http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v10i11.22

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

# Eosinophilic Angiocentric Fibrosis in Upper Eyelid Conjunctiva: A Case Report

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**Abstract**
*Eosinophilic angiocentric fibrosis is a rare lesion of head and neck region included in the spectrum of Immunoglobulin G4 (IgG4) related disease. It has characteristic histologic findings of onion skin fibrosis and prominent eosinophilia. In the present case, the subject is a 45 year old female presenting with a painless mass in right upper lid of five years duration.*
**Keywords:** *Eosinophilic angiocentric fibrosis (EAF) IgG4 related disease conjunctiva.*

## Introduction

Eosinophilic angiocentric fibrosis (EAF) is a relatively rare disease with an indolent course. It is represented by fibro inflammatory lesions of unknown origin.[1]It usually affects the nasal cavity, paranasal sinuses and orbit. Very rarely, it can affect the lacrimal gland, respiratory tract, retro orbit, and gingiva.[2] Till date, approximately 79 cases of EAF have been reported in the scientific literature worldwide, including 61 in the nasal cavity, 23 in the paranasal sinuses, 13 in the orbit, 4 in the respiratory tract, 3 in the lacrimal gland, 1case each in the gingiva and retro orbital location. Only one case depicting bilateral eyelid conjunctival involvement has been recently published.[2]

Herewith we present a case of a 45 years old female who presented with a painless mass in the right upper lid.

## Case Report

A patient aged 45 years female presented with a mass in the right upper eyelid conjunctiva of five years duration. The patient had a history of allergic sinusitis. The results of routine blood investigations were within normal limits except for serum IgG4 levels, which were raised. Other screening tests for autoimmune diseases were negative. Biopsy examination revealed marked chronic inflammation and stromal fibrosis. The inflammatory infiltrate comprised of mature lymphocytes, plasma cells and eosinophils. The stromal fibrosis showed concentric lamellar deposits of collagen and focal storiform fibrosis. There was an absence of lymphoid follicle formation or granuloma formation. Immunohistochemical staining for IgG and IgG4 was positive, which further supported the diagnosis of EAF.

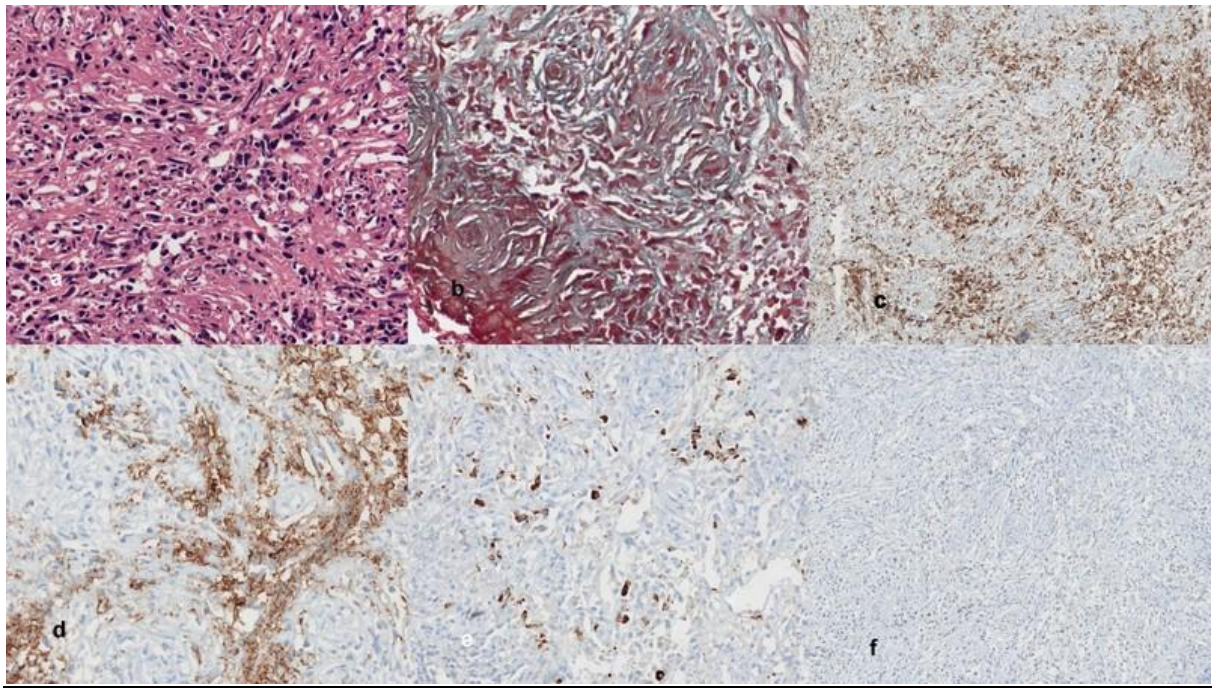


**Figure 1**

a) Photomicrograph showing dense lymphoplasmacytic infiltrate and stromal fibrosis.(Hematoxylin and Eosin stain; 400X magnification)
b) Photomicrograph showing characteristic perivascular whirling fibrosis. (Masson Trichrome stain; 400X magnification)
c) CD 45 IHC X 200 X magnification: Photomicrograph showing dense lymphocytic infiltration.(CD 45 IHC; 200 X magnification)
d) IgG IHC X 400X magnification: Photomicrograph showing large number of IgG positive plasma cells.(IgG IHC; 400X magnification)
e) Photomicrograph showing increased number of IgG4 plasma cells.(IgG4 IHC; 400X magnification)
f) Photomicrograph showing negative staining. (Pancytokeratin IHC; 200X magnification)

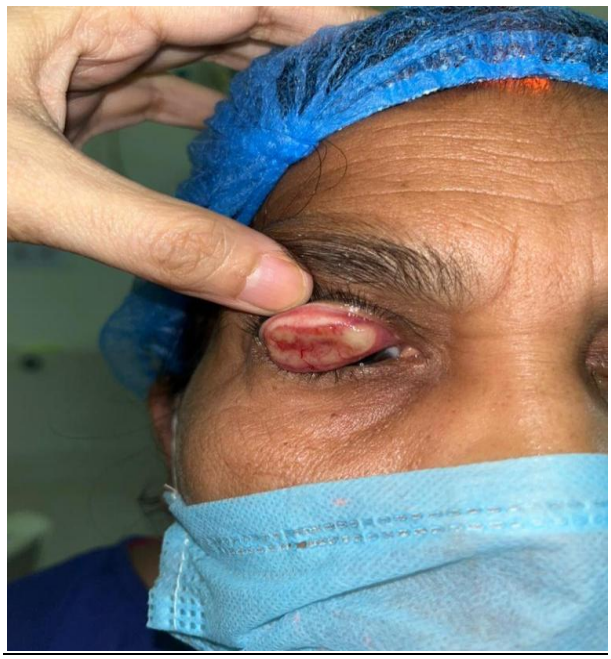

**Figure 2:** Colour photograph of the patient showing a mass lesion in the right upper eyelid conjunctiva.

**Discussion**

EAF is a very rare condition of unknown etiology. It was first described by Holmes and Panje in 1983 as "intranasal granuloma faciale".[3]In 1985, Roberts and McCann reported three similar cases and coined the histologically descriptive name.[4]The pathogenesis of EAF is unclear and has been attributed to surgical manipulation, trauma and hypersensitivity.[5]Recently, Deshpande et al suggested that EAF belongs to the spectrum of IgG4-related diseases, which are characterised by tumefactive lesions, increased number of IgG4 positive plasma cells and increased IgG4 to IgG positive plasma cell ratio.[6]

The pathogenesis of IgG4 related diseases is not fully understood, but there is increasing evidence that the interaction of clonal cytotoxic T cells and activated B cell subsets plays an important role in

the release of profibrotic cytokines.[7]The exact role of eosinophils in EAF, otherwise seen in a minority of IgG4- related diseases is also unclear. Tissue eosinophilia, classically due to immune mediated hypersensitivity and recruitment by IL-5, is known to produce cytotoxic inflammatory mediators leading to stimulation of fibroblasts.[7]

The characteristic histologic findings are the basis for establishing the diagnosis. According to Roberts and McCann, the disease demonstrates a spectrum of histologic findings depending upon the the stage of the disease.[8.9]Both patterns can be seen in the same biopsy, suggesting a continually evolving process. Eosinophilic vasculitis without fibrinoid necrosis is common in early lesions occurring in a patchy manner and focally involving capillaries and venules. Late lesions show dense perivascular onion skin whirling fibrosis with decreasing inflammatory cell infiltration.[10]

The differential diagnosis of EAF includes Wegener's granulomatosis, Churg Strauss Syndrome, granuloma faciale and infections.[11]

EAF is a progressive yet indolent benign disorder without reported malignant transformation. Till date, no previous case of EAF related mortality is reported.[11]No definitive treatment has been identified, however, most of the cases are treated with surgical resection alone or in combination with medical therapy.[12-14] EAF is a relatively less known entity. As a clinicopathologic diagnosis, it is important for pathologists to be aware of this condition in order to direct clinical workup of IgG4- related diseases for better patient management.

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