A Rare Pediatric Case of Lacrimal Gland Pleomorphic Adenoma

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Summary: Pleomorphic adenoma of the lacrimal gland is a rare benign finding in adults, and extremely uncommon in children. These tumors often present with increased proptosis or hypoglobus, and they invariably require operative excision. While many of these lesions can be removed through a subcranial or transorbital exposure, larger and more posterior tumors occasionally necessitate an intracranial approach. This report describes a large lacrimal gland pleomorphic adenoma in a 16 year-old patient that required use of an expansive intracranial exposure to ensure complete excision including the capsule. We contrast this case and its management to other pediatric cases described in the literature. (*Plast Reconstr Surg Glob Open 2019;7:e2435; doi: 10.1097/GOX.0000000000002435; Published online 24 September 2019.*)

L acrimal gland pleomorphic adenoma (LGPA) is the most common benign epithelial tumor of the lacrimal gland in adults, typically arising in the third to fifth decade of life.[1] Most cases present with inferomedial globe displacement, lateral upper lid fullness, exophthalmos, and blepharoptosis. Less common findings include diplopia or changes in vision and/or reduced extraocular movement. These signs and symptoms, coupled with characteristic features on computed tomography (CT) or magnetic resonance imaging (MRI), are usually sufficient to secure the diagnosis and obviate the need for biopsy, which may increase the risk of recurrence along the biopsy path. Treatment of LGPA consists of intact capsule excision, if possible, to reduce the risk of recurrence or malignant transformation. Successful excision of an intact capsule yields an excellent prognosis with a reported recurrence rate <3% after 5 years.[2]

Lacrimal fossa lesions are much less common in children, comprising 0.3%–4% of orbital space-occupying lesions[3] and most are inflammatory pseudotumors. The differential diagnosis of lacrimal fossa tumors includes dermoid cysts, vascular tumors, inflammatory lesions, lymphoma, expleomorphic adenoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, adenocarcinoma not

otherwise specified, and other very rare epithelial malignant neoplasms. LGPA is exceedingly rare in children <20 years of age. We report a LGPA in a 16-year-old patient treated by an anterior cranial base approach. The collection and evaluation of protected patient health information was HIPAA compliant.

CASE PRESENTATION

A 16-year-old male presented with a 2-year history of slowly progressive proptosis and right globe inferior displacement. The patient denied headache, numbness, orbital pain, or visual changes. His family first brought the facial asymmetry to his attention. He had no significant medical or surgical history, medications, or family history of orbital tumors.

Physical examination demonstrated proptosis (7 mm confirmed by Hertel exophthalmometry) and hypoglobus (1 cm) without blepharoptosis, periorbital sensory deficit, or epiphora (Fig. 1). Uncorrected visual acuity was 20/20 with normal intraocular pressure, normal anterior segment, pupillary, and fundal findings. Extraocular movements were intact, without weakness, limitations, diplopia, or pain.

CT and MRI, demonstrated $2 \times 2\,cm^2$ contrast-enhancing homogenous soft tissue mass in the superolateral right orbit extraconal space with intraconal space breach and anterior-inferior globe displacement. The tumor encroached upon the superior rectus muscle but optic nerve was spared; the remainder of the brain and contralateral orbit were normal. No significant bony erosion was present. The patient deferred treatment until completion of school and 3-month interval imaging demonstrated further enlargement (measuring $3.2 \times 2.2 \times 1.8\,cm^3$) (Fig. 2).

Fig. 1. Preoperative photograph demonstrating proptosis and hypoglobus.
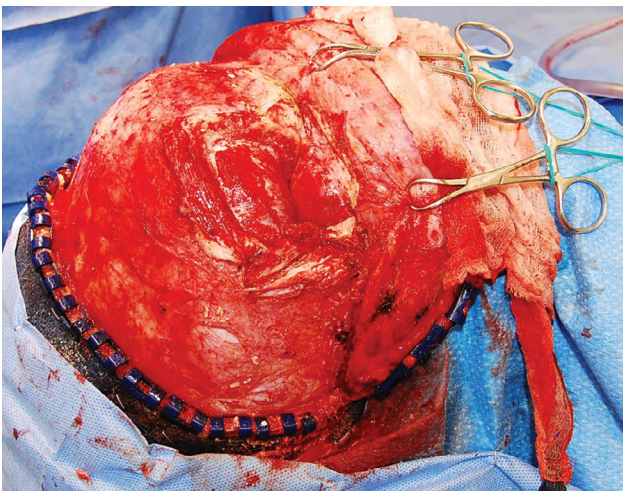

Fig. 2. Coronal view, gadolinium-enhanced MRI demonstrating well-circumscribed right superolateral orbital mass

To avoid biopsy tract seeding, biopsy was not performed. Due to the size and involvement of the mass with the extraocular muscles, an interdisciplinary team of neurosurgery, plastic surgery, and ophthalmology approached the mass
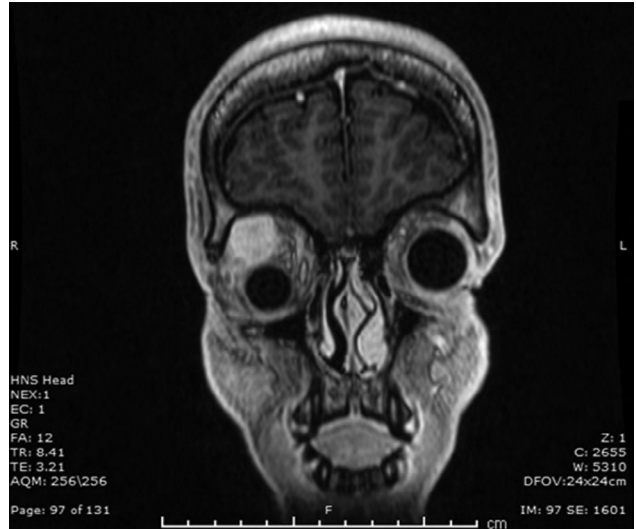

Fig. 3. Bicoronal approach, right front-orbital craniotomy, including removal of orbital roof.

through an intracranial approach. After coronal incision and fronto-orbital craniotomy, the right orbital roof was removed to provide wide superior/lateral exposure of the tumor (Fig. 3). There was no evidence of bony erosion or infiltration of the surrounding tissue. The periorbita was opened and the tumor meticulously dissected from superior and lateral rectus muscles without loss of any rectus muscle, and the supraorbital nerve allowing en-bloc resection with an intact capsule. The orbital roof, supraorbital bar, and frontal bone window were anatomically reconstructed utilizing absorbable hardware. Intraoperative frozen section pathology demonstrated glandular neoplasm with squamous differentiation thought to be benign. Final pathologic examination demonstrated an encapsulated glandular neoplasm with squamous differentiation consistent with pleomorphic adenoma.

At 11-month follow-up, the patient was found to have resolution of proptosis, no hypoglobus, normal extraocular movements without strabismus, excellent frontal contour, and well-healed coronal incision (Fig. 4). He denied any double vision, changes in vision, or dry eye.

DISCUSSION

Pleomorphic adenoma, the most common benign epithelial tumor, is also referred to as "benign mixed tumor" due to presence of mesenchymal and epithelial elements.[2] Malignant transformation within a pleomorphic adenoma is known as expleomorphic adenoma or malignant mixed tumor and may occur with prolonged tumor duration or incomplete resection.[2] LGPA has been rarely reported in the children and is not seen in newborns or infants; the youngest reported case was in a 5-year-old child (Table 1). Bajaj et al[3] reported 119 pediatric orbital space-occupying lesions, finding a 0.8% incidence of LGPA, whereas larger case series of pediatric space-occupying orbital lesions reported no cases of pleomorphic adenoma.[14]

The most common clinical findings of pediatric LGPA are similar to those reported in adults—lateral upper lid

Fig. 4. Postoperative photograph demonstrating resolution of proptosis and hypoglobus.

fullness, inferior globe displacement, exophthalmos, diplopia, blepharoptosis, reduced ocular mobility, or changes in vision.[2] One distinction between adults and children is the duration of symptoms before seeking care. While the average interval between symptoms onset and seeking care in adults has been reported to be 1–2 years,[2] our review of 10 reported cases of pediatric LGPA found a mean interval of 4.3 months in children <16 years of age (Table 1). Proptosis was the most commonly reported complaint, followed by blepharoptosis and periorbital or eyelid "swelling" (Table 1). We suspect heightened parental vigilance is responsible for the reduced interval observed in pediatric presentation.

Clinical findings coupled with diagnostic imaging are the basis for diagnosing lacrimal tumors. High-resolution CT, ultrasonography, and MRI have all been reported modalities of diagnostic imaging in pediatric LGPA cases with CT being most common. Rose et al[1] proposed an algorithm for the preoperative management of lacrimal gland masses with scores assigned to clinical and radiologic findings. Clinical findings concerning for malignancy in adults include patient presentation within 10 months of onset, persistent pain, and sensory loss.[1] Radiologic findings that may suggest malignancy include irregular mass shape, calcification within the tumor, bone invasion or destruction, and molding along the lateral orbital wall or globe.[1] In benign masses, this algorithm assists in the avoidance of biopsy, which could lead to biopsy tract seeding of tumor cells with a higher risk of recurrence or malignant transformation. If a decision is made to proceed with biopsy, it is typically performed through an anterior approach with either incisional or fine needle aspiration (FNA) with biopsy tract excision at the time of definitive treatment. Our confidence in the diagnosis was strong and preoperative biopsy was avoided to reduce the risk of recurrence.

The choice of surgical approach for pediatric lacrimal gland neoplasms is dependent upon tumor location, size, and pathology as well as surgeon experience. Commonly reported direct surgical approaches include anterior, later-

Table 1. Pediatric Case Reports of LGPA in the Literature

Author	Age	Complaints	Duration (mo)	Exam Findings	Tumor Size (cm)	Surgical Approach
Gupta S. 2013[4]	5 y	Painless progressive supraorbital fissure swelling	12	Ptosis	2.85×1.26×1.27	FNA; LO
Faktorovich 1996[5]	6 y	Blepharoptosis	6	Immobile nontender palpable upper eyelid mass	2.0×1.3×1.8	LO
Gupta A. 2013[6]	7 y	Painless, progressive, non-pulsatile proptosis	2	3 mm proptosis; inferomedial globe displacement; upgaze restriction	2.7 x 3.1	USG FNA; LO
Chen 2005[7]	9 y	Proptosis	2	3 mm proptosis	3.5 x 2 x 1.5	LO
Korchak 2015[8]	9 y	Proptosis and diplopia on far gaze	2	4 mm proptosis; 85% restriction in superior and temporal gaze of affected globe	2.3×1.6×1.2	Biopsy + LO
Cates 2002[9]	10 y	Painless eyelid swelling	3	3 mm proptosis; inferomedial globe displacement; non-tender lacrimal gland mass	1.8×1.8	LO
Vijayakumar 2013[10]	11 y	Slow progressive swelling of lateral aspect R eye	6	Firm painless mass of lateral R orbit	4×3×2	Modified LO
Mercado 1998[11]	15 y	Proptosis; 4 y intermittent swelling of eyelid	1	2 mm proptosis; firm non-tender mass fixed to superolateral orbital rim	4×3×3	SLO
Perez 2006[12]	16 y	Blepharoptosis	60	Palpebral ptosis	2.5	NR
Chandrasekhar 2001[13]	17 y	Progressive proptosis	36	5 mm proptosis; inferolateral displacement of R globe; diplopia on superior and right lateral gaze	2.5 ovoid	SLO

FNA, Fine Needle Aspiration Biopsy; LO, Lateral Orbitotomy; SLO, Superolateral Orbitotomy.

al, and superolateral orbitotomy. Anterior orbitotomy has been described for incisional biopsy and excision of lesions in the extraconal anterior two-thirds of the orbit. In this approach, an upper eyelid crease, infrabrow, or supraorbital incision is made without need for bone removal. Lateral orbitotomy provides deeper exposure than the anterior approach and is the most commonly described approach to lacrimal gland and extraconal lateral orbital lesions. In this approach, osteotomies performed through a lazy-S pattern incision proceeding from the lateral brow posterolaterally along the zygomatic arch allow removal of the lateral orbital wall. Of course, this approach leaves a visible lateral facial scar, which may be objectionable in a child.

Superolateral orbitotomy is less commonly reported, but has been advocated for improved visualization of the superior orbit compared with the lateral orbitotomy and has been advocated for intraconal tumors and those near the orbital apex.[16] In this approach, the lateral orbital rim, a large portion of the superior orbital rim, and lateral orbital wall are temporarily removed through a coronal incision.[16] Variations of this procedure include a frontotemporal craniotomy with en bloc removal of the orbital roof, superior orbital rim, and lateral orbital wall. Regardless of the approach, the goal of treatment is mass excision en-bloc with capsule intact to avoid myxoid component leakage.[2] Even with complete excision, a 3% recurrence rate after 5 years has been reported.[2] We elected for an intracranial exposure with orbital roof removal to reduce the risk of recurrence by ensuring that the capsule and tumor were completely excised.

Our review of the English literature found 10 case reports of pediatric LGPA (Table 1); only 1 case report did not state the surgical approach chosen.[12] Preoperative biopsy (FNAC) was performed in 2 patients.[6,15] The justification given for preoperative biopsy was adherence to an algorithm for superotemporal orbital masses[15] and as the result of a radiological recommendation.[6] Lateral orbitotomy was the most commonly reported surgical approach, described in 7 reports. None of the case reports describing the LO approach provided a postoperative photograph and only 1 reported the postoperative exam, limited to a comment that the patient was symptom free at 1 year.[15] Superolateral orbitotomy approach was reported in 2 cases. Both authors commented that the approach improved visualization and ensured complete excision, although neither report mentioned intraconal extension of the tumor. Only one case report[13] provided a postoperative photograph, demonstrating resolution of proptosis/hypoglobus. Each of the reported cases emphasized the importance of intact capsule excision to limit recurrence and malignant transformation, which is aggressive and may necessitate orbital exenteration.

CONCLUSIONS

LGPA is very rare in children. The presenting symptoms are similar to those observed in adults, but the duration of symptoms is appreciably shorter. Diagnosis hinges on a combination of clinical and radiographic findings. Diagnostic biopsy is rarely indicated. The risk of recurrence and/or malignant transformation is dependent upon the completeness of excision. For larger lesions and those with intraconal extension, coronal incision and fronto-orbital craniotomy provides excellent access and reduces visible facial scars.

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