"An eye for a tooth:" Orbital teratoma in an adult

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Abstract:

A 27-year-old female presented with progressive prominence of her right eye since childhood. On examination, a pinkish mass filling the right orbit with no evidence of an eyeball was seen. She also had bilateral preauricular skin tags. Imaging revealed a mass with solid and cystic components in the right orbit, with a well-formed tooth *in situ*, suggestive of a teratoma. The tumor was removed and histopathology confirmed the diagnosis. Orbital teratoma in an adult is a rare presentation. The presence of preauricular skin tags in association with orbital teratoma is a new finding.

Keywords:

Germ cell tumor, orbit, preauricular skin tag, teratoma

Introduction

Teratoma is a congenital neoplasm containing structures derived from all three germ cell layers.[1] The patients usually present in the immediate newborn period.[2] We present an extremely rare case of a mature orbital teratoma in an adult.

Case Report

A 27-year-old female presented with the complaints of progressive prominence of the right eye since her childhood. On inspection, there was a pinkish nontender mass, cystic to firm in consistency, filling her right orbit with marked conjunctival prolapse [Figure 1a]. Facial examination showed bilateral preauricular skin tags [Figure 1b]. The left eye was within normal limits. The patient gave an alleged history of enucleation of the right eye during her infancy. Magnetic resonance imaging showed a complex solid, and a cystic mass with fat-fluid levels [Figure 2a]. Dense low signal intensity was seen at the apex suggestive of teeth [Figure 2a-arrow]. The right eyeball was absent. The imaging features were consistent with a teratoma. There was a gross expansion of the orbital volume on the right side. The mass was excised completely through an anterior transconjunctival approach under general anesthesia. The postoperative period was uneventful. Histopathology examination demonstrated derivatives of all three germ layers, along with teeth, hair follicles, mucinous glands, blood vessels, muscle fibers, and nerve fibers [Figure 2b]. There was no evidence of atypia or malignancy. A final diagnosis of a benign mature teratoma was made. We reviewed the patient after 8 weeks and she further underwent dermis fat grafting for socket reconstruction. She is awaiting customized prosthesis fitting for cosmetic rehabilitation [Figure 2c].

Discussion

Teratomas constitute 6.6% of childhood tumors. Orbital teratoma is a rare but important differential diagnosis for rapidly progressive unilateral proptosis in a newborn as the orbit is a typical location for primary extragonadal germ cell tumors. Females are affected more than males with a ratio of 2:1. The clinical presentations include proptosis, stretching of the lid over a fluctuating mass, or exposure keratopathy.[3,4] There can be diffuse enlargement of the bony orbit. The eyeball is normally developed in most of the cases but undergoes secondary degenerative changes due to displacement by the tumor. Rarely, life-threatening complications can occur due to intracranial extension. To date,

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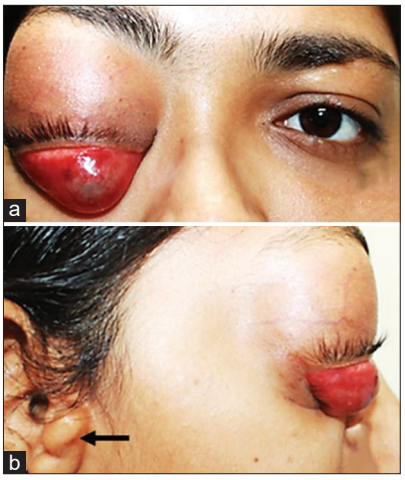

Figure 1: (a) Right orbital mass with conjunctival prolapse (b) right preauricular skin tag (arrow)

primary orbital teratomas have been reported in four adults.[5] Imaging shows multiloculated cystic masses with calcification, bone, and ossification.[4,5] The presence of preauricular skin tags as an association with orbital teratomas has not been noted previously. In the absence of any other ocular or systemic abnormality, it was presumed to be a sporadic association, and further genetic studies were not requested. The presence of a tooth inside a tumor is pathognomonic of a teratoma, which has been reported erstwhile in the orbit of two patients.[4]

Management of an orbital teratoma includes complete surgical excision with preservation of the eyeball if possible, or exenteration, followed by socket reconstruction.[5]

In all likelihood, the surgical intervention in the childhood of this patient did not accomplish complete removal of the tumor. However, in the absence of erstwhile surgical records, it is impossible to posit if the tumor was removed at all. The late presentation of the patient was due to her financial constraints and lack of awareness about cosmetic options. Cosmetic rehabilitation is essential in patients in whom the globe cannot be salvaged. Close follow-up is mandatory as orbital teratomas are known to recur and may even undergo malignant transformation.

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 initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

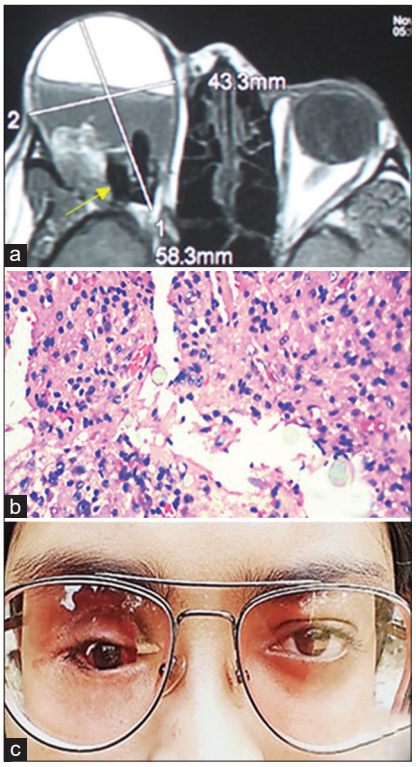

Figure 2: (a) Magnetic resonance imaging T1 axial section: heterogenous mass with fat-fluid levels with hypointense teeth at the apex (arrow); (b) microphotograph showing features of teratoma (H and E, ×40); (c) patient at last follow-up

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