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## Ectopic Lacrimal Gland Presenting as an Orbital Mass in Childhood

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# Ectopic Lacrimal Gland Presenting as an Orbital Mass in Childhood

Proptosis in children may be an ominous sign suggesting rhabdomyosarcoma, lymphoma, metastatic neuroblastoma, Ewing sarcoma, eosinophilic granuloma, or orbital cellulitis, among others [1]. Ectopic lacrimal gland tissue within the orbit is an uncommon etiology of proptosis in infants and children [2], although it may be more common in adults [2]. We describe two cases of proptosis in children associated with ectopic lacrimal gland tissue within the orbit and describe the features suggesting this diagnosis on CT of the orbit.

## Case Reports

### Case 1

A 16-month-old boy developed proptosis suddenly, 5 days before examination. Neuroophthalmic examination revealed visual fixation to be central, steady, and maintained in both eyes. There were 4 mm of proptosis and resistance to retropulsion of the right globe. The pupils, ocular motility, and ophthalmoscopy were normal. CT revealed a multilobular, enhancing mass lesion involving the superior, lateral, and medial intraconal areas of the orbit, without evidence of bone erosion (Fig. 1). Surgical exploration of the orbit with biopsy of the mass disclosed ectopic lacrimal gland tissue distributed throughout the orbit. Histopathology revealed normal-appearing glandular elements of lacrimal tissue with scattered foci of mature lymphocytes. A normal bone-marrow biopsy and hemogram excluded malignancy of the hematopoietic system.

### Case 2

A 6-month-old girl had been born with nonprogressive proptosis of the right globe. Neuroophthalmic examination revealed visual fixation was central and steady in both eyes. The right globe had 5 mm of proptosis and was resistant to retropulsion. Exposure keratitis was present in the right eye and was caused by the proptosis. The pupils, eye movements, and ophthalmoscopy were normal. CT disclosed a multilobular enhancing mass involving the intraconal space (Fig. 2). Surgical exploration with biopsy of the mass revealed ectopic lacrimal gland tissue. Histopathologic examination showed normal-appearing lacrimal tissue, without evidence of round cell infiltration. Corneal exposure was treated with topical lubricants.

## Discussion

Ectopia of the lacrimal gland is the presence of this glandular tissue at a site where it ordinarily does not occur. Our two cases clearly illustrate ectopic lacrimal gland tissue within the orbit, outside the lacrimal fossa. In our first case, mild inflammation may have contributed to the sudden appearance of proptosis [2], whereas proptosis

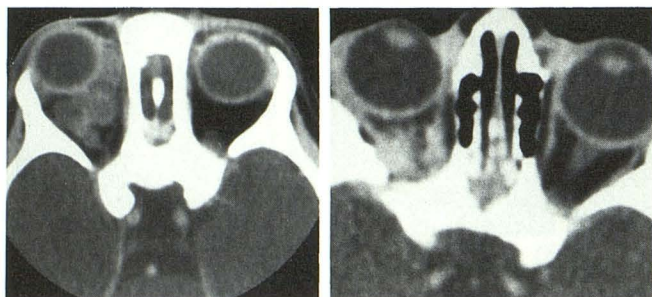
in our second patient was nonprogressive. The benign nature of the orbital masses in both patients was suggested by the lack of ocular motility disturbance, although loss of vision and ophthalmoparesis has been described with ectopia of the lacrimal gland [2-4].

Ectopic lacrimal tissue is an uncommon etiology of proptosis, therefore it may be incorrectly diagnosed as a neoplasm. Enhancing multilobular masses of consistency similar to that of the normal lacrimal gland and the absence of either erosion of the orbit or involvement of the paranasal sinuses on CT were characteristic of both our cases. Only four reports have described the CT appearance of ectopic lacrimal tissue within the orbit [4-7]. Margo et al. [5] described a case with an enhancing mass adjacent to and involving the lacrimal gland and fossa; however, histopathology of their case revealed dense fibroconnective tissue replacing normal lacrimal glandular elements, unlike the normal features of the ectopic lacrimal gland tissue in both of our cases. Salvolini et al. [7] showed a case with dacryoadenitis, in addition to the ectopic lacrimal gland, which appeared in a superior-lateral position unlike the distribution in the intraconal orbit in our two cases. In the case described by Appel and Som [6], concomitant orbital varix, in addition to the ectopic lacrimal gland tissue, made indistinct the CT features that distinguish these two entities. Jacobs et al. [4] depicted ectopic lacrimal tissue in an inferior location on CT. Rush and Leone [8] mentioned a CT scan in their case report, but it was not shown. Thus, the CT and histopathologic features of our two cases of lacrimal gland ectopia were more clear than some of those previously reported [5-7].

Involvement of the adjacent paranasal sinuses is characteristic of orbital cellulitis in children, but it is not observed with orbital pseudotumor or ectopia of the lacrimal gland. Both pseudotumor of the orbit and ectopia of the lacrimal gland are uncommon causes of proptosis in children. Changes of the orbital bone are more frequent with malignant tumors, but such changes are absent with orbital pseudotumor and ectopia of the lacrimal gland [4-7]. Therefore, multilobular, enhancing orbital masses and the absence of bony or paranasal sinus involvement on CT may suggest ectopic lacrimal gland.

Ectopic lacrimal gland tissue may result in benign cystic lesions of the orbit [2] or neoplastic transformation of ectopic lacrimal tissue to pleomorphic adenoma and adenocarcinoma [2, 9]. Therefore, surgical biopsy and histologic confirmation of the identity of the orbital lesion may be necessary for accurate diagnosis.

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**1** **2**  
Fig. 1.—Case 1: Multilobular enhancing mass in intraconal area of orbit above optic nerve and adjacent to it.  
Fig. 2.—Case 2: Enhancing mass in intraconal space, distant from lacrimal gland.

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