Pediatric episcleral osseous choristomas: a case report

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Dear Editor,

We are writing to present a rare episcleral osseous choristomas in a 12-year-old girl. Choristomas are congenital benign mass composed of normal tissue located in an abnormal site[1]. Ocular choristomas can be categorized as limbal dermoid, lipodermoid, single-tissue choristomas (i.e., lacrimal gland, respiratory, and osseous choristomas), and complex choristomas[2]. Osseous choristoma is the rarest form of oculart choristomas comprising mature bone tissue, which often localizes in the superior temporal site of sclera, beneath the eyelid[3]. The relatively asymptomatic feature often results in a late presentation of the mass. We herein report a rare case of a 12-year-old girl with an episcleral osseous choristoma adhered tightly to the sclera in her right eye.

Ethical Approval This study complied with the tenets of the Declaration of Helsinki. Written informed consent was obtained from the patient’s mother for publication of this case report and any accompanying images.

CASE REPORT

A 12-year-old girl was referred to our eye center for management of mass in her right eye without a remarkable medical, traumatic, or family history. The mass was first noticed 2 years ago, and had slightly enlarged during the last 1mo. The girl complained about progressive foreign body sensation in the upper eyelid when blinking. On examination, her best corrected visual acuity was 20/20 in both eyes using the standard visual acuity chart at 5 m, with intraocular pressure documented to be 15 mm Hg bilaterally as measured by non-contact tomometer (TX-F, Canon Inc., Tokyo, Japan). Her eyes were orthophoric and extraocular movements were in normal limit without pain. Slit-lamp examination revealed a subconjunctival nodule (about 5×4 mm3) in the superior temporal quadrant at 8 mm behind the limbus. The nodule was firm, with clear margin and poor mobility. Nutrient vessels and inflammation surrounding the mass were noted (Figure 1A). Ultrasonic biological microscopy and B-scan confirmed a superior temporal located solid mass showing medium to high density indicating calcium components (Figure 1B, 1C). The calciferous mass was further confirmed by orbit computed tomography (CT), presenting as a high-density nodule (7×3×6 mm3) located in the lateral superior region of the eyeball (Figure 1D-1F). Dilated fundus photography and optical coherence tomography were unremarkable (Figure 1G, 1H), while visual field examination revealed slightly decreased sensitivity in the right eye (Figure 1I). All the examinations and assessments were performed in JSIEC. A suspected diagnosis of episcleral osseous choristoma was made after a thorough examination.

Informed consent was obtained and surgical excision and biopsy of the lesion was conducted under local anesthesia. During the operation, the mass was found whitish and adhered tightly to the sclera, with lack of neovascularization or necrosis (Figure 1J). The mass was carefully dissected and removed to avoid potential damage to the extraocular muscles and the sclera, using 15-degree ophthalmic knife (Alcon Inc.) and microscissors (Wenchuang Inc., China; Figure 1K-1M). The underlying sclera was nearly half as thick as normal after excision of the mass, as evaluated with a microscope on high magnification. Tenon’s capsule and conjunctiva were then sutured to close the incision site. Tobramycin and dexamethasone eye drops (TobraDex®; Alcon Inc., Fort Worth, Tex, USA) was administered q2h for 3d and then qid for a week, and tobramycin and dexamethasone eye ointment (TobraDex®) was applied qn for 10d, postoperatively. The mass was sent for pathological evaluation under hematoxylin and eosin staining. Histology of the specimen demonstrated...
mature bone without atypia (Figure 1N) and diagnosis of episcleral osseous choristoma was confirmed. Patient’s complaint of foreign body sensation had largely relieved 2wk postoperatively. At the 1-year follow-up, ocular examination showed normal anatomic and functional outcomes in her right eye and no local recurrence was found (Figure 1O).

DISCUSSION
In this study, we present a rare episcleral osseous choristomas in a 12-year-old child. Slit-lamp examination revealed a subconjunctival whitish nodule in the superior temporal quadrant of the eyeball. Ocular imaging examination revealed a high-density lesion indicating its calcium components.
Surgical dissection and removal of the lesion was conducted. Pathologic specimens revealed mature bone tissue without atypia and diagnosis of episcleral osseous choristomas was confirmed.

Episcleral osseous choristomas are the rarest form of ocular choristomas, with only less than 70 cases that have been reported in the literature since it was first described by Von Graefe in 1863[6-8]. Osseous choristoma is a benign mass comprising normal bone tissue in an epibulbar location[7]. In the eye, they are mainly found at the superior temporal region of the episclera (74%), and can be either freely mobile or firmly attached to the sclera or the extraocular muscles[8]. In addition, osseous choristomas slightly skew female (69%) and the right eye (76%)[9], and are rarely combined with systemic abnormalities[9]. Our case supports these statistical conclusions.

Episcleral osseous choristomas is believed to be congenital and grows very slowly[5]. Although some insults, such as trauma and inflammation, might trigger development of the mass[9], no malignant transformation or intraocular extension have ever been reported. In our case, the patient complained about enlargement of the mass and progressing foreign body sensation, likely due to the growth of tissues surrounding the osseous choristomas, such as connective tissue and fibroadipose tissue. Another possibility was that she has grown up and could palpate and identify the mass.

A pediatric epibulbar mass might be differentiated from several conditions including epibulbar dermolipoma, myxoma, prolapsed orbital fat, Kaposi’s sarcoma, sebaceous carcinoma and other malignant tumors, epibulbar foreign body, etc[10-12]. In this case, a high density of the lesion was found on CT scanning, indicating calcium components which may differentiate it from a dermolipoma and myxoma[13]. In addition, the mass was found solid and whitish and biological microscopic examination indicated calcium components, which should be differentiated from extraocular extension of retinoblastoma with calcification[14]. However, no abnormality was found in the fundus, suggesting less possibility of a retinoblastoma in this girl. Besides, the girl presented no pain or swelling of the eye, which did not support the fact of inflammatory disorders such as dacryoadenitis and sarcoidosis[15]. No systemic disorders between the first visit and the 1-year follow-up were found, indicating a small possibility of malignant lesions, such as Kaposi’s sarcoma[16]. A surgical biopsy is generally necessary to confirm the diagnosis and rule out these malignant disorders.

The management of an episcleral osseous choristomas includes conservative management and surgical excision. Observation can be alternative if the mass is typical with no complaints by the patient. Surgery is suggested in the following condition: discomfort caused by the sizeable mass (i.e., foreign body sensation, irritation, and pain), requirement for diagnosis, and cosmensis[17-18]. In our case, the osseous choristomas was surgically removed due to discomfort and patient demand. During operation, we noted that the lesion was adherent firmly to the underneath sclera. It seems hard to estimate the depth of invasion of the lesion. Therefore, careful dissection of the lesion should be taken to avoid iatrogenic globe perforation.

It is of great importance that a preoperative orbit CT may help evaluate the extent of scleral involvement and guide the choice of surgical procedures.

Ocular osseous choristomas are very rare ocular tumors. We herein described a pediatric episcleral osseous choristomas with a thorough examination, treatment and prognosis, which provided substantial information that an episcleral osseous choristoma should be distinguished when a mass is found in pediatric individuals. Future direction of research may be exploring whether ocular osseous choristomas is associated with systemic abnormalities as well as conducting genetic studies to find out specific mutations of the tumor.

In summary, we report a case of episcleral osseous choristomas in a 12-year-old girl, which localized in the superior temporal site of the episclera. In clinical practice, an epibulbar osseous choristoma should be considered when a mass is found in the superotemporal quadrant underlying the bulbar conjunctiva with characteristic calcification, particularly in pediatric individuals who had eye discomfort instead of visual defects.

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