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NASOPHARYNGEAL ADMANTINOMATOUS CRANIOPHARYNGIOMA IN 14 YEARS OLD BOY- A CASE REPORT STUDY

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ABSTRACT

Craniopharyngiomas are rare benign epithelial tumours arising from the pituitary stalk or gland. The sellar and parasellar region is the most commonly involved sites but at times tumors extend below the sellar floor involving the sphenoid sinus, invade the pharynx and reach upto the nasal cavities. Here is a case of 14 years old boy presenting with nasal cavity mass leading to bilateral obstruction and he was operated to excise the mass lesion. Grossly a grayish white cystic mass lesion was obtained after excision biopsy. On cut section, cyst contained a greyish brown thick liquid like material, with the microscopic findings of densely packed nodules of well differentiated epithelium along with stellate reticulum and wet keratin consistent with the diagnosis of craniopharyngioma. Clinical features along with imaging characteristics (except site of the lesion) and histopathological findings were all consistent with primary admantinomatous craniopharyngioma,

KEYWORDS: Craniopharyngioma, Adamantinomatous, Nasopharyngeal.

INTRODUCTION

Craniopharyngiomas are rare benign epithelial tumors which arise from the pituitary stalk or gland. , It accounts for 1–5% of all primary intracranial tumors.[i] These slow-growing non-glial tumours shows bimodal peak by affecting at one hand children and adolescents and, on the other hand ,adults over 50 years of age.[ii] The sellar and parasellar region of the central nervous system is the most commonly involved sites but at times tumours may extend below the sellar floor involving the sphenoid sinus ; invading the pharynx and reaching upto the nasal cavities.The infrasellar craniopharyngioma may then originate anywhere along the tract of the obliterated craniopharyngeal duct, which would include the sphenoid bone, vomer, and nasopharynx. , Here we are presenting a case of nasal cavity craniopharyngioma.

CASE REPORT

A 14-year-old boy presented to Otorhinolaryngeal O.P.D. with the chief complaints of bilateral nasal obstruction, epistaxis, and headache in the presence of a purely nasopharyngeal mass. General physical examination was normal and laboratory blood tests were unremarkable.

MRI showed complex solid/cystic lesion with heterogenous signal intensity. Contrast enhanced T1-weighted images showed ring enhancement in solid component and peripheral wall.

A few days later, the patient underwent complete surgical resection of the mass and the specimen came for the histopathological examination.

The histopathological examination revealed: Grossly a greyish white cystic mass was received at our department. On cut section, cyst contained a greyish brown thick liquid like material. The excised tumour was further processed and routinely stained with haematoxylin and eosin. Microscopically tissue section showed cords and nodular whorls of well differentiated epithelium along with irregular trabeculae lined by palisading columnar epithelium. These compact areas were merging with loosely knitted epithelial tissue forming stellate reticulum. Pale nodules of wet keratin were found in both the dense and loose areas of the tissue. The histomorphology of the lesion was consistent with the diagnosis of craniopharyngioma.

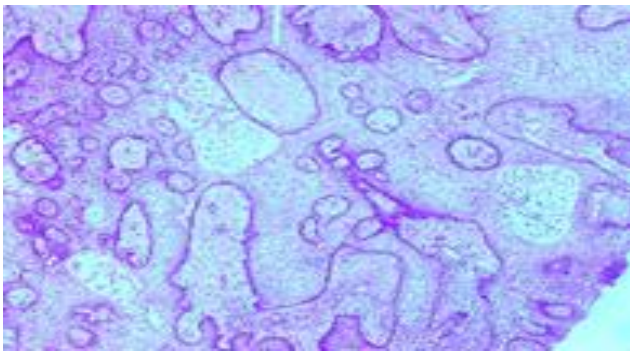

Fig.1a: 4x magnification H &E showing cords & nodular whorls of well differentiated epithelium along with irregular trabeculae

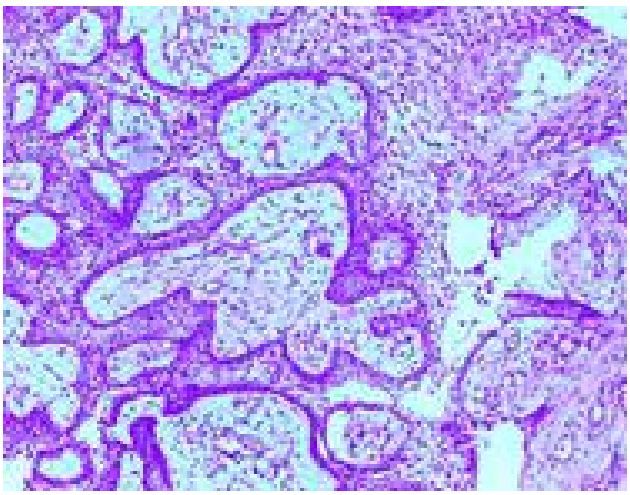

Fig.1b:10x Magnification H&E Showing Irregular Trabeculae Lined By Palisading Columnar Epithelium

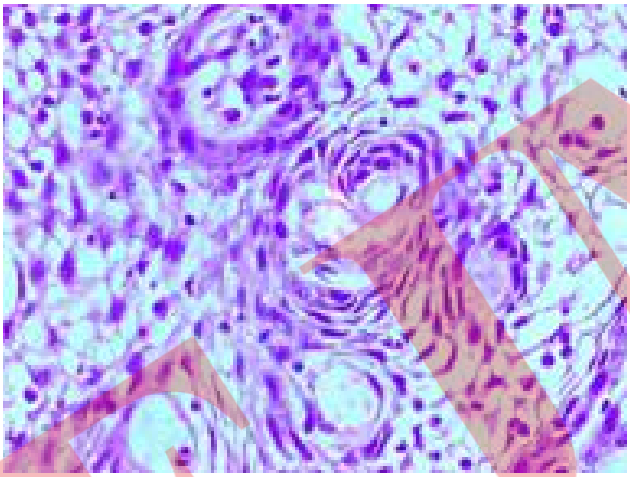

Fig.1c: 40 X Magnification H&E Showing Whorls Of Well Differentiated Epithelium

DISCUSSION

Craniopharyngiomas are benign but aggressive epithelial neoplasms, with almost similar frequency in children and adults; though a slightly higher incidence could be seen in the age group between 5 and 15 years. Equal gender distribution has been noted in groups of children.

Though 90 % tumours are located in supra-sellar location yet in about 5% of cases infra-sellar extension of the tumour is found. Rarely, it may arise primarily in unusual locations, such as the nasopharynx, sphenoid bone, third ventricle, pineal gland, sylvian fissure and cerebellopontine angle.[vi]

Histologically craniopharyngiomas are divided into two subtypes: the adamantinomatous and papillary types. The adamantinomatous variant is almost nine times more common than papillary variant and are pathologically distinct too. Adamantinomatous subtype, which are seen in children at infrasellar/suprasellar location, are predominantly cystic lobulated masses,. Squamous papillary craniopharyngiomas, on the other hand, are predominantly solid or mixed solid-cystic masses, which are often observed in adults at supra-sellar location. The solid components of the mass show an intense and inhomogeneous enhancement with some areas showing necrosis and rare foci of calcification. Both adamantinomatous and squamous-papillary subtypes could be seen in 15% of craniopharyngiomas.

Embryology - According to Warwick and Williams, during the fourth week of gestation ; in the roof of the stomodeum just anterior to the oropharyngeal membrane; an ectodermally lined diverticulum develops known as Rathke's pouch. An infundibulum descends as a neural outgrowth from the floor of the third ventricle (diencephalon) of the embryonic brain. Rathke's pouch ascends cranially traversing the mesenchyme to meet the neuroectoderm of the infundibulum (neurohypophysis) and differentiates into the anterior lobe of the pituitary, the adenohypophysis. While traversing this course, Rathke's pouch forms a cord of cells joining the stomodeal ectoderm to the future adenohypophysis in embryo. This cord gets disintegrated later in embryonic life, leaving an obliterated craniopharyngeal canal. It is a tract that runs from the anterior part of the hypophyseal fossa of the sphenoid bone to the junction of the posterior septum of the nose with the palate, which is the stomodeal end of the recess. The pharyngeal hypophysis (functioning adenohypophyseal tissue), which remains in adults, is a caudal remnant of this cord.

Types of origin – Different theories have been postulated regarding the origin of craniopharyngioma which take into account the different histologies along with the characteristic location of these tumors.[xii] The theories which explains the origin of adamantinomatous infrasellar craniopharyngioma, are based on the embryologic development of the adenohypophysis. In 1899, it was first time postulated by Mott and Barret that these tumours might have originated from the remnants of the pharyngeal hypophysis.[vii] In 1904 Erdheim proposed that the craniopharyngioma arose from the remnants of the obliterated craniopharyngeal duct, which in turn suggested that these tumours can arise anywhere along the tract of migration of Rathke's pouch from the vomer, the roof of the nasopharynx, through the midline sphenoid bone beneath the floor of the sella turcica.[viii] On the other hand ,the metaplastic theory states that the squamous papillary subtype is a result of metaplasia of squamous epithelial cell rests that are remnants of the part of the stomadeum that contributed to the buccal mucosa.

CONCLUSION

In conclusion, clinical features which includes age of the patient along with the imaging characteristics (except location) and histopathological findings are all compatible with primary admantinomatous craniopharyngioma. As the location of the tumour in purely infra-sellar region is a very uncommon incidence that's what make us to present this case as a report.

ABBREVIATIONS

O.P.D. –Out patient department

MRI- Magnetic resonance imaging

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