

EASTERN OPHTHALMIC PATHOLOGY SOCIETY MEETING-WASHINGTON DC, SEPTEMBER 2018.

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Material Submitted: Pre-meeting PowerPoint circulated as not enough material.

Title: Bilateral lacrimal sac inflammatory polyps with allergic mucin.

Clinical History: A 57 y old man presented to the ENT surgeons with nasal stuffiness and a runny nose of a few years duration. This was associated with swellings in both medial canthal areas, larger on the left side compared to the right. The swellings according to his community caregiver had been present for 3 years and getting progressively larger. A CT-scan confirmed the presence of the chronic sinus disease with grade 3 nasal polyposis associated with bilateral obstruction of the nasolacrimal ducts. There were well-defined bilateral canthal swellings-2.5 on the left side and a 2cm on the right side. The left swelling appeared to be bi-lobed and extending into the anterior orbit and subcutaneous tissue.

The patient underwent nasal polypectomy and the histology of this showed typical inflammatory nasal polyps. 4 months later he underwent bilateral endoscopic DCRs for suspected bilateral lacrimal sac mucoceles and to exclude tumour and fungal infection. Biopsies of both lacrimal sacs were carried out with placement of drainage tubes. Swabs taken at the time of the surgery grew E.coli from the right side and group B streptococcus from the left side. 4 months post-surgery, the canthal swellings have much improved but the DCR tubes have extruded and there appears to be re-development of the swelling on the left side. Further imaging is planned with view to carrying out further surgery. All fungal cultures have been negative to date.

Ophthalmic Pathology:

Macroscopic: From the left lacrimal sac was received yellow/greyish gelatinous tissue with a rubbery feel, 9x8x6 mm. From the right side was receive gelatinous greyish material 5x3x3 mm

Microscopy:

The biopsy from the left side showed chronically inflamed polypoid pieces of mucosal tissue that were focally lined by ciliated epithelium. The stromal tissue was oedematous and congested and infiltrated by chronic inflammatory cells inclusive of eosinophils. Accompanying the tissue was layered mucin containing tiered colonies of dense eosinophils with Charcot-Leyden crystals. No fungi were identified. Gram positive cocci were present in the specimen. The right lacrimal sac biopsy was identical to the left side except no mucin or bacterial colonies were present. ***The appearances were those of bilateral lacrimal sac inflammatory polyps with allergic mucin.***

Discussion:

Typical inflammatory nasal polyps are the most common type and are due to recurrent attacks of rhinitis (allergic, inflammatory). They affect 30 year olds upwards and are often associated with asthma, chronic rhinitis and aspirin intolerance. These polyps can involve the nasal cavity and paranasal sinuses. Histologically they show respiratory epithelium, often with squamous metaplasia, oedematous and loose stroma with hyperplastic mucous glands and an inflammatory infiltrate, comprising lymphocytes, plasma cells, eosinophils, neutrophils and mast cells. The mucosa may be ulcerated or infected, the basal membrane may be thickened, there may be bizarre stromal cells (large and pleomorphic) due to reactive changes and there may be a prominent glandular¹ component. Seeing nasal-type inflammatory polyps in both lacrimal sacs is highly unusual.

These inflammatory polyps were associated with allergic mucin. Allergic mucin is thick, highly viscous, and said to be similar to peanut butter or mechanical grease. Microscopically, the mucin often takes on a chondroid lamellated appearance. Trapped between the mucin lamellae are sheets of eosinophils, frequently with the presence of eosinophilic breakdown products and/or Charcot-Leyden crystals that can easily be seen with H&E staining. Allergic mucin is one of the features that are found in allergic fungal rhinosinusitis² (AFS). In 1994, Bent and Kuhn published their major diagnostic criteria centered on the histologic, radiographic, and immunologic characteristics of the disease, of which allergic mucin is one of the major criterion for diagnosing AFS³. Allergic fungal sinusitis affects immunocompetent individuals from the third to fifth decades of life. Allergic fungal sinusitis is usually non-invasive and follows a benign course. Patients present with a history of chronic allergy, asthma, nasal polyps, and sinusitis, which is often refractory to medical therapy². Chest radiographs are often normal, but CT scans of the nasal sinuses reveal areas of high attenuation and increased density. Bony erosion is rare, but remodelling can occur. Characteristic allergic mucin is seen in histologic specimens and serves to remind the pathologist to be aware of and look for fungal organisms. Following surgical removal and diagnosis, low-dose corticosteroid therapy and, in some cases, immunotherapies have been used as long-term adjunctive treatments with varying success. The immunologic mechanism behind allergic fungal sinusitis has been thought to be a combined type I and type III immune hypersensitivity response, mainly because of the similarity to mucoid impaction of the bronchus and acute bronchopulmonary fungal disease².

There have been 3 documented cases of AFS involving the lacrimal sac. These are summarised below in the table:

Authors	Clinical features	Histological features
4. Facer ML, Ponikau JU, Sherris DA. Eosinophilic fungal rhinosinusitis of the lacrimal sac. <i>Laryngoscope</i> 2003;113:210–4.	69-year-old male with a history of nasal polyps and prior sinus surgery presenting with a cystic lesion in the right medial canthal region.	Findings consistent with AFS with positive staining for fungal hyphae on Chitinase immunofluorescence stains.
5. Kim C, Kacker A, Chee RI, Lelli GJ. Allergic fungal sinusitis causing nasolacrimal duct obstruction. <i>Orbit</i> 2013; 32:143–145.	54-year-old female with a history of asthma and nasal polyps presenting with left dacryocystitis unresponsive to topical and systemic antibiotics	Marked eosinophilia “highly suggestive of allergic fungal sinusitis’ ‘However, GMS stain for fungal elements was negative.
6. Pao KY, Yakopson V, Flanagan JC, Eagle RC Jr. Allergic fungal sinusitis involving the lacrimal sac: a case report and review. <i>Orbit</i> . 2014;33(4):311-3	70-year- male with tearing from the left eye for 6 weeks. The patient noted that a “bump” developed overlying his left lacrimal sac one-month prior. History of seasonal allergies, nasal polyps and right nasolacrimal duct obstruction. Previous uncomplicated right external DCR with Crawford silicone stent placement and an uncomplicated nasal polypectomy 3 years prior to presentation	Sheets of amphophilic allergic mucin containing laminated aggregates of eosinophils in various stages of degeneration, multiple Charcot-Leyden crystals and fungal hyphae confirmed by GMS stain . The fungi were found within the mucin and did not invade the surrounding tissue.

As can be seen from the table, AFS with lacrimal sac involvement typically have a history of chronic rhinosinusitis, allergic rhinitis atopy and nasal polyps. The presentation of AFS involving the lacrimal sac and lacrimal sac tumors are similar and may cause epiphora, proptosis, blepharoptosis, ophthalmoplegia, recurrent dacryocystitis and a mass in the medial canthal area. However, AFS patients lack bloody discharge and lymphadenopathy and don't usually present with a firm mass that extends above the medial canthal tendon

Facer and colleagues postulated that distal obstruction, often due to nasal polyposis and distension of the lacrimal sac leads to chronic reflux of eosinophilic mucin into the nasolacrimal system⁴.

In our case, the mucin did not contain fungal elements. Reports of AFS have described cases with allergic mucin but without concomitant fungus⁷. The absence of such organisms in otherwise typical allergic mucin brings into question the role of fungi in allergic fungal sinusitis. A study performed by Lara and Gomez⁷ showed that the clinical presentation and radiographic findings of AFS were similar in groups with and without fungi. Incidence, age, and gender distribution were similar to data reported previously. However, the amount of allergic mucin was much greater in the group with fungus than in the group without fungus. They concluded that the presence of allergic mucin is not unique to allergic fungal sinusitis, but rather is the result of a process that could have other aetiologies⁷. While not always causative to the disease, the fungus continues to fuel the process and is likely an entrapped bystander. They suggested that allergic fungal sinusitis was more appropriately termed allergic mucinous sinusitis or eosinophilic mucinous rhinosinusitis given the fungal negative cases⁷.

References:

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3. Bent JP 3rd, Kuhn FA. Diagnosis of allergic fungal sinusitis. *Otolaryngol Head Neck Surg*. 1994;111(5):580-588.
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7. Lara JF, Gomez JD. Allergic mucin with and without fungus: a comparative clinicopathologic analysis *Arch Pathol Lab Med*. 2001 Nov;125(11):1442-7.