

Case Report

Periorbital necrotizing fasciitis in a case of lid and conjunctival amyloidosis

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Received: 01 September 2020

Accepted: 16 September 2020

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ABSTRACT

Necrotizing fasciitis is a severe, progressive, rapidly spreading infection of the subcutaneous soft tissue and underlying fascia. Periorbital necrotizing fasciitis is rare owing to the excellent blood supply of the area. Ocular amyloidosis is relatively uncommon- that of the eyelid is typically associated with systemic associations whereas amyloidosis of the conjunctiva is often localised with no other associations. Authors report a case of a 40-year-old female with a 4-year history of eyelid and conjunctival amyloidosis who presented with necrotic ulcers and eschars in the upper and lower lid with purulent discharge, conjunctival mass and a dry looking cornea in the left eye. She was a known case of rheumatoid arthritis (RA) on treatment in the last 2 years. The presentation, investigations and management are described.

Keywords: Necrotizing fasciitis, Amyloidosis, Eyelid, Conjunctiva

INTRODUCTION

Periorbital necrotizing fasciitis is a progressive, rapidly spreading and destructive polymicrobial infection involving the skin, subcutaneous and deep soft tissue¹. It is commonly associated with traumatic events in patients with compromised immune systems, diabetes, chronic steroid use and alcoholism. Common organisms include *Streptococcus pyogenes* and *Staphylococcus aureus* and less commonly, facultative and anaerobic microorganisms. Patients usually present with pain, swelling, inflammation, necrosis and microvascular thrombosis.

Amyloidosis is a diverse, heterogeneous group of disorders characterized by the deposition of hyaline extracellular material into various tissues throughout the body including the eye and ocular adnexa.² Ocular involvement of amyloidosis has been reported in the adnexa, extraocular muscles, levator palpebrae muscle,

conjunctiva, cornea, lens, anterior uvea and trabecular meshwork, as well as vitreous and retina. Patients present with ocular pain, oedema, ptosis, diplopia, epiphora, mass or tissue infiltration.

Authors describe a patient with a history of localized ocular amyloidosis who presented with severe periorbital necrotizing fasciitis.

CASE REPORT

A 40-year-old female presented to the institute with complaints of swelling over the eyelids, ocular pain and purulent discharge from the left eye in the past 4 days. Furthermore, she had a 4-year history of localized ocular amyloidosis. It had begun in the left eye as a swelling in the upper eyelid and a conjunctival mass. Biopsy of the mass had revealed eosinophilic deposits with positive congo red staining and green birefringence on polarized light, confirming it as amyloid. This amyloid was

consistent with AL amyloidosis (primary amyloidosis). Local ultrasonography had revealed a phlebolith with multiple calcific foci on the medial aspect of upper eyelid of left eye. MRI of the orbit had revealed left optic neuritis, early changes of meningitis, inflammatory changes in the periorbital soft tissues with apparently normal appearing eyeballs. The patient was then referred to an onco haematologist for evaluation of systemic amyloidosis. But hemogram, renal and liver function tests, 2D echocardiogram, ECG, CT scan of chest and abdomen were not suggestive of systemic amyloidosis. Patient was on conservative management since then. She was also a known case of RA on oral prednisolone and hydroxychloroquine for 2 years.



Figure 1: Gross appearance on presentation.



Figure 2: Periorbital oedema and erythema with necrosis and eschar.



Figure 3: Anterior segment of conjunctival mass, dry lustreless cornea with copious purulent discharge.

On physical examination, there was widespread left sided facial oedema extending up to the neck (Figure 1). The left periorbital area was oedematous and erythematous with periocular skin necrosis and eschar (Figure 2). The upper lid tissue was extremely friable with copious purulent discharge in the left eye. Owing to the significant skin and tissue changes, the affected eye could not be assessed for visual acuity- only finger counting close to face. In anterior segment examination, there was chemosis and a conjunctival mass with papules which bled easily. The cornea was dry, hazy and lustreless (Figure 3). Details of the iris, pupil and lens were limited because of corneal haze. There was no view of the fundus and eye ball movements were restricted in all directions. The other eye was normal, with visual acuity of 6/6 and normal fundus examination. She was started on topical lubricants and antibiotics.

MRI revealed large multilobulated lesion in left pre-orbital region extending into retro-orbital region extraconal in lateral part, abutting anterior part of lateral rectus muscle and left lacrimal gland and also into the temporal region. (Figure 4 and 5). No significant diagnostic intracranial abnormality was seen. Blood investigations showed leucocytosis, raised ESR and CRP. Steroids were discontinued on physician's advice. After surgical opinion, pus sample was sent for culture and antibiotic sensitivity which was positive for methicillin-resistant *staphylococcus aureus* (MRSA) sensitive to vancomycin. She was started on injectable vancomycin and local antiseptic ointment. Debridement of devitalized tissue was done. After 5 days, injectable meropenem was added. Following this, she showed local improvement; the purulent discharge and skin erythema gradually reduced over the next 4-5 days. In spite of this, visual loss was permanent. She was later referred to a higher center for oculoplastic surgery after control of ongoing active infection.

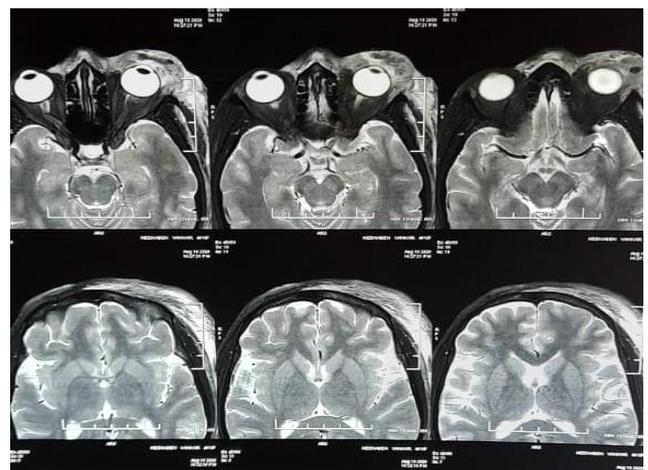


Figure 4: Transverse section of periorbital involvement- soft tissue oedema and thickening with normal globe.

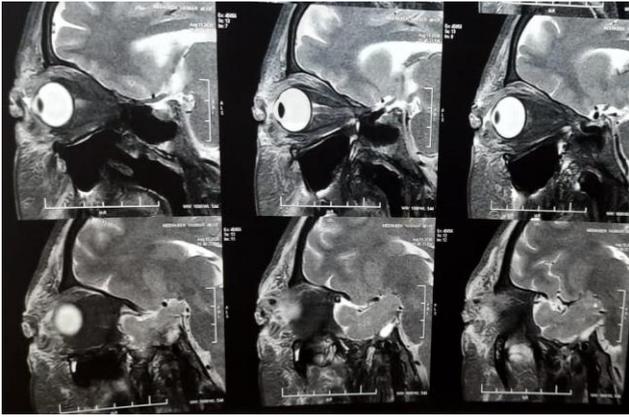


Figure 5: Sagittal section of gross periorbital soft-tissue infiltration with retro-orbital extension.

DISCUSSION

Although necrotizing fasciitis occurs in the face and neck, isolated periorbital necrotizing fasciitis is relatively uncommon.³ Common organisms associated include Group A beta-haemolytic *Streptococcus pyogenes* (50% cases). Others include *Staphylococcus aureus*, *Pseudomonas*, *Staphylococcus agalactiae*, *Streptococcus viridians*, *Staphylococcus epidermidis*, *Propionibacterium acnes*, *Clostridia* etc. Most common inciting factor for periorbital necrotizing fasciitis is penetrating or surgical trauma. It is common in patients with immunosuppression, diabetes, chronic steroid use and alcoholism.⁴ Risk of necrotizing fasciitis is high in patients with comorbidities and diseases like chronic renal failure, peripheral vascular disease, rheumatological disease and systemic malignancy. Especially, steroids and chemotherapeutic agents used in above conditions may increase the incidence.

Necrotizing fasciitis of the eyelids behaves differently from elsewhere in the body. Eyelid skin is thin and lacks subcutaneous tissue. Rich blood supply of the orbicularis acts as a barrier between the skin and the underlying periorbita, preventing the spread of infection to the orbit. The dermis is attached firmly at the nasojugal fold medially and to the malar fold laterally which prevents spread of inflammation. Once this barrier is broken, the infection spreads via fascial planes to the neck, increasing mortality. Spread of inflammation over the nasal bridge to the contralateral eyelids results in bilateral disease.⁴

Prominent cutaneous changes of severe inflammation include vesicle formation, hyperaemia, and a reddish or purplish hue.³ Presence of red, tense and swollen skin, leucocytosis and tissue emphysema strongly raises clinical suspicion.⁵ Contrast CT is the investigation of choice- findings include increased soft-tissue opacity and thickness. MRI is preferred for soft-tissue extension.⁶

Early high-dose antibiotics with prompt surgical debridement is associated with lower mortality. Mild

cases may respond to antibiotic therapy alone.⁷ Standard antimicrobial therapy should consist of a combination of beta-lactam antibiotics, such as penicillin or cephalosporin and clindamycin. If not diagnosed and treated rapidly, necrosis of the orbicularis muscles, post septal tissues, and anterior fat of the orbit can occur.⁸

Amyloidosis is characterised by misfolded proteins deposited within extracellular space in various tissues and organs, including the orbit. Amyloidosis is categorized as localized or systemic. Classic forms are light-chain (AL) and amyloid-A protein (AA) amyloidosis. Most common form of local amyloidosis is caused by deposition of monoclonal immunoglobulin light chains or by a usually benign B-cell or plasma-cell clone.⁹

Ophthalmologic involvement in amyloidosis is very rare. Signs include eyelid malposition, globe displacement, abnormal eyelid motility, diplopia, ptosis, subconjunctival haemorrhage, conjunctival mass, corneal opacity, increased intraocular pressure, vitreous floaters, and abnormal fundal appearance. Patient may initially present with a mass of unknown origin. Amyloidosis of the conjunctiva and eyelid is typically benign. When confined to the conjunctiva the amyloid tends to be localised, whereas cutaneous lesions are associated with systemic disease.¹⁰ The patient showed evidence of both lid and conjunctival amyloid but no systemic associations.

CT and MRI are important in localizing involved orbital structures. Final diagnosis of ocular amyloidosis requires tissue biopsy that shows red-green dichroism with Congo red staining in polarized light. Systemic examination and investigations are required to rule out systemic amyloid and neoplastic plasma-cell disease.¹¹

Management varies depending on symptoms and visual impairment by amyloid protein deposition. For eyelid amyloidosis, surgical approach alone may be associated with a high risk of incomplete excision and local recurrence. Demirci et al reported a 27% recurrence rate after local surgical debulking.¹² Radical excision may cause both structural and functional impairment of eyelid. Conjunctival lesions are often conservatively managed by lubrication with artificial tears. If recalcitrant, local excision or surgical debulking may be performed.¹²

Ocular manifestations of RA include scleritis, episcleritis, keratoconjunctivitis sicca, keratitis and less commonly chorooiditis, retinal vasculitis, episcleral nodules, retinal detachment and macular oedema.¹³ Periorbital necrotizing fasciitis and ocular manifestations of amyloidosis are seldom associated with RA.

The patient was on steroids for RA for 2 years, thus immunocompromised, which might have precipitated necrotizing fasciitis. Injectable vancomycin and meropenem were started for MRSA. Regular antiseptic dressing and frequent debridement of necrotic tissue was

done. Soft tissue features gradually improved with treatment but vision could not be salvaged. Patient was referred to higher center for oculoplastic management.

CONCLUSION

Most of the time, periocular and orbital amyloidosis occur as a primary localized disease, which is treated by surgical debulking and observation. Simultaneous presence of both periorbital necrotizing fasciitis and ocular amyloidosis, both of which are very uncommon, signifies compounded overall risk, especially in patients with altered immune systems. Advanced necrotizing fasciitis can cause extensive periorbital tissue destruction and can spread to the neck which is potentially life threatening. Ocular amyloidosis can cause permanent vision threatening complications as well as cosmetic deformities. Hence, thorough history taking, examination, early diagnosis and prompt treatment is paramount in the management of these conditions.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Das AD, Gore AS, Vasava IG, Anurag S. Yadav. Periorbital necrotizing fasciitis in a case of lid and conjunctival amyloidosis. *Int J Res Med Sci* 2020;8:3720-3.