Case Report

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# Ectopic lacrimal gland causing intermittent proptosis

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**Abstract:**

Unilateral proptosis secondary to ectopic lacrimal gland tissue is rare. The most common site of ectopic lacrimal gland tissue is at the bulbar conjunctiva and limbal area. Although uncommon, intraorbital ectopic lacrimal gland tissue may mimic other ominous symptoms of intraorbital neoplasm in childhood. We present a rare case of intraconal ectopic lacrimal gland tissue in a 14-year-old girl with intermittent proptosis since childhood associated with subconjunctival hemorrhage and excruciating pain. She underwent lateral orbitotomy with orbital mass excision that resulted in good outcomes and no recurrence was seen at 6 months after the surgery.

**Keywords:**

Ectopic lacrimal gland, intermittent proptosis, subconjunctival hemorrhage

## Introduction

The lacrimal gland comprises the orbital lobe located at the orbital fossa and a smaller palpebral lobe located at the temporal side of the superior fornix.[1] Lacrimal gland tissue that located at other site is defined as ectopic lacrimal gland tissue.[1,2] The most common site of the ectopic gland tissue is the bulbar conjunctival and limbal area.[1-3] Orbital involvement is relatively uncommon.[3,4] The ectopic lacrimal gland is derived from the invagination of the supratemporal embryonic conjunctival epithelium into the anterior orbital soft tissue and occasional deeper migration into the orbital soft tissue.[2,5,6] The clinical presentation of the ectopic lacrimal gland varies depending on its exact location and pathology.[5] On reviewing the literature, only one case report of ectopic lacrimal gland tissue was found with intermittent episodes of proptosis with conjunctival congestion.[2] We describe a rare case of retroorbital intraconal ectopic lacrimal gland tissue presenting with intermittent bouts of proptosis associated with intolerable pain and subconjunctival hemorrhage during the acute presentation.

## Case Report

A 14-year-old girl with no known medical illness presented with intermittent subconjunctival hemorrhage with proptosis for 3 years of age. It was associated with left eye pain, headache, and vomiting. It was not related to fever or straining maneuver and resolved by itself after 3–4 days. However, the attacks had been increasing in frequency for 1 year, associated with the worsening of symptoms. On examination, vision for both eyes was 6/6 with no relative afferent pupillary defect. The left eye appeared proptosed with patches of subconjunctival hemorrhage in the inferotemporal quadrant [Figure 1], and its extraocular movements were markedly restricted. Computed tomography (CT) scan of the orbit showed left eye proptosis with the distance from the interzygomatic line to the anterior surface of the globe, measuring 23.7 mm [Figure 2]. The left eye proptosis resolved spontaneously a few days later. Magnetic resonance imaging (MRI) showed a well-defined, lobulated, and intraconal lesion within the left orbit measuring

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2.0 cm × 1.1 cm and located between the optic nerve and lateral rectus muscle, displacing the nerve medially and the muscle laterally [Figure 3]. The patient underwent left lateral orbitotomy with excision of the orbital mass. The microscopic section revealed ectopic lacrimal gland tissue with hyperplastic changes and scattered foci of mature lymphocyte, and no abnormal vascular component or varices was seen [Figure 4].

## Discussion

The ectopic lacrimal gland is uncommon and rarely reported. The age of onset of the ectopic lacrimal gland tissue can vary drastically from as young as 5 months to older adults, but it is usually presented in childhood.[2,7-9] In our literature review, we found 31 cases of ectopic lacrimal gland tissue with various clinical presentations.[2-4,6,7,10] The most common clinical presentation of the orbital ectopic lacrimal gland was progressive proptosis.[2-4,6,7,10] Of that, only one case report described a patient with intermittent episodes of proptosis, similar to that seen in our patient.[2] However, none of these cases reported with intolerable pain and subconjunctival hemorrhage. The patient in this case report, apart from intermittent proptosis, also presented with intolerable pain and subconjunctival hemorrhage, which required hospital admission.

Intermittent proptosis is characterized by a brief episode of proptosis. There are a few causes of intermittent proptosis, of which orbital varices are the most common (90% of cases).[11] Orbital varices are congenital venous malformations, normally diagnosed in early childhood to late middle age. The head position, usually lowering of the head and valsalva maneuver causes engorgement of the varix and leads to intermittent proptosis.[12] Other uncommon causes include highly vascular orbital neoplasms that are prone to periodic congestion, such as orbital lymphangioma and recurrent orbital hemorrhage.[13]

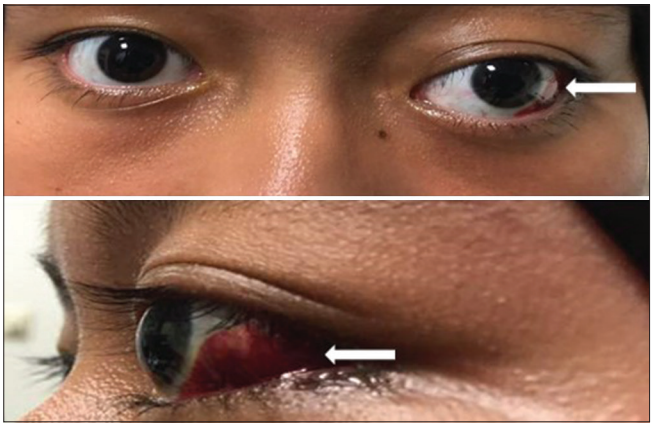

**Figure 1:** Left eye proptosis with subconjunctival hemorrhage inferolateral (arrow)

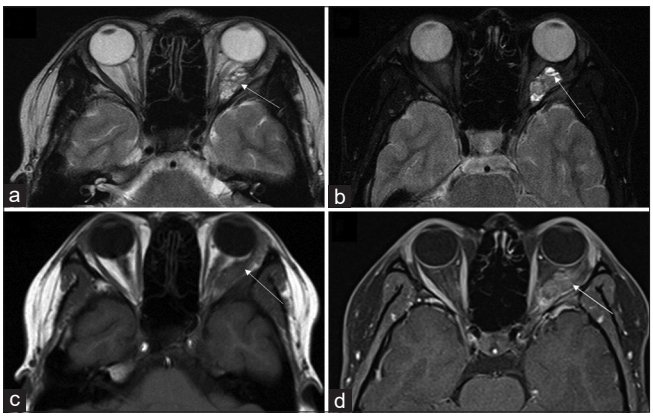


**Figure 3:** T2-weighted image (a) showing a fairly well-defined heterogeneous hyperintense left intraconal mass between the optic nerve and lateral rectus muscle, with medial displacement of the optic nerve and mild proptosis. T2 fat-saturated image (b) showing the mass has a cystic component. T1-weighted image (c) showing this mass is an isointense to the recti muscle and optic nerve. T1 fat-saturated postcontrast image (d) showing the mass with heterogeneous contrast enhancement (arrow)

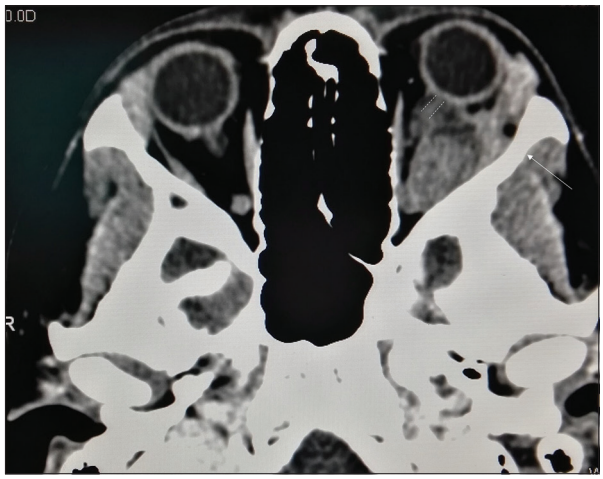

**Figure 2:** The lesion abuts the lateral orbital wall with thinning of the adjacent cortex (arrow). The optic nerve is pushed medially by this lesion with obliteration of the fat plane (dotted line)

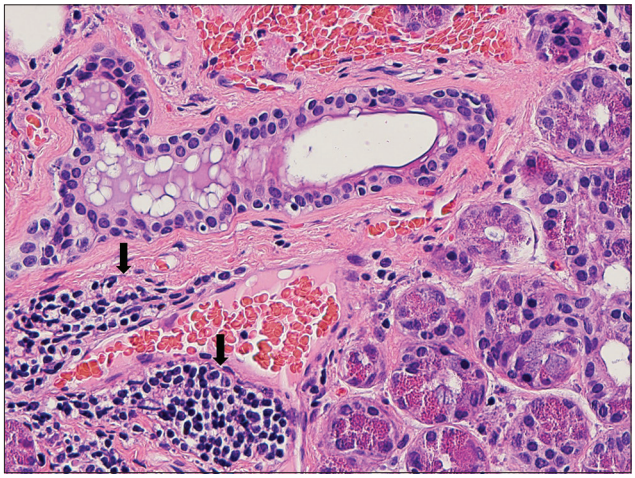

**Figure 4:** Acini are lined by secretory cells of serous type with granular cytoplasm. The interlobular duct is lined by two layers of low columnar cells. Note the adjacent lymphocytic infiltrates (arrow) with no abnormal vascular component or varices (H and E stain, ×400)

The exact mechanism of intermittent proptosis in ectopic lacrimal gland tissue is unknown and may be because of an underlying inflammatory process. However, what initiates the inflammatory reaction is unclear. Green and Zimmerman postulated that the secretion of the ectopic lacrimal gland is not drained adequately, and the resultant accumulation of the secretory product may initiate cyst formation, inflammation, and proptosis.[2] The ectopic lacrimal gland indeed does not has a proper ductal system, and an inflammatory reaction can be initiated.[4] In our patient, the histological examination of the excised tissue demonstrated an inflammatory process. Subconjunctival hemorrhage that presented during the bout of proptosis could be due to venous congestion secondary to the inflammatory process.

Although CT and MRI are the common imaging modalities used for the investigation of the ectopic lacrimal gland, they are not diagnostic.[3-5,10] Histopathology is crucial for the definitive diagnosis and differentiation from a neoplastic growth. Complex and simple choristoma are the most common histopathological subtypes of the ectopic lacrimal gland.[14] Neoplastic transformation of the ectopic lacrimal gland, for example, to pleomorphic adenoma and adenocarcinoma, is extremely rare but has been reported.[5,14]

In conclusion, intermittent proptosis and subconjunctival hemorrhage are rare presentations of ectopic lacrimal gland tissue. Although it carries a good prognosis, neoplastic transformation is possible and needs to be ruled out.

## Declaration of patient consent

We have obtained all appropriate patient and guardians consent form. In the form, the patient and her parents have given their consent for her image and other clinical information to be reported in the journal. The patient and parents understand their name and initials 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

The authors declare that there are no conflicts of interests of this paper.

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