Annular elastolytic giant cell granuloma of conjunctiva: A case report

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Annular elastolytic giant cell granuloma is a condition characterized histologically by damaged elastic fibers associated with preponderance of giant cells along with absence of necrobiosis, lipid, mucin, and pallisading granuloma. It usually occurs on sun-damaged skin and hence the previous name actinic granuloma. A similar process occurs on the conjunctiva. Over the past three decades only four cases of conjunctival actinic granuloma have been documented. All the previous patients were females with lesions in nasal or temporal bulbar conjunctiva varying 2-3 mm in size. We report a male patient aged 70 years presenting with a 14 mm × 7 mm fleshy mass on right lower bulbar conjunctiva. Clinical differential diagnoses were lymphoma, squamous cell carcinoma *in situ* and amyloidosis. Surgical excision followed by histopathology confirmed it to be a case of actinic granuloma. This is the first case of isolated conjunctival actinic granuloma of such a large size reported from India.

Key words: Actinic granuloma, conjunctiva, granuloma annulare, necrobiosis lipoidica

Annular elastolytic giant cell granuloma, as the name implies, is a giant cell rich granulomatous inflammation in response to elastolysis or damage to elastic tissue fibers. It almost always occurs as cutaneous lesions in patients with sun damaged skin. Though a possibly related process can occur in the conjunctiva, very few cases have been reported so far. Over the past three decades only four cases of conjunctival actinic granuloma have been documented.

Case Report

A 70-year-old male patient presented with painless, gradually increasing swelling in the right lower bulbar conjunctiva for 6 months followed by redness and watering for last 3 weeks.

Ocular examination revealed a 14 mm × 7 mm red fleshy mass in the lower bulbar conjunctiva in the right eye [Fig. 1]. Vision in the right eye was 20/100 with early cataractous change. The left eye was pseudophakic and had a total retinal detachment with only PL. The patient had no significant past medical history.

The conjunctival mass was clinically diagnosed to be a squamous cell carcinoma *in situ*, lymphoma or amyloidosis. Excision biopsy followed by histopathological examination was done. Histopathology revealed a localized granulomatous inflammation with histiocytes around a homogeneous material along with giant cells and chronic inflammatory cells [Figs. 2 and 3]. Van Gieson and Congo red stains were performed. Congo red stain was negative. Van Gieson stain demonstrated the complete absence of elastic tissue at the center of the granuloma [Fig. 4].

The patient was followed up for 2 years. There was no further recurrence of the disease.

Discussion

Annular elastolytic giant cell granuloma is a condition characterized histologically by damaged elastic fibers surrounded by numerous giant cells and absence of necrobiosis, lipid, mucin, and pallisading of the granuloma.[1] It almost always occurs on sun exposed skin, such as face, neck, dorsum of hand, forearm, and arm and hence the previous name actinic granuloma; however there are few reports occurring in sun-protected sites. A possibly related process occurs on the conjunctiva.[2] The term actinic granuloma was coined by O Brien in 1975, who described similar histological features in cutaneous lesions of patients with sun-damaged skin.[3] He described the pathogenesis as an attempt to repair the damaged connective tissue. Subsequently this concept was disputed by Ragaz and Ackerman,[4] who believed that actinic granuloma was a variant of granuloma annulare.[5,6] McGrae postulated that actinic granuloma represented a cell-mediated immune response to actinically altered elastotic fibers with a preponderance of helper T cells in the lymphocytic infiltrate.[1]

The occurrence of conjunctival actinic granuloma in isolation is a rare entity. In the past four decades only four cases have been

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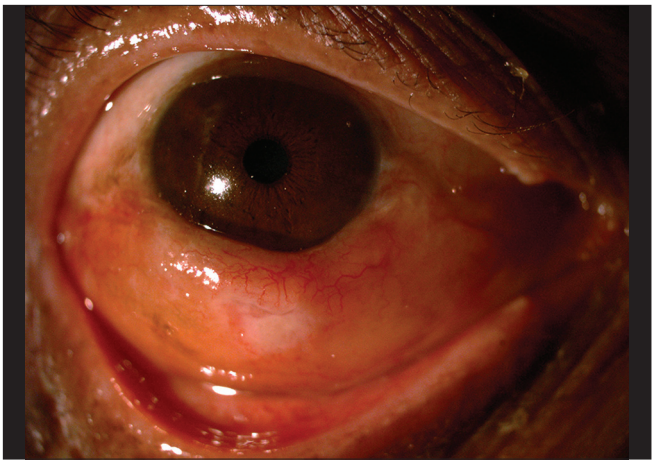

Figure 1: Clinical photograph showing 14 mm × 7 mm red fleshy mass in right lower bulbar conjunctiva

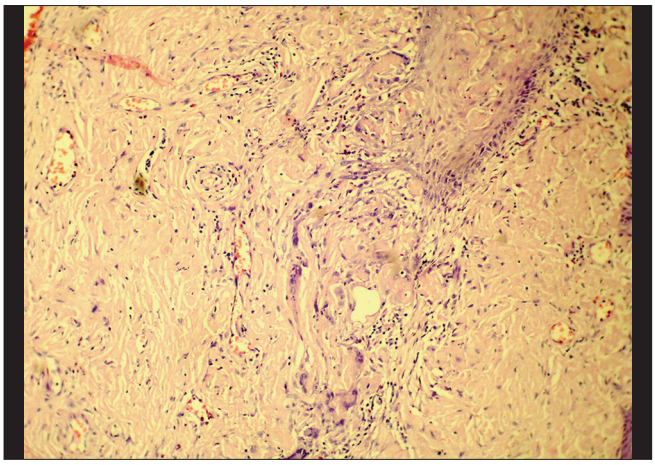

Figure 2: Microphotograph showing subepithelial granulomatous reaction in right conjunctiva. H and E stain ×100

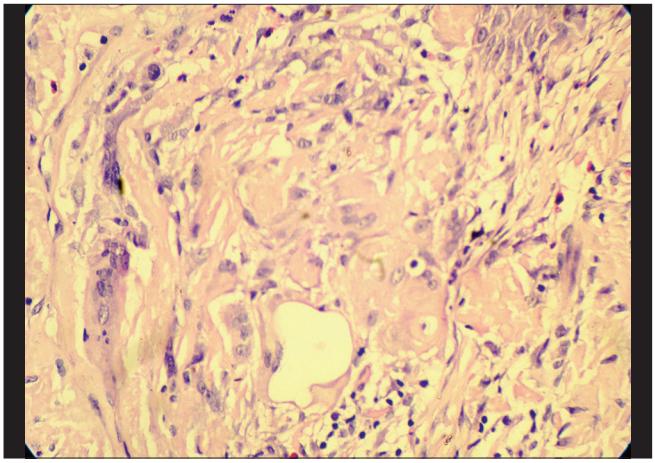

Figure 3: Microphotograph showing giant cells, inflammatory cells, and histiocytes in the granuloma. [H and E stain ×400]

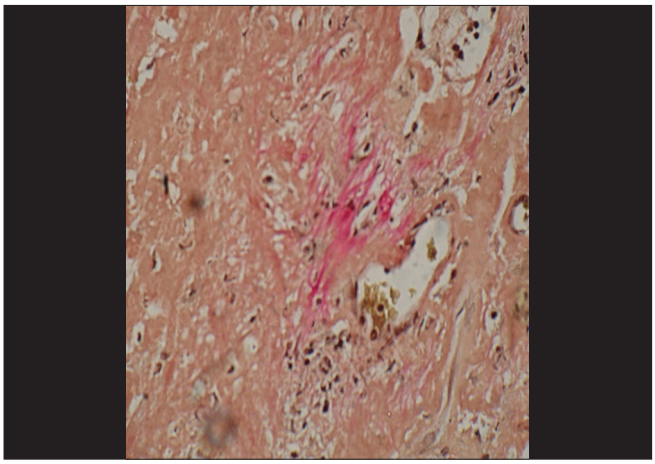

Figure 4: Microphotograph showing the absence of elastic tissue in the centre of granuloma. Van Geison stain ×400

documented.[7] All the previous cases were females, our case being the first such lesion occurring in a male patient. The size of all the previous lesions varied between 2 and 3 mm occurring in nasal or temporal bulbar conjunctiva. In those cases the clinical differential diagnoses were pinguicula, pinguiculitis, Bowens disease, and conjunctival nevus. In our case the size of the lesion was much larger. 14 mm × 7 mm, a fleshy mass involving the whole of the lower bulbar conjunctiva. The clinical differential diagnoses were squamous cell carcinoma *in situ*, mucosa associated lymphoid tumor (MALTOMA), amyloidosis, and leukemic deposit.

The hematological investigations of the patient were within normal limits.

Surgical biopsy followed by histopathology demonstrated a granulomatous reaction in the subepithelial lamina propria. A large number of giant cells along with lymphoplasmacytic infiltration were seen. There was elastotic degeneration of the connective tissue. There was no evidence of any neoplastic change or amyloid deposition.

Histological differential diagnoses were infectious and noninfectious granulomatous reaction. Among the infectious agents mycobacterial, fungal, and parasitic lesions can occur. The absence of caseous necrosis excludes tuberculosis and the prominent eosinophilic response invited by fungal and parasitic lesions was not present in this case.

The noninfectious granulomatous inflammation includes foreign-body giant cell reaction, granuloma annulare or pseudoheumatoid nodule, and necrobiosis lipoidica. No foreign body could be detected in the lesion. Granuloma annulare presents with abundant mucin, partial loss of elastic fibers and radial arrangement of epithelioid histiocytes (pallisading granuloma). In Necrobiosis lipoidica central elastolysis is absent.

The complete loss of elastic tissue in the central zone as documented by connective tissue stain is used as the primary basis for separating annular elastolytic giant cell granuloma from granuloma annulare and necrobiosis lipoidica.

Thus actinic granuloma of conjunctiva is a distinct clinical, histological, and immunological lesion. Though rare, the clinician should consider the possibility of actinic granuloma presenting as a fish flesh mass in the conjunctiva and pathologist should consider the possibility of granulomatous inflammation in association with elastolysis and does not necessarily imply the presence of foreign bodies or infection.

Regarding medical treatment Hydroxychloroquine, Clofazimine, Dapsone, intralesional, and systemic steroid has been used in annular elastolytic giant cell granuloma (AEGG) of skin.[8,9] No trial has been documented for conjunctival lesion. Topical steroid had little effect.[7] Surgical excision is considered to be the treatment of choice.

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