Management of Extensive Epibulbar Choristoma Associated With Microphthalmos
A Rare Clinical Entity

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Choristomas are growths of histologically normal tissue in an abnormal location. Although typically not visually symptomatic, they may occur as extensive lesions that interfere with normal ocular development.1-4 We report 2 cases of unilateral extensive epibulbar choristomas associated with microphthalmia and describe our surgical management.

Report of Cases

Patient 1
An otherwise healthy, 1-month-old girl presented with a large lesion that extended from the right orbit (Figure 1A). The child was born of unrelated parents after an uneventful pregnancy and delivery. The lesion was present from birth and was well circumscribed and covered with skin. On ocular ductions, the lesion would partially move, presumably owing to some attachments to the extraocular muscles. Examination of the upper eyelid margin revealed erosion. No discernible ocular structures were identified on the right side. Orbital computed tomography revealed a cystic lesion that protruded from the right orbit with the rectus muscles and optic nerve attached to the posterior aspect of the mass (Figure 1B). Comparison of the orbital volume of the fellow, normal side revealed no significant bony hypoplasia.

To discern the identity of the mass, excision and reconstruction were performed for functional and aesthetic reasons. A subtotal resection was performed to the level of the conjunctiva resulting in no residual tumor extruding from the orbit. Gross examination revealed that the lesion was covered by skin, and deeper evaluation revealed abundant adipose tissue and, at the core, a small mass of dark, uvea-like tissue (Figure 1C).

To reconstruct the socket, we harvested the skin that covered the anterior aspect of the lesion. This free graft was then secured to the cut edge of the conjunctiva (Figure 1D), and an ocular conformer was inserted. One month after surgery, a customized prosthesis was fabricated to the satisfaction of the parents and surgeons, although independent, objective documentation of the outcome was not obtained.

Pathologic examination revealed the lesion to be covered anteriorly by mature, keratinized stratified squamous epithelium and dermis. Deep to the dermis was adipose tissue and collagenous connective tissue that surrounded a small irregular cystoid structure (Figure 2A). The wall of the small cyst was composed of a coarsely organized layer of collagenous fibers with no mature intraocular structure inside except for some neuroglial and uveal tissues (Figure 2B).

Patient 2
A 2-year-old girl presented with a large, fingerlike mass that protruded from the right orbit at birth (Figure 3A). The child was born of healthy, unrelated parents after a normal pregnancy. This lesion was associated with a coloboma of the right upper eyelid and erosion of the lower eyelid margin. No systemic abnormalities or hypoplasia of the affected orbit were found. Ophthalmic examination revealed a normal left eye.

The surgical management of this patient required 3 separate operations to achieve a functional and cosmetically acceptable outcome.

IMPORTANCE Extensive epibulbar choristomas associated with microphthalmia have rarely been reported. We report 2 cases and describe our experience in the diagnosis and management of this challenging entity.

OBSERVATIONS The clinical features, surgical and histopathologic findings, and aesthetic results of surgical treatment were recorded.

CONCLUSIONS AND RELEVANCE Surgical management of extensive epibulbar choristoma associated with microphthalmos using a simplified method of partial tumor excision and skin grafting can result in improved cosmetic outcomes.

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acceptable outcome. During the first operation, we debulked the tumor and reconstructed the fornix with a piece of skin harvested from the anterior aspect of the lesion. A conformer was placed and held in position by tarsorrhaphy. The coloboma was not addressed at this point. Six months after the initial operation (Figure 3B), we opened the eyelids and found that the socket was contracted. During the second operation, we reconstructed the socket with a full-thickness skin graft from the medial upper arm. The upper eyelid coloboma was addressed by resection and eyelid margin reconstruction. One year after the second operation (Figure 3C), frontalis suspension was used to correct the ptotic eyelid. Follow-up after 2 years revealed improved function and cosmetic satisfaction by the parents and surgeons (Figure 3D). Gross and pathologic examination revealed findings similar to patient 1 with skin anteriorly, abundant adipose tissue in the interstitium, and a core of disorganized uveal tissue.

Discussion

The primary treatment goal of extensive epibulbar choristoma associated with microphthalmos should be focused on achieving cosmesis in young patients because vision is always lost. Traditionally, epibulbar choristomas have been treated by evisceration, enucleation, or even subtotal exenteration. Huang and colleagues reported a modified method in which they debulked the external portion of the tumor and closed the wound at the conjunctival level, leaving the residual tumor in the orbit to stimulate bony orbit development. Their method was rationalized on the benign nature of choristomas. They concluded that the mass should be excised at an early age when local mechanical erosion to the eyelid occurs and the appearance is disfiguring. We performed 2 such similar operations for our 2 patients. We debulked the anterior portion of the tumor, but instead of direct closure, we opted to use the skin from the anterior surface of the tumor and secured the cut edge of the conjunctiva to reconstruct the socket.

A. A 1-month-old girl with a large lesion protruding from the right eyelid tissue. B. Orbital computed tomography revealed a cystic lesion protruding from the right orbit, with the rectus muscles and optic nerve attached to the posterior aspect of the mass. C. The tumor was partially resected at the conjunctival level. Gross examination revealed that it was covered by skin, and deeper evaluation revealed abundant adipose tissue and, at the core, a small mass of uvea-like tissue. D. A piece of skin was harvested from the anterior surface of the tumor and secured to the cut edge of the conjunctiva to reconstruct the socket.

A. The lesion was covered by keratinized, stratified squamous epithelium and fat-abounded stroma (hematoxylin-eosin; original magnification ×10). B. The wall of the microphthalmic globe was composed of a coarsely organized layer of collagenous fiber. Some disorganized neuroglial structure, uveal tissue, and connective tissue were present inside the wall (hematoxylin-eosin; original magnification ×50).
Lesion to avoid donor site morbidity, but when necessary the upper arm provides a suitable source of skin with minimal functional and cosmetic impairment.

An additional consideration in patients with epibulbar choristoma associated with microphthalmos is the potential for sympathetic ophthalmia. When excising a cyst with potential uvea, care must be taken to completely remove all remnants of uveal tissues to minimize the risk to the fellow eye.

In conclusion, use of a simplified method of partial tumor excision and skin grafting can result in cosmetic outcomes as noted in our patients and documented in the second case.

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