Anastomosing islands of palisading epithelial cells. [H & E, 20 X]

DISCUSSION

Craniopharyngiomas are benign but aggressive epithelial neoplasms. They are most commonly located extra axially in the sellar or suprasellar area in 90% of cases [8]. Although this benign tumor exhibits slow growth, it typically shows adherence to surrounding structures, including the floor of the third ventricle [9], adjacent pituitary stalk, and vessels. Grossly, 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. 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 [10]. 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.

Regarding origin of the craniopharyngioma there are two main hypotheses - embryogenetic and metaplastic. Embryogenetic theory relates to development of the adenohypophysis and transformation of the remnant ectoblastic cells of the craniopharyngeal duct and the involutedrathke pouch, while metaplastic theory relates to the residual squamous epithelium (derived from stomodeum and normally part of the adenohypophysis), which may undergo metaplasia [11]. 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.

Symptoms and clinical findings are related to the craniopharyngioma's localization and mass effect with compression of the surrounding structures. The main symptoms are defects in the visual fields (bitemporal or homonimushemianopsia), varying signs of pituitary insufficiency (diabetes insipidus, amenorrhoea, diminished libido and cachexia), and the symptoms of increased intracranial pressure (headache, vertigo and cranial nerve deficit) that occur relatively late in the course [12]. The signs of pituitary dysfunction appear early in the cases in which the tumor expands within the sella.

Our patient presented with defects in the visual fields, headache and vomiting since last 8-9 months. Time interval between onset of symptoms and diagnosis ranges from 1-2 years. Skull X-ray study, tomograms, and CT scan were the most common radiographic techniques employed in evaluation of the diesease. Magnetic resonance image clearly showed the tumor extension, cystic portions, mixed intensity signal, inhomogeneous or heterogeneous enhancement [13-22].

Essentially, 2 main management options are available for Craniopharyngioma: Attempt at gross total resection or planned limited surgery followed by radiotherapy [23]. Gross total surgical removal is the treatment of choice. Recurrence rates can be as high as 20%. However, extent of surgical resection is certainly important in the long-term recurrence rates of these tumors. After total removal, the chance of tumor recurrence after 5 years is 13%. However, that rate increases to 58% with subtotal resection [24]. This is because of hidden "fingers" of tumor which often invade the hypothalamic region. Such recurrence usually occurs in the vicinity of the primary tumor site [25].

SUMMARY

The diagnosis of craniopharyngioma requires imaging studies in the form of MRI, computed tomography (CT), serum cortisol level, cytological examination of cystic fluid and the most useful diagnostic test microscopic examination of the tumor because histopathology is the gold standard for the final diagnosis.

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