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Case Report

Adamantinomatous Craniopharyngioma-A rare Tumor arising from remnant of Rathkes pouch

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Abstract

Craniopharyngioma is a rare tumor arising from squamous rests located at any point along the invagination of the primitive stomodeum, Rathke's pouch, from the nasopharynx to the hypothalamus. There are two peaks in the occurrence of adamantinomatous craniopharyngioma, one in the first to second decade and the other in the fifth decade. They are generally sporadic and their molecular pathogenesis is poorly defined. There are two clinicopathological forms of craniopharyngiomas-papillary squamous type, and adamantinomatous craniopharyngioma. The classical adamantinomatous variant, which affects people of all ages consists of palisading columnar cells that look like ameloblast of embryonic tooth buds. It has poor prognosis when compared to papillary craniopharyngioma. It has an underlying mutation in β-catenin and translocation of the protein to the nucleus, which can be demonstrated by immunohistochemistry.

Keywords: craniopharungioma, adamantinomatous, poor prognosis, β Catenin,.

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INTRODUCTION

Craniopharyngioma is a rare tumor arising from squamous rests located at any point along the invagination of the primitive stomodeum, Rathke's pouch, from the nasopharynx to the hypothalamus [1]. There are two peaks in the occurrence of adamantinomatous craniopharyngioma, one in the first to second decade and the other in the fifth decade. These tumors have a bimodal age distribution. More than 50% of patients are children or adolescents and the peak incidence occurs between five and 14 years of age; there is also a second, smaller peak in the sixth decade of life [2,3]. Despite this, tumors may become symptomatic at any age. They are generally sporadic and their molecular pathogenesis is poorly defined.

CASE REPORT

A 18 year old male presented with history of intermittent headaches, nausea and vomiting headache for more than 3months. In addition, he reported diplopia, tinnitus for the past three weeks. Neurological examination showed he was ataxic and had hearing loss in his left ear, a mild left-side facial paresis. Other examinations were normal. On radiological examination showed a heterogeneous enhancement of the partially cystic lesion in the left cerebellopontine angle that compressed the adjacent brainstem and cerebellum. The patient underwent a left suboccipital retrosigmoid craniectomy. The post-operative course was uneventful and deficits in the cranial nerves improved three months after surgery.

Grossly, single cystic lesion which let out mineral oil like material and also solid areas noted. Microscopically, showed multiple cystic spaces surrounded by peripheral palisading nuclei and clusters of eosinophilic cells lacking nuclear staining, and surrounding piloid gliosis.(Fig.1). The stellate reticular pattern and reversal of polarization resemble adamantinomatous lesion of tooth (ameloblastoma)(Fig.2). Immunohistochemical results showed β-Catenin positive cells immediately adjacent to wet keratin or within whorled squamous nests rather than tumor cells (Fig.3).

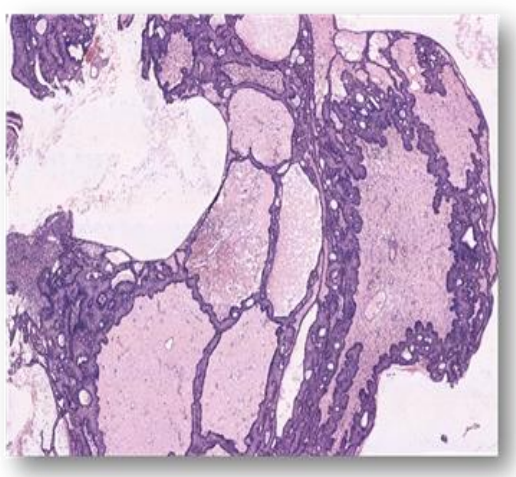

Fig.1: Multiple cystic spaces surrounded by peripheral palisading nuclei

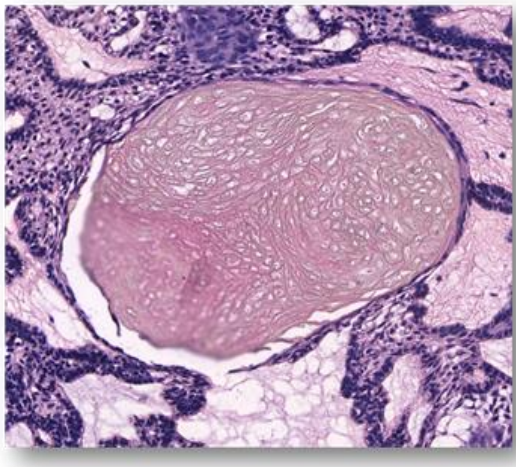

Fig.2: clusters of eosinophilic cells lacking nuclear staining.

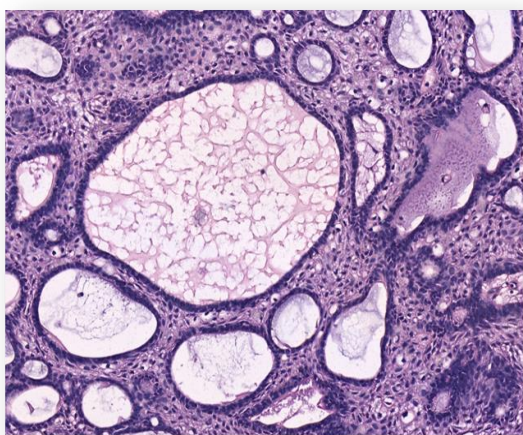

Fig.3: Stellate reticular pattern and reverse polarization which resemble adamantinomatous lesion

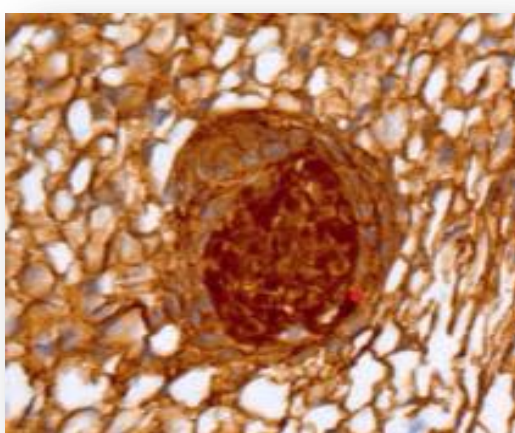

Fig.4: On β-Catenin showed positive for cells immediately adjacent to wet keratin or within whorled

DISCUSSION

Craniopharyngiomas is an ectopic craniopharyngioma that originates from migrated or trapped ectopic squamous cells of the craniopharyngeal duct during the rotation of the adenohypophysis in the embryonic period in the cerebellopontine angle(CPA). Growth of these ectopic cells can form a CPA craniopharyngioma [1,4]. They are generally sporadic and their molecular pathogenesis is poorly defined. These tumors are characteristic with peripherally palisaded nuclei, stellate reticulum, ghost cells, localised calcification, and often surrounding piloid gliosis with rosenthal fibre formation [5]. Although positive is often confined to cells within squamous morules, occasionally surrounding clusters of wet keratin (ghost cells), this type is now known to be related with beta catenin mutation and WNT pathway activation, as well as nuclear beta catenin immunoreactivity [6]. They are characteristic of peripherally palisaded nuclei, clusters of eosinophilic cells devoid of nuclear staining, so-called ghost cells (also known as wet keratin), focal calcification.

There are two clinicopathological forms of craniopharyngiomas. The papillary squamous type, which mostly affects adults mainly consists of mature stratified squamous cells, may be metaplastic and has a better prognosis. The classical adamantinomatous variant, which affects people of all ages consists of palisading columnar cells that look like ameloblast of embryonic tooth buds [7].

Papillary craniopharyngioma is composed of nonkeratinizing stratified squamous epithelium that is devoid of a keratohyalin layer (unlike epidermoid cyst) or wet keratin, ghost cells, or calcification (unlike adamantinomatous craniopharyngioma) Re-evaluation of adamantinomatous craniopharyngiomas for the presence of mesenchymal components may be required to determine where these biphasic tumours provide some insights to their histogenesis [8].

It has an underlying mutation in β-catenin and translocation of the protein to the nucleus, which can be demonstrated by immunohistochemistry. It should be noted that nuclear β-catenin staining is never seen in all tumor cells, but instead concentrates in cells immediately adjacent to wet keratin or within whorled squamous nests. Craniopharyngiomas are almost exclusively seen in adults and is related to mutations in BRAFV600E, which is paralleled by BRAF VE1 immunoreactivity [9].

CONCLUSION

We found that β -catenin gene mutation and overexpression in craniopharyngioma were unique in the adamantinomatous form. The literature says that adamantinomatous and papillary craniopharyngiomas are genetically distinct as well as histologically and clinically. Biphasic craniopharyngiomas will require more research to identify their significance.

Conflict of Interest

There is no conflict of interest.

Financial support and sponsorship

Nil

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published.

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