Orbital involvement in lacrimal drainage disorders

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Abstract:

Lacrimal sac is situated anterior to the orbital septum, which acts as a barrier, thus limiting the posterior migration of the pathologies affecting the lacrimal drainage system. Certain pathologies can breach this barrier and secondarily involve the orbit causing significant clinical manifestations. This posterior migration of pathology also has a significant influence on the management and outcomes. The present paper will discuss the lacrimal pathologies which secondarily involve the orbit and its influence on the management and outcomes.

Keywords:

Acute dacryocystitis, dacryocystocele, dacryology, lacrimal drainage system pathology, lacrimal sac tumors

Introduction

Lacrimal sac is situated in the lacrimal sac fossa, which is a part of the medial orbital wall. However, it is situated anterior to the orbital septum and is not considered a part of the orbital tissues.[1] There are strong attachments of orbital septum to the posterior lacrimal crest, along with other barriers, which prevent the spread of lacrimal pathologies to the orbit.[2] Lacrimal fascia, posterior limb of medial canthal tendon, deeps heads of preseptal and pretarsal orbicularis are the other barriers which prevent posterior migration of pathologies.[2] However, a breach of these barriers can happen due to varied reasons, leading to the orbital involvement secondary to the lacrimal pathologies.

The present paper discusses the various lacrimal pathologies that can secondarily involve the orbit with the resultant clinical manifestations and significant influence on the management and outcomes.

Congenital Dacryocystocele with Orbital Extension [Figure 1]

Congenital dacryocystocele is a rare presentation constituting 0.1%–0.3% of congenital nasolacrimal duct obstruction cases. It presents as a bluish cystic mass in the medial canthal region with or without the presence of intranasal cysts with anatomical obstruction at the valve of Hasner and functional obstruction at the valve of Rosenmuller.[3-6] The congenital dacryocystoceles have a high rate of posterior spread of infection and hence, orbital cellulitis and sepsis are commonly encountered.[7-12] Bernardini *et al*. described four cases of orbital proptosis and dystopia due to posterior extension of the dacryocystocele into the orbit. Two cases were spontaneous, one secondary to infection leading to weakening of the thin sac wall and posterior barriers, and the fourth one was iatrogenic due to forceful compressions.[13] Carrere and Lewis also described a case of a 9-month-old infant who presented with telecanthus and dystopia of the globe due to posterior and lateral extension of the dacryocystocele.[14]

The treatment of these cases includes probing along with marsupialization of the intranasal cysts if present. Dacryocystorhinostomy is the definitive treatment in refractory cases.

Acute Dacryocystitis with Orbital Extension [Figure 2]

Acute dacryocystitis is defined as "a medical urgency which is clinically characterized by rapid onset of pain, erythema, and swelling, classically below the medial canthal tendon with or without preexisting epiphora mainly resulting from the

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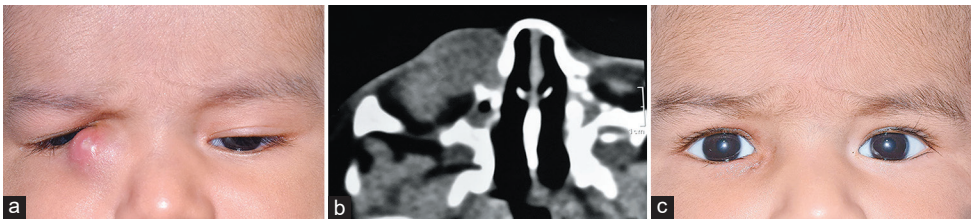

Figure 1: External photograph of a 4-month-old child with the right eye swelling in the lacrimal sac region displacing the medial canthus superiorly and telecanthus (a). Computed tomography image, axial cut, showing a heterogeneous cystic swelling on the right side extending from the lacrimal sac fossa displacing the globe temporally and extending into the anterior orbit (b). Color photograph of the child showing resolution of the swelling post probing (c)

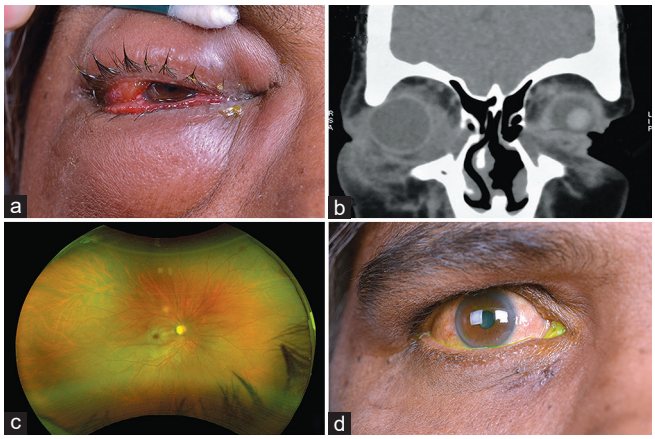

Figure 2: External photograph of the right eye showing gross preseptal edema involving the upper and lower eyelids, conjunctival congestion, and chemosis (a). Computed tomography image, coronal cut, showing right-sided lacrimal sac swelling with orbital cellulitis (b) and fundus photograph of the right eye showing central retinal artery occlusion (c). Color photograph of the right eye after endoscopic dacryocystorhinostomy (d)

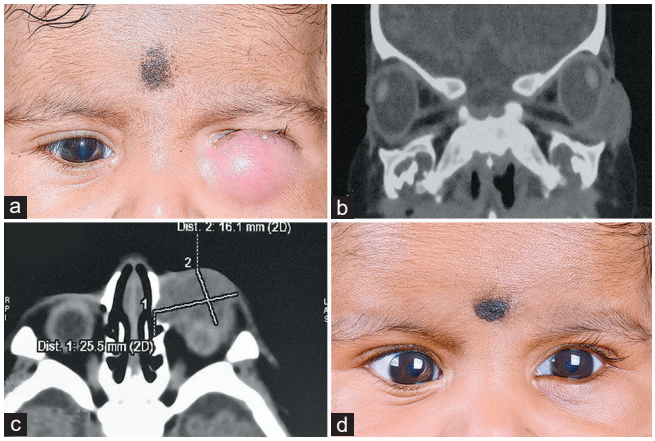

Figure 3: External photograph showing left side gross swelling in the lower eyelid with active inflammation (a). Computed tomography scan, sagittal cut, showing a well-defined heterogeneous swelling reaching the mid orbit (b), and axial cut showing well-demarcated anterolateral diverticula arising from the lacrimal sac (c). Color photograph showing resolution of the left eye after surgical correction (d)

acute infection of the lacrimal sac and perisac tissues."[1,15] Generally, acute dacryocystitis presents with preseptal cellulitis since lacrimal sac is situated anterior to the orbital septum. Posterior extension in the form of orbital cellulitis and orbital abscess can happen if the posterior barriers weaken.[2,15-27] Various theories for posterior extension of acute dacryocystitis have been suggested. Repeat episodes of acute dacryocystitis can cause distension of the soft tissues, and hence, weaken the posterior barriers, which then predisposes to orbital spread of the infection.[16] Other proposed theories include the spread of infection through the ethmoid sinus and lamina papyrecea,[17] or hematogenous spread to the orbit due to other preexisting systemic conditions in immunocompromised individuals.[16]

Intraconal extension of the abscess is another possibility. Once the posterior barriers are breached, the anterior and inferior location of the lacrimal sac predisposes to spread of infection between the medial and inferior rectus muscles directly to the intraconal space, leading to the formation of an intraconal abscess.[20,21] This can lead to a risk of deterioration of vision and even a permanent loss of vision.

Loss of vision due to acute dacryocystitis can also be associated with the development of panophthalmitis and subsequent orbital manifestations.[28] Postulated mechanisms could be a direct spread through microscopic perforations in the sclera or indirect spread through hematogenous route in an immunosuppressed patient.[28] Development of optic neuritis or vascular occlusions secondary to acute dacryocystitis can also lead to loss of vision. Compression of the optic nerve due to mass effect by the orbital abscess can lead to central retinal artery occlusion or ophthalmic artery occlusion, leading to vision loss. The orbital veins being valveless can cause venous occlusion, and loss of vision in the event of the development of any thrombophlebitis. Hence, superior ophthalmic vein thrombosis and cavernous sinus thrombosis can be rare complications of an acute dacryocystitis and would obviously present with their orbital manifestations. Direct invasion of the virulent organism, toxic vasculitis occluding fine pial feeder vessels, or mass effect on optic nerve can cause ischemia, leading to optic neuritis and vision loss.[18,27,29-32]

Lacrimal Sac Diverticula with an Orbital Extension [Figure 3]

Lacrimal sac diverticula are outpouching of the lacrimal sac and can be congenital or acquired.[33,34] These outpouchings can

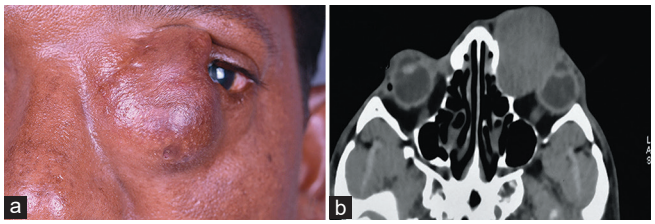

Figure 4: Solitary Fibrous Tumor of the Lacrimal Sac: External photograph of the left eye showing a gross swelling at the medial canthus with orbital extension and temporal dystopia (a). Computed tomography scan, axial image, showing a mass lesion in the bony lacrimal fossa region with orbital extension, and globe dystopia (b)

be in any direction arising from the lacrimal sac and etiologies can either be congenital weak walls, acquired infections, or posttraumatic.[35,36] Majority of the diverticula arise from the lateral wall, as the sac is covered only by the periorbital or lacrimal fascia offering least resistance to the expansion.[37-40] Anteriorly, resistance to the sac expansion is by the lacrimal fascia, medial canthal tendon, and the orbicularis muscle. Posteromedially, the bony lacrimal sac fossa acts as a deterred for the formation of diverticula toward itself.[1,41] Recurrent dacryocystitis can also cause a localized weakness of the lacrimal sac wall. Most commonly, these diverticula are seen along the inferior orbital rim but can extend anteriorly or posteriorly, sometimes also into the ethmoid sinus.[41] Besides these mechanical nature of orbital involvement by the lacrimal sac diverticula, recurrent diverticulitis can also involve orbit secondary to the development of preseptal or orbital cellulitis.

The lacrimal drainage pathway may or may not be occluded and a high degree of clinical suspicion is required to diagnose these lesions. Computed tomography (CT) scan and CT dacryocystography are tools employed for diagnosis of these lesions and their orbital extensions.[33,41]

Lacrimal Sac Tumors [Figure 4]

Lacrimal sac tumors can be classified into four categories: epithelial, lymphoproliferative, melanocytic, and mesenchymal and can be benign or malignant.[42] Epithelial sac tumors form the majority and account for 60%–94% of all the lacrimal sac tumors.[42] Of the malignant, 90% are of epithelial origin.[42]

Epithelial lacrimal sac tumors can be primary or secondary. The primary tumors are the most common, of which, squamous cell carcinoma forms the majority. Recurrence rate for these tumors is very high and ranges from 11% to 66% and mortality >50% for recurrent tumors.[43-47] Presentation is mainly as epiphora, bloody tears, and mass lesion at the medial canthus or with features of acute dacryocystitis. Extension into orbit, manifesting as proptosis, has been seen 30% of these cases, majority with squamous cell carcinoma (18%).[48]

Lymphoproliferative tumors constitute approximately 11% of lacrimal sac malignancies.[42,49,50] Of these, the most commonly seen is the diffuse large B-cell lymphoma.[50] The presentation of these tumors can vary from only epiphora to

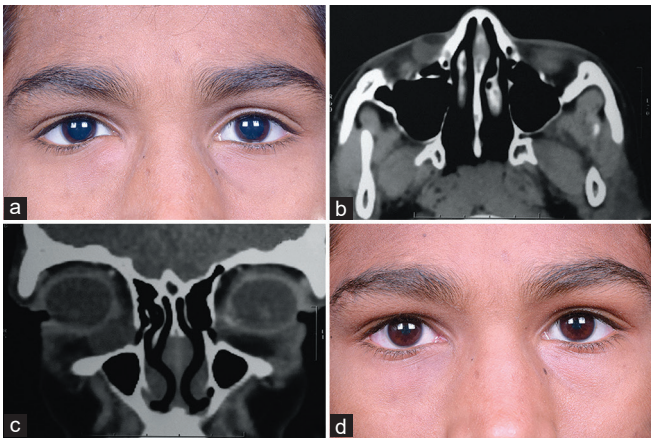

Figure 5: Dermoid cyst as a masquerader: External photograph demonstrating the right eye swelling in the lower eyelid medially (a). Computed tomography scans, axial, and coronal cuts, showing a heterogeneous swelling in the anterior orbit with air–fluid level separate from the lacrimal sac (b and c). Clinical photograph showing resolution of the swelling on the lower eyelid after surgical excision (d)

mass lesion in the lacrimal sac area. It can also present as an acute dacryocystitis following tumor infiltration of the lacrimal passages. Direct extension into the orbit and sinonasal can present with proptosis, dystopia, and motility restriction.[50] It is interesting to note that orbital extension of these lesions is seen in approximately 6% of the reported cases in the literature.[50]

Primary melanocytic tumors of lacrimal sac are uncommon and constitute 4%–5% of lacrimal sac tumors.[42] Extension to orbit mostly present in the later stages of the disease and is a very rare finding.[51-53] Mesenchymal tumors form approximately 12%–14% of lacrimal sac tumors, most of them benign, but with a malignant potential.[42] The most commonly seen tumor is fibrous histiocytoma and most of them present with orbital extension.[54] A series of solitary fibrous tumors of the lacrimal sac and nasolacrimal duct (11 cases in literature) showed orbital extension in four cases (36%).[55]

The treatment for these lesions is generally en bloc excision with complete removal of orbital and/or nasal component. This is followed by radiotherapy and/or systemic chemotherapy depending on the histopathology features and presence of metastasis.[42]

Masquerades [Figure 5]

Several nonlacrimal lesions in the anatomical zone of lacrimal drainage system may have orbital involvement, and hence, may be misdiagnosed. Inferomedial orbital swellings can masquerade as dacryocystocele or lacrimal sac swellings with or without epiphora but without much proptosis. High degree of suspicion along with clinical and radiological tests can help clinch the diagnosis. Lacrimal sac irrigation in these cases is generally patent unless there is gross mechanical compression on the lacrimal drainage passages by the orbital masses. Serial sections of the CT or magnetic resonance imaging may help distinguish these masses from the lacrimal

sac. However, if the compression is gross, it may be difficult to differentiate these lesions from the lacrimal drainage pathway. CT-dacryocystography may be helpful in resolving such dilemmas.[56]

The differential diagnosis of these lesions includes dermoid or epidermoid cysts, cavernous hemangiomas, lymphangiomas, orbital solitary fibrous tumors, encephalocele, glial heterotopia, benign fibrous histiocytoma, sinus mucoceles, hygroma, and ectopic lacrimal gland.[11,56-58] It is imperative to differentiate these conditions from lacrimal drainage disorders, as the management greatly differs.

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Conflicts of interest

There are no conflicts of interest.

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