Eosinophilic angiocentric fibrosis of the sinonasal tract: a case report and review of the literature

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Abstract

Eosinophilic angiocentric fibrosis (EAF) is a rare chronic benign disorder of unknown etiology and is characterized by submucosal thickening and fibrosis in the upper respiratory tract. In this report, we describe a case of EAF in the nasal cavity of a woman who underwent elective surgery for division of adhesions and has had no recurrence during 2 years of postoperative follow-up. A review of the literature on the clinical manifestations of EAF, sites of lesions, management, and outcomes identified 48 articles that included 72 cases. A summary of these reports is presented, including our present case. The most common anatomic site involved was the nose (77.8%), the most common manifestation was nasal obstruction (66.7%), and the most common treatment modality was surgical resection (83.3%). After surgery, 36% of patients remained free of EAF. The most common pharmacologic agent used was a corticosteroid (38.9%).

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Introduction

Eosinophilic angiocentric fibrosis (EAF) is a non-malignant inflammatory disease that reportedly can affect almost any area of the face, including the nose,[1] orbits, eyelids,[2] larynx,[3] and subglottis.[4] There have also been occasional reports of lower respiratory tract involvement with airway narrowing.[5,6] In 2011, Deshpande et al classified EAF as part of the spectrum of immunoglobulin (Ig)G4-related systemic diseases and observed that it was characterized by a tendency to form tumefactive lesions in the affected organ.[5]

The term "eosinophilic angiocentric fibrosis" was coined in 1985 when Roberts and McCann published their initial description of three cases.[7] They described EAF as a mucosal lesion that is accompanied by thickening of the submucosal connective tissues. The disease starts as a focal perivascular subepithelial exudate of eosinophils with a collection of plasma cells and lymphocytes without fibrinoid necrosis. These foci form widespread areas of perivascular fibrosis with marked angiocentric rotation. This process leads to thickening and turbidity of the mucosa, which becomes adherent to the underlying structures. As the fibrosis progresses, the lymphoblastic component recedes but the eosinophils remain. The fibrosis does not resolve, and the resulting stenosis requires surgical resection. Noting that these lesions occurred in the upper respiratory tract, Roberts and McCann described this disorder as "upper respiratory eosinophilic angiocentric fibrosis" to cover its main features.

The histopathologic characteristics of EAF include a dense fibrotic stroma with a perivascular "onion skin-like" whorling pattern and a dense inflammatory infiltrate consisting of lymphocytes, plasma cells, eosinophils, and some neutrophils. Modest acute neutrophilic inflammation with focal endothelial proliferation,[1] a collagen bundle winding around the vessels in an onion-skin pattern, and eosinophils, lymphocytes, and plasma cells[2] may be present.

This report describes a case of EAF in the nasal cavity in a woman who underwent elective surgery for division of adhesions and has had no recurrence during 2 years of postoperative follow-up. a review of the literature on the clinical manifestations of EAF, sites of lesions, management, and outcomes is presented. This report provides an opportunity to discuss clinically relevant issues and raise awareness of EAF as a disease entity.

Case report

A woman in her early 60 s was referred to our center with a complaint of nasal obstruction and postnasal drip. She had a history of hypertension, for which she was on treatment. Four years earlier, she had undergone antrostomy for nasal obstruction. At that time, the histopathologic diagnosis was mild mucositis, and she made a symptomatic recovery. A recent clinical examination had revealed obstruction of the right nares and deviation of the nasal septum to the right. Further clinical examinations, including a complete blood count, C-reactive protein, antinuclear autoantibodies, antineutrophil cytoplasmic antibodies (ANCA), and peripheral antineutrophil cytoplasmic antibodies (p-ANCA), were in the normal range (Table 1). A chest radiograph was normal. Magnetic resonance imaging (MRI) of the paranasal sinuses revealed relative thickening of the anterior portion of the nasal septum extending anterolaterally and involvement of the nasal cartilages with extension up to the subcutaneous tissue (measuring approximately 2.6×1.8 cm) (Figure 1). Fungal granuloma was suggested as a differential diagnosis on MRI. She underwent elective surgery for division of adhesions.

Table 1. Results of blood and tissue examinations in a patient with eosinophilic angiocentric fibrosis.

Parameter*	Result	Unit	Reference range
Blood glucose	5	mmol/L	3.89–6.11
Urea	1.24	mmol/L	0.54–1.60
Creatinine	1	mg/dL	0.5–1.5
Sodium	154	mmol/L	135–147
Potassium	4.9	mmol/L	3.5–5
Phosphorus	1.29	mg/dL	0.81–1.62
LDH	470	U/L	240–480
RBC	4.44	$\times 10^6/\mu L$	3.53–4.66
WBC	3.3	$\times 10^3/\mu L$	3.0–7.8
Hemoglobin	114	g/L	120–170
Hematocrit	38.9	mL/12 hours	36–53
Platelets	151	$\times 10^3/\mu L$	150–450
ESR 1st hour	41	mm/hour	<20
C-reactive protein	0.09	mg/dL	<0.3
PT	13	s	11–13
PTT	31	s	19–35
T4	54.6	ng/mL	45–120
TSH	2.39	µIU/mL	0.3–5
Tuberculin test	16	mm	<5
ANA	5	U/mL	0–10
C-ANCA	1.70	U/mL	<10
P-ANCA	1.80	U/mL	<10
ACE	37	IU/L	8–72
Acid-fast staining	Negative	–	–
CD68 (IHC)	Positive in background histocytes	–	–
CD34 (IHC)	Negative	–	–
CD1a (IHC)	Negative	–	–
CD99 (IHC)	Negative	–	–
BCl-2 (IHC)	Positive	–	–
B-catenin (IHC)	Negative	–	–
S100 (IHC)	Negative	–	–
Ki-67 (IHC)	Positive in 3%–5%	–	–
Vimentin (IHC)	Positive	–	–

*Some values and normal ranges shown in the table are expressed in the conventional units measured by the devices and laboratory kits used at our institution.
ACE, angiotensin-converting enzyme; ANA, anti-nuclear antibodies; C-ANCA, anti-PR3 antibody; ESR, erythrocyte sedimentation rate; IHC, immunohistochemistry; LDH, lactate dehydrogenase; P-ANCA, anti-myeloperoxidase antibody; PT, prothrombin time; PTT, partial thromboplastin time; RBC, red blood cells; TSH, thyroid-stimulating hormone; WBC, white blood cells.

Histologic examination of resected small tissue fragments revealed perivascular whorling of bland collagen fibers (onion skin-like appearance) with resulting obliteration of the vessel lumens (characteristic of the fibrotic stage of EAF) (Figure 2a). Some eosinophils were noted within the fibrosis. There were fragments of normal mucosa, overlain by respiratory-type epithelium, which included a moderate chronic
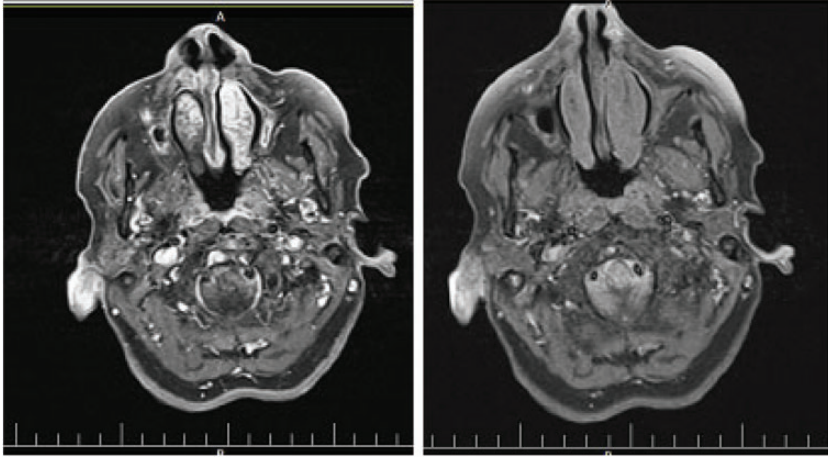
Figure 1. Axial T1-weighted images of the nasal cavity with and without contrast.

inflammatory cell infiltrate. Although a small number of eosinophils and neutrophils traversed the vessel walls, there was no evidence of fibrinoid necrosis or vasculitis. No giant cells, necrosis, or granuloma formation were present. Trichrome staining highlighted collagen whorls around vessels (Figure 2c). Specific stains for fungi, bacteria, and acid-fast bacilli were negative. In an immunohistochemical study, vimentin was strongly and diffusely positive in fibrotic areas of spindle cells and collagen bundles. These perivascular fibrotic areas were nonreactive with smooth muscle actin (Figure 2d) and CD34.

Discussion

The PubMed database was searched for all case reports on EAF published from inception in 1983 to 1 August 2022. All reports published in English that included the keywords "eosinophilic angiocentric fibrosis" OR "EAF" were extracted. The search yielded 47 eligible reports that included 71 cases of EAF.

The most commonly involved site was the nose (77.8%), the most common clinical manifestation was nasal obstruction (66.7%), and the most common management modality was surgical resection (83.3%). After surgery, 36% of patients were free of EAF without recurrence. The most common pharmacologic agent used was a corticosteroid (38.9%). The clinical manifestations, sites of lesions, management, and outcomes are summarized in Table 2.

EAF is a rare lesion of the sinonasal or upper respiratory tract, most commonly presenting with prolonged obstructive symptoms, a recurrent sinus mass, thickening of the mucosa, and deformity of the nose because of destruction of cartilage. A definitive diagnosis of EAF relies on histopathologic findings that include inflammation, infiltration of eosinophils, fibrotic bundles around the arteries, and an onion skin-like pattern with fibrotic stroma, usually without any signs of malignancy.[5,9–12]

Our case was a woman in her early 60 s who had a 7-year history of a progressive nasal sinus mass. During this time, the mass had progressed to the frontal lobe and affected her vision. Furthermore, there was destruction of the nasal septum and appearance of the saddle nose deformity.
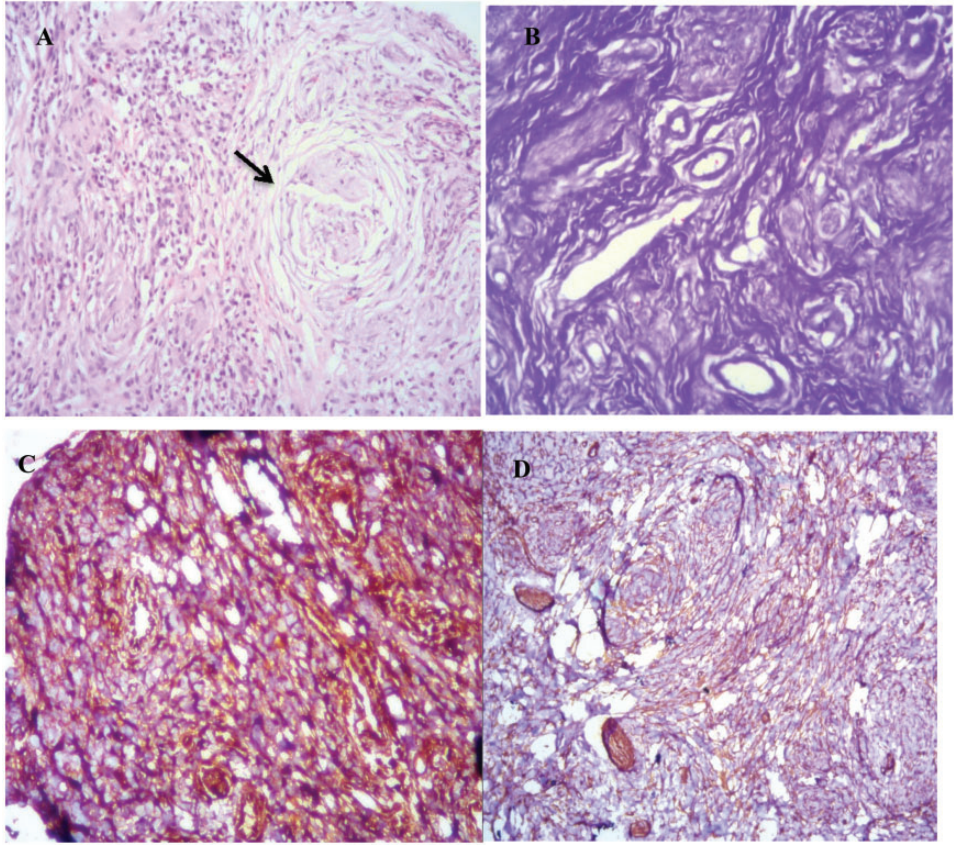
Figure 2. Histopathologic photomicrograph showing (a) fibrocollagenous tissue with perivascular infiltration of eosinophils accompanied by extensive areas of perivascular fibrosis (arrow) showing a characteristic angiocentric whorling with an onion skin-like pattern (hematoxylin and eosin staining, ×40 objective). (b) Masson trichrome staining shows perivascular fibrosis in the typical concentric pattern (×40 objective). (c) Diffuse positive staining for vimentin on immunohistochemistry (×40 objective) and (d) Negative staining for smooth muscle actin on immunohistochemistry (×40 objective).

EAF was diagnosed based on histopathologic, immunologic, and MRI findings

The symptoms of EAF are very vague in the early stages, the most common being nasal obstruction, epistaxis, respiratory problems, epiphora, proptosis, decreased sense of smell, and allergies to substances such as wool, plants, and carpet fluff. The disease is diagnosed radiologically by computed tomography or MRI. Soft tissue swelling, sinus opacification, thinning of the surrounding bone, turbidity of the nasal cavity and sinuses with or without bone erosion, sclerosis, and focal bone destruction are noted in radiographic reports.[10]

Histopathologic findings depend on the stage of the disease. Although there is no clear boundary between early and late lesions, the early stage is characterized by inflammatory eosinophil-rich vascular fibrotic lesions, and the late stage by dense perivascular (onion skin-like) fibrosis with fewer inflammatory cells. The biopsy can distinguish between the early and later stages of the disease.[9]

Table 2. Summary of cases of eosinophilic angiocentric fibrosis reported to date in the literature.

Author (year of publication)	Age/Sex	Clinical symptoms	Site of lesion	Management	Follow-up	Reference
Holmes and Panje (1983)	49/M	Nasal obstruction	Nose (lateral wall)	Intralesional triamcinolone, single resection	PD	21
Roberts and McCann (1985)	27/F	Nasal congestion	Nose (septum and lateral wall)	Antihistamine, nasal and systemic steroids, multiple resections	PD	7
	59/F	Nasal congestion	Nose (septum and lateral wall)	Intralesional steroids, multiple resections	PD	
Roberts and McCann (1997)	54/F	Nasal stuffiness	Nose (septum)	Multiple resections, radiotherapy	NA	22
	50/F	Nasal stuffiness	Nose (lateral wall)	Multiple resections	PD	
Altemani et al. (1997)	54/F	Nasal obstruction	Nose (septum and lateral wall)	Multiple resections	PD	23
Mataei et al. (2000)	51/M	Nasal obstruction	Nose (septum)	Single resection	FD	24
Thompson and Heffner (2001)	28/M	Nasal obstruction, epistaxis, pain	Nose (septum and lateral wall), maxillary sinus	Nasal and systemic steroids, single resection	PD	12
	49/F	Nasal obstruction	Nose (septum)	Nasal and systemic steroids, single resection	PD	
	64/F	Nasal obstruction	Nose (septum and lateral wall), maxillary sinus	Nasal and systemic steroids, single resection	PD	
Burns et al. (2001)	38/M	Nasal obstruction and swelling	Nose (septum and lateral wall)	Intralesional and systemic steroids, multiple resections, dapsone, YAG laser	PD	25
Loane et al. (2001)	42/M	Nasal obstruction, postnasal drip	Nose (septum)	Immunosuppression, multiple resections	NA	26
Pereira et al. (2002)	52/M	Nasal obstruction	Nose (septum)	Partial resection	PD	27
Owa et al. (2002)	41/M	Nasal obstruction	Nose (septum)	Single resection	FD	28
Goldman (2003)	50/F	Nasal obstruction	Nose (septum & lateral wall)	Intralesional steroids, multiple resections	PD	29
Tabaee et al. (2003)	79/M	Nasal congestion, swelling, tenderness	Nose (septum and lateral wall)	Single resection	FD	30
Onder and Sungur (2004)	45/M	Nasal obstruction	Nose (septum)	Single resection	NA	31

(continued)

Table 2. Continued.

Author (year of publication)	Age/Sex	Clinical symptoms	Site of lesion	Management	Follow-up	Reference
Chinell et al. (2004)	31/F	Nasal obstruction	Nose (septum)	Biopsy and dapsone	PD	32
Nguyen et al. (2004)	31/F	Nasal obstruction	Nose (septum)	Single resection	NA	33
Narayan and Douglas (2005)	72/F	Nasal obstruction and swelling	Nose (septum and lateral wall)	Biopsy	PD	34
Paun et al. (2005)	37/F	Epistaxis, epiphora	Nose (lateral wall)	Multiple resection, oral steroids and azathioprine	PD	1
	68/M	Nasal obstruction	Nose (septum)	Single resection	FD	
	57/F	Nasal obstruction	Nose (septum), orbit	Single resection, dapsone, hydroxychloroquine, azathioprine, systemic steroids	PD	
Yung et al. (2005)	58/F	Nasal obstruction	Nose (lateral wall)	Single resection	FD	35
	66/F	Recurrent epistaxis	Nose (septum)	Biopsy	PD	
	45/F	Recurrent epistaxis, nasal obstruction	Nose (septum, lateral wall), maxillary sinus	Biopsy, oral prednisone, azathioprine	PD	
Holme et al. (2005)	72/F	Nasal obstruction	Nose (septum, lateral wall)	Biopsy, pulsed dye laser, dapsone and clofazimine	PD	36
Slovik et al. (2006)	45/M	Nasal obstruction	Nose (septum)	Biopsy	PD	37
Clauser et al. (2006)	31/M	Nasal obstruction	Nose (septum)	Single resection	PD	38
Watanabe and Moriwaki (2006)	51/M	Nasal obstruction	Nose (septum)	Single resection	FD	39
Nigar et al. (2007)	67/F	Nasal obstruction and swelling	Nose (septum)	Single resection	PD	40
Jain et al. (2008)	31/F	Epiphora, orbital mass	Sinus (maxillary, ethmoid), orbit	Multiple biopsies, steroids, single resection	PD	41
	57/M	Nasal congestion, epiphora, proptosis	Nose (lateral wall), multiple sinuses, lacrimal gland	Steroids, single resection	PD	
	27/F	Nasal obstruction	Nose (lateral wall)	Biopsy	NA	
	51/F	Nasal mass, obstruction, and epiphora	Nasal (lateral wall)	Single resection	PD	

Table 2. Continued.

Author (year of publication)	Age/Sex	Clinical symptoms	Site of lesion	Management	Follow-up	Reference
Kosarac et al. (2008)	19/F	Nasal congestion, face pain	Right maxillary sinus	Single resection	FD	13
	31/F	Nasal obstruction	Nose (right nasal cavity)	Single resection	FD	
	49/M	Nasal obstruction	Nose (septum)	Single resection	FD	
Deshpande et al. (2011)	63/M	Pulmonary infiltrate and mass in sinus cavities	Nose, lacrimal gland, lung	Single resection, steroids	PD	5
	81/F	NA	Left nose	Single resection, steroids	PD	
	31/F	NA	Nose, orbit, maxillary sinus, ethmoid	Single resection, steroids	PD	
	54/F	NA	Right lacrimal gland	Single resection, steroids	PD	
	55/M	NA	Orbit	Single resection, steroids	PD	
Nogueira (2011)	NA	NA	Subglottic	CO_2 laser, corticosteroids, dapsone	FD	42
Yang et al. (2011)	26/F	Nasal swelling	Nose (septum)	Single resection	PD	43
	16/M	Nasal swelling	Nose (septum)	Single resection	PD	
	62/F	Nasal swelling	Nose (right nasal lateral wall)	Single resection	PD	
	28/M	Nasal swelling	Nose (septum)	Single resection	FD	
	24/F	Nasal swelling	Nose (septum)	Single resection	FD	
	73/M	Nasal swelling	Nose (left nasal lateral wall)	Single resection	FD	
Benlemlih et al. (2012)	86/F	Vision loss in left eye	Left eye	Steroids	PD	44
Li et al. (2013)	27/F	Nasal obstruction, headache, epistaxis, facial pain	Nose	Single resection	FD	18
Kim et al. (2014)	29/F	Dyspnea, nasal obstruction	Lower respiratory tract	Steroid (prednisone), bronchodilator (fluticasone + salmeterol, doxofylline) 3 months	PD/waning of symptoms	6
Lloyd et al. (2015)	45/M	Vision loss in left eye, epistaxis, headache	Left eye	Steroids, azathioprine cotrimoxazole	PD	45

Table 2. Continued.

Author (year of publication)	Age/Sex	Clinical symptoms	Site of lesion	Management	Follow-up	Reference
Okamato et al. (2015)	69/F	Chronic cough	Lung (right hilar area)	Single resection	FD	46
Faramarzi et al. (2015)	35/M	Nasal obstruction, epiphora	Nose	Single resection, steroids	FD	47
Jin et al. (2016)	31/F	Nasal obstruction, sinus pain	Nose	Single resection	PD	48
	58/M	Nasal obstruction, rhinorrhea	Nose	Observation without surgery	PD	
Hardman et al. (2017)	62/F	Saddle nose deformity, shortness of breath	Nose	Single resection, steroids	FD	49
Gorostis et al. (2017)	61/M	Vision loss in right eye, pain, proptosis, eyelid edema, nasal obstruction	Right ethmoid, orbit	Single resection	FD	50
Keogh et al. (2017)	56/F	Globus sensation, hoarseness, stridor	Subglottal	Single resection, Dexamethasone	FD	4
Heft et al. (2017)	45/F	Nasal obstruction, hyposmia	Paranasal sinus	Single resection	FD	51
Sazgar et al. (2019)	45/M	Nasal obstruction	Nose	Single resection	FD	52
Legare et al. (2018)	58/M	Left periorbital edema, nasal discharge	Left periorbital	Corticosteroid/ rituximab	PD	53
Němec et al. (2020)	NA	Nasal obstruction	Nose	Single resection	PD	54
Okuyama et al. (2020)	55/F	Conjunctival and eyelid swelling	Upper eyelid, conjunctiva	Single resection, Fluorometholone	FD	2
Saenz-Ibarra et al. (2020)	37/F	Nasal deformity, facial pain	Nose	Single resection	FD	55
Heedari et al. (2021)	69/M	Periorbital edema, epiphora, retroocular pain	Left orbit	Steroids	PD	56
Han et al. (2021)	32/M	Nasal obstruction	Nose	Single resection, nasal steroid, antihistamine	FD	57

Table 2. Continued.

Author (year of publication)	Age/Sex	Clinical symptoms	Site of lesion	Management	Follow-up	Reference
Daneshi et al. (2022)	33/F	Blindness in right eye, decrease in visual acuity in left eye	Supra sellar mass	Single resection	PD	58
Farina et al. (2022)	76/M	Nasal obstruction	Nose	Single resection	FD	59
Present report (2022)	55/F	Vision loss, nasal obstruction	Nose	Single resection, corticosteroids	FD	–

F, female; FD, free of disease; M, male; PD, persistent disease; NA, not available.

The differential diagnosis of EAF includes neoplasms and non-infectious granulomatous diseases, such as Wegener's granulomatosis, eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome or allergic granulomatosis), granuloma faciale (GF), and Kimura disease.[13]

The histopathologic findings of Wegener's granulomatosis include granulomatous vasculitis, geographic necrosis, and a positive C-ANCA test.[14] Churg–Strauss syndrome is characterized by eosinophilic vasculitis, fibrinoid necrosis with granuloma, and a positive P-ANCA test.[15] In Kimura disease, dense lymph masses with prominent germinal centers are dominant, and although fibrosis is present, it lacks the typical angiocentric rotation pattern characteristic of EAF.[16] GF is a benign skin disease of unknown cause that is severely limited by plaques and skin nodules tending to the facial area but mucosal involvement is also possible.[17] EAF is also referred to as an extracutaneous GF lesion.[18] Histopathologic examination of GF shows diffuse infiltration of eosinophils, neutrophils, and lymphoid cells at the surface. The primary lesion shows vasculitis, and although fibrosis exists, it is neither prominent nor concentrated in layers.[17]

Immunoglobulin (Ig)G4-related diseases, like EAF, are a complex of inflammatory diseases that affect various organs, including the pancreas, lungs, kidneys, and salivary glands.[19] IgG4 increases in only 50% of patients. Elevated plasma cell concentrations associated with IgG4 and an IgG4 to IgG ratio of >0.4 support a diagnosis of EAF.[5]

Immunohistochemistry and flow cytometry examinations can aid in the clarity of lesion components, although most are not necessary for EAF. Flow cytometry usually shows no unusual T-cell populations, plasma cells are polyclonal, and infiltrating cells are a combination of neutrophils,

histiocytes, and eosinophils. On immuno-histochemistry, vimentin is positive, smooth muscle actin is negative or may be positive in a small number of cells, S100 is negative, and diagnostic bacterial, acid-fast, and fungal staining tests are all negative.[9]

The treatments used for EAF to date include surgery (endoscopic sinus surgery, lateral rhinotomy, open rhinoplasty, septo-plasty, curettage, and lesion excision), cor-ticosteroids (systemic and/or intralesional), and immunosuppressive agents (azathio-prine, dapsone, clofazimine, tacrolimus, hydroxychloroquine) either alone or in combination. However, in some cases, the disease has not been completely cured and has relapsed.[20]

EAF of the sinonasal tract is a benign but progressive disease. Surgery is helpful in patients with obstructive symptoms. Recurrence is rare but typically occurs at the site of the primary lesion. Therefore, EAF is considered to be a progressive dis-ease. The disease may progress in some patients, even with corticosteroid therapy. Therefore, close follow up is essential.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

The research protocol obtained ethical approval from and was performed under the supervision of the review board of Kermanshah University of Medical Sciences. This report is presented in accordance with the CARE guidelines.[8] Written and verbal informed consent was obtained from the patient. The ethical standards relevant to patient confidentiality were observed at all times.

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References

1. Paun S, Lund VJ and Gallimore A. Nasal fibrosis: long-term follow up of four cases of eosinophilic angiocentric fibrosis. *J Laryngol Otol* 2005; 119: 119–124.
2. Okuyama S, Yazu H, Ito Y, et al. Eosinophilic angiocentric fibrosis in bilateral upper eyelid conjunctivas: a first case report. *Am J Case Rep* 2020; 21: e924042–e.
3. Fageeh NA, Mai KT and Odell PF. Eosinophilic angiocentric fibrosis of the sub-glottic region of the larynx and upper tra-chea. *J Otolaryngol* 1996; 25: 276–278.
4. Keogh I, O'Connell R, Hynes S, et al. Eosinophilic angiocentric fibrosis as a sten-osing lesion in the subglottis. *Case Rep Otolaryngol* 2017; 2017: 2381786.
5. Deshpande V, Khosroshahi A, Nielsen GP, et al. Eosinophilic angiocentric fibrosis is a form of IgG4-related systemic disease. *Am J Surg Pathol* 2011; 35: 701–706.
6. Kim WJ, Kim YI, Kim JE, et al. Unexplained persistent dyspnea in a young woman with eosinophilic angiocentric fibro-sis. *Respir Care* 2014; 59: e72–e76.
7. Roberts PF and McCann BG. Eosinophilic angiocentric fibrosis of the upper respiratory tract: a mucosal variant of granuloma faciale? A report of three cases. *Histopathology* 1985; 9: 1217–1225.
8. Gagnier JJ, Kienle G, Altman DG, et al. The CARE guidelines: consensus-based clinical case reporting guideline development. *Headache* 2013; 53: 1541–1547.
9. Ahn J and Flanagan M. Eosinophilic angio-centric fibrosis: a review and update of its association with immunoglobulin G4-related disease. *Arch Pathol Lab Med* 2018; 142: 1560–1563.
10. Han SC, Park JH and Hong SN. Eosinophilic angiocentric fibrosis invading the nasal septum: a case report and review of literature. *Ear Nose Throat J* 2020; 100: 557–561.

11. Helliwell TR. Non-infectious inflammatory lesions of the sinonasal tract. *Head Neck Pathol* 2016; 10: 32–39.

12. Thompson LD and Heffner DK. Sinonasal tract eosinophilic angiocentric fibrosis: a report of three cases. *Am J Clin Pathol* 2001; 115: 243–248.

13. Kosarac O, Luna MA, Ro JY, et al. Eosinophilic angiocentric fibrosis of the sinonasal tract. *Ann Diagn Pathol* 2008; 12: 267–270.

14. Lamprecht P and Gross WL. Wegener's granulomatosis. *Herz* 2004; 29: 47–56.

15. Chakraborty RK and Aeddula NR. Churg Strauss Syndrome (Allergic Granulomatosis). In: *StatPearls. StatPearls Publishing, Treasure Island (FL)*; 2019.

16. Zhang G, Li X, Sun G, et al. Clinical analysis of Kimura's disease in 24 cases from China. *BMC Surgery* 2020; 20: 1.

17. Nikolovska S, Gocev Đ and Damevska K. Granuloma faciale–a difficult diagnosis?–A case report. *Serbian J Dermatol Venereol* 212; 4: 32–38.

18. Li Y, Liu H, Han D, et al. Eosinophilic angiocentric fibrosis of the nasal septum. *Case Rep Otolaryngol* 2013; 2013: 267285.

19. Tokura Y. Spectrum of IgG4-related skin disease and differential diagnoses. *Hong Kong J Dermatol Venereol* 2020; 28: 100–110.

20. Fang CH, Mady LJ, Mirani NM, et al. Sinonasal eosinophilic angiocentric fibrosis: a systematic review. *Int Forum Allergy Rhinol* 2014; 4: 745–752.

21. Holmes DK and Panje WR. Intranasal granuloma faciale. *Am J Otolaryngol* 1983; 4: 184–186.

22. Roberts PF and McCann BG. Eosinophilic angiocentric fibrosis of the upper respiratory tract: a postscript. *Histopathology* 1997; 31: 385–386.

23. Altemani AM, Pilch BZ, Sakano E, et al. Eosinophilic angiocentric fibrosis of the nasal cavity. *Mod Pathol* 1997; 10: 391–393.

24. Matai V, Baer S, Barnes S, et al. Eosinophilic angiocentric fibrosis. *J Laryngol Otol* 2000; 114: 563–564.

25. Burns BV, Roberts PF, De Carpentier J, et al. Eosinophilic angiocentric fibrosis affecting the nasal cavity. A mucosal variant of the skin lesion granuloma faciale. *J Laryngol Otol* 2001; 115: 223–226.

26. Loane J, Jaramillo M, Young HA, et al. Eosinophilic angiocentric fibrosis and Wegener's granulomatosis: a case report and literature review. *J Clin Pathol* 2001; 54: 640–641.

27. Pereira EM, Millas I, Reis-Filho JS, et al. Eosinophilic angiocentric fibrosis of the sinonasal tract: report on the clinicopathologic features of a case and review of the literature. *Head Neck* 2002; 24: 307–311.

28. Owa AO, Boyle S and Gallimore AP. Eosinophilic angiocentric fibrosis as a cause of nasal obstruction. *Rhinology* 2002; 40: 41–43.

29. Goldman NC. Angiocentric eosinophilic fibrosis. *Otolaryngol Head Neck Surg* 2003; 128: 445–446.

30. Tabaee A, Zadeh MH, Proytcheva M, et al. Eosinophilic angiocentric fibrosis. *J Laryngol Otol* 2003; 117: 410–413.

31. Onder S and Sungur A. Eosinophilic angiocentric fibrosis: an unusual entity of the sinonasal tract. *Arch Pathol Lab Med* 2004; 128: 90–91.

32. Chinelli PA, Kawashita MY, Sotto MN, et al. Granuloma faciale associated with sinonasal tract eosinophilic angiocentric fibrosis. *Acta Derm Venereol* 2004; 84: 486–487.

33. Nguyen DB, Alex JC and Calhoun B. Eosinophilic angiocentric fibrosis in a patient with nasal obstruction. *Ear Nose Throat J* 2004; 83: 183–184, 186.

34. Narayan J and Douglas-Jones AG. Eosinophilic angiocentric fibrosis and granuloma faciale: analysis of cellular infiltrate and review of literature. *Ann Otol Rhinol Laryngol* 2005; 114: 35–42.

35. Yung A, Wachsmuth R, Ramnath R, et al. Eosinophilic angiocentric fibrosis–a rare mucosal variant of granuloma faciale which may present to the dermatologist. *Br J Dermatol* 2005; 152: 574–576.

36. Holme SA, Laidler P and Holt PJ. Concurrent granuloma faciale and eosinophilic angiocentric fibrosis. *Br J Dermatol* 2005; 153: 851–853.

37. Slovik Y, Putterman M, Nash M, et al. Eosinophilic angiocentric fibrosis of the

sinonasal tract in a male patient with chronic bowel inflammation. *Am J Rhinol* 2006; 20: 91–94.

38. Clauser L, Mandrioli S, Polito J, et al. Eosinophilic angiocentric fibrosis. *J Craniofac Surg* 2006; 17: 812–814.

39. Watanabe N and Moriwaki K. Atypical eosinophilic angiocentric fibrosis on nasal septum. *Auris Nasus Larynx* 2006; 33: 355–358.

40. Nigar E, Dhillon R, Carr E, et al. Eosinophilic angiocentric fibrosis and extra-facial granuloma faciale. *Histopathology* 2007; 51: 729–731.

41. Jain R, Robblee JV, O'Sullivan-Mejia E, et al. Sinonasal eosinophilic angiocentric fibrosis: a report of four cases and review of literature. *Head Neck Pathol* 2008; 2: 309–315.

42. Nogueira A, Lisboa C, Duarte AF, et al. Granuloma faciale with subglottic eosinophilic angiocentric fibrosis: case report and review of the literature. *Cutis* 2011; 88: 77–82.

43. Yang BT, Wang YZ, Wang XY, et al. Nasal cavity eosinophilic angiocentric fibrosis: CT and MR imaging findings. *AJNR Am J Neuroradiol* 2011; 32: 2149–2153.

44. Benlemlih A, Szableski V, Bendahou M, et al. [Eosinophilic angiocentric fibrosis: a form of IgG4-related systemic disease?]. *Ann Pathol* 2012; 32: 271–275. (French)

45. Lloyd AP, Benzimra JD, Warfield AT, et al. Optic neuropathy secondary to eosinophilic angiocentric fibrosis. *J Neuroophthalmol* 2015; 35: 45–47.

46. Okamoto K, Motoishi M, Kaku R, et al. A surgical case of eosinophilic angiocentric fibrosis of the lung. *Surg Case Rep* 2015; 1: 52.

47. Faramarzi M, Dadgarnia MH, Moghimi M, et al. Nasal eosinophilic angiocentric fibrosis with orbital extension. *Head Neck Pathol* 2015; 9: 426–429.

48. Jin CJ, Perez-Ordonez B and Witterick I. Eosinophilic angiocentric fibrosis of the sinonasal tract. *BJR Case Rep* 2016; 2: 20150419.

49. Hardman J, Toon C and Nirmalananda A. An unusual presentation of eosinophilic angiocentric fibrosis. *J Surg Case Rep* 2017; 2017: rjx234.

50. Gorostis S, Bacha M, Gravier S, et al. Right ethmoid eosinophilic angiocentric fibrosis with orbital extension. *Eur Ann Otorhinolaryngol Head Neck Dis* 2017; 134: 351–354.

51. Heft Neal ME, Rowan NR, Willson TJ, et al. A case report and systematic review of eosinophilic angiocentric fibrosis of the paranasal sinuses. *Ann Otol Rhinol Laryngol* 2017; 126: 415–423.

52. Sazgar AA, Kia S and Akbari A. Nasal framework reconstruction in patient with eosinophilic angiocentric fibrosis. *Indian J Otolaryngol Head Neck Surg* 2019; 71: 2031–2035.

53. Legare N, Frosh S, Vasquez JB, et al. Eosinophilic angiocentric fibrosis: a sino-orbital masquerader. *BMJ Case Rep* 2018; 2018: bcr201722367.

54. Němec I, Mikolaj M and Hrabal P. Eosinophilic angiocentric fibrosis as a cause of nasal obstruction. *Acta Chir Plast* 2020; 60: 59–61.

55. Saenz-Ibarra B, Ceceñas-Falcon LA, Cardenas-De la Garza JA, et al. Nasal eosinophilic angiocentric fibrosis with IgG4-positive plasma cell infiltration. *Malays J Pathol* 2020; 42: 137–141.

56. Heedari HM, Ciotoracu AC, Mitulescu TC, et al. A clinical case of orbital inflammatory pseudotumor as the primary expression of eosinophilic angiocentric fibrosis. *Rom J Ophthalmol* 2021; 65: 411–418.

57. Han SC, Park JH and Hong SN. Eosinophilic angiocentric fibrosis invading the nasal septum: a case report and review of literature. *Ear Nose Throat J* 2021; 100: 557–561.

58. Daneshi SA, Taheri M, Fattahi A, et al. The primary brain eosinophilic angiocentric fibrosis, a rare case report. *Prague Med Rep* 2022; 123: 113–119.

59. Farina J, Broggi G, Federico C, et al. Eosinophilic angiocentric fibrosis of the nasal cavities: a report of an uncommon lesion with emphasis on the etiology and differential diagnosis. *Medicina (Kaunas)* 2022; 58: 865.