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TITLE OF CASE

Recurrent atypical fibroxanthoma of the limbus

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SUMMARY

We report an unusual presentation of recurrent atypical fibroxanthoma of the limbus. Clinical and histological appearance, as well as management are discussed and the current literature is reviewed.

BACKGROUND

Atypical fibroxanthomas are uncommon cutaneous neoplasms and belong to the group of fibrohistiocytic tumors. They most commonly occur in actinic skin of the head and neck region in older Caucasian men. We report an unusual case of recurrent atypical fibroxanthoma near the limbus and discuss the pathophysiology of atypical fibroxanthoma.

CASE PRESENTATION

An 81-year old man presented with foreign body sensation of his left eye and a rapidly growing ocular surface lesion. He reported that the lesion had arisen rapidly and was now causing irritation when blinking. He was unable to give a precise timeframe for his complaints. There was no history of pain or bleeding associated with the lesion.

Past ocular history was significant for excision of a lesion from the left nasal limbus in 2007, performed in a rural centre in another State. The patient also gave a history of a previous excision of a similar lesion from the left eye some time prior to 2007 but no further medical details were available. The patient had also undergone bilateral cataract surgery in 2007.

On examination best-corrected visual acuity was 6/7.6 bilaterally. A limbal lesion was noted (*Figure 1).* Further ocular examination revealed bilateral pseudophakia but was otherwise unremarkable. Intraocular pressure was normal at 11mmHg OD, 12 mmHg OS.

Examination demonstrated a discreet, smooth, rounded vascular nodule at 11 o'clock on the left limbus. It appeared to be tethered to the conjunctiva and had a feeder vessel.

The histopathological report from the excision in 2007 was obtained and demonstrated morphological features consistent with atypical fibroxanthoma. Resection margins from this excision were involved. Immunohistochemistry revealed positivity for SMA, and negativity for AE1/3, CK5/6, desmin, calponin, CD68, MelanA, HMB45 and S100.

At that time, he was followed up yearly for monitoring of recurrence, but no regrowth of any nodule or lesion was noted. When the patient moved to his current location in 2010, he was strongly recommended to continue with ophthalmic monitoring, but this did not occur.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis for a recurrent lesion at the limbus could include ocular surface squamous neoplasia, conjunctival inclusion cyst, pterygium, naevus, melanoma, limbal dermoid, epithelioma, and squamous cell carcinoma.

TREATMENT, OUTCOME AND FOLLOW-UP

The patient subsequently underwent excision biopsy of the current lesion. Histopathology revealed an atypical fibroxanthoma, which had been narrowly but completely excised along the deep edge (0.5mm) and superficial margins. Histology demonstrated features of atypical fibroxanthoma, namely a highly pleomorphic spindle cell proliferation with enlarged irregular nuclei. There was no evidence of squamous dysplasia or melanocytic junctional proliferation (*Figure 2).* Further immunohistochemical stains were positive for CD10, but negative for S100, MelanA, p63, CK5/6.

The patient continues to be under regular ophthalmological surveillance to monitor for possible recurrence given that two previous excisions had been performed.

DISCUSSION

We have reported the presence of a rare neoplasm at the limbus, found to be an atypical fibroxanthoma (AFX). The case demonstrates the recurrence of AFX at this location, after

previous incomplete excision.

Atypical Fibroxanthomas are relatively uncommon cutaneous neoplasms and have only been reported twice in the literature as ocular surface lesions [1 2]. They are most common in the Caucasian population, with a 4:1 male-female ratio and occur in actinic skin, predominantly in the head and neck region of older people [3 4].

AFX has historically been thought to follow a benign course, although local recurrences and metastases have been reported [5-8]. Some recurrences may also represent insufficient excision margins of the initial tumor [8], but AFX can recur in a previously treated area. Recurrence rate has been reported as approximately 7-9% for cutaneous lesions [4].

Fibroxanthomas belong to the group of fibrohistiocytic neoplasms, which also includes malignant fibrous histiocytoma. Histologically as well as immunohistochemically, they appear to show similar features [4 9] and it is now thought that these may represent a spectrum of disease rather than distinct disease entities [4].

Histologically, AFX is characterized by spindle and pleomorphic cells [3 8]. Atypical fibroxanthoma must be differentiated from melanoma, leiomyosarcoma and squamous cell carcinoma, especially the spindle cell variants. It remains a diagnosis of exclusion [3 4]. Immunohistochemically, AFX commonly expresses vimentin and CD10, both of which can be non-specific [3 4]. AFX is only very rarely positive for desmin and stains consistently negative for S100 [4].

While limbal AFX lesions are extremely rare [10], one previous case report describes atypical fibroxanthoma of the limbus in a patient with previous penetrating eye injury and subsequent light perception vision [1]. Engelbrecht et al. described a large, nodular, opaque mass on the central cornea with extensive vascularization, obscuring view of the anterior chamber and the patient subsequently underwent lamellar keratectomy and complete excision of the limbal tissue. Histopathology later showed high-grade intraepithelial squamous neoplasia (in situ carcinoma) overlying an atypical fibroxanthoma.

A different case described primary AFX of the palpebral conjunctiva [2]. The patient presented with a nodule in the conjunctiva after previous evisceration some 50 years prior for trauma. The patient was treated with excision and adjuvant cryotherapy to the surgical bed.

One more case of ophthalmic recurrent AFX is included in a study on treatment of ocular neoplasms with topical mitomycin C therapy [6], but no details of the location and features of the

AFX were provided.

Our reported case describes a limbal atypical fibroxanthoma, which differs significantly in its morphology compared with the only other case reporting a recurrent ocular AFX. This report presents a case of recurrent AFX of the limbus, which to the authors' best knowledge, has not previously been reported in the literature.

LEARNING POINTS/TAKE HOME MESSAGES

- Atypical fibroxanthoma may occur as limbal lesion and recurrence is possible.
- As pathology may be unusual, biopsy is useful to guide management of lid and limbal lesions.
- Incomplete excision margins may warrant long term follow-up.

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Figure captions

Figure 1 – Atypical fibroxanthoma of the limbus

Figure 2 – Histological Section of the limbal atypical fibroxanthoma

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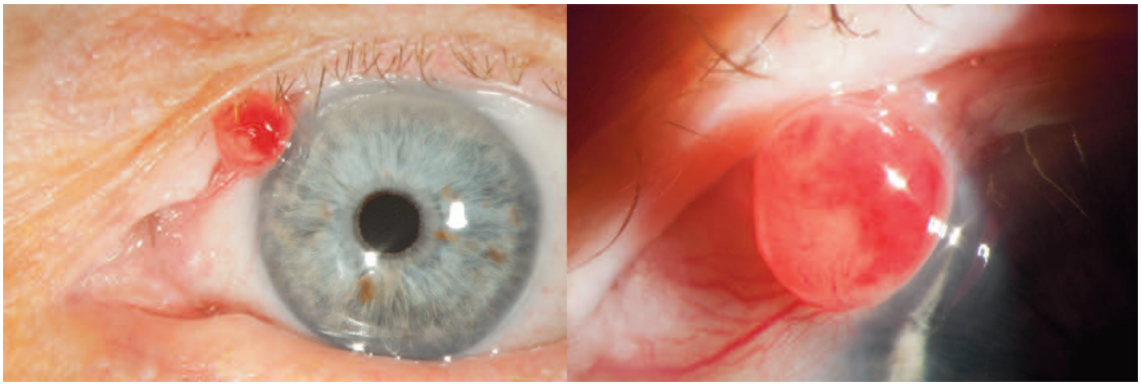

Figure 1

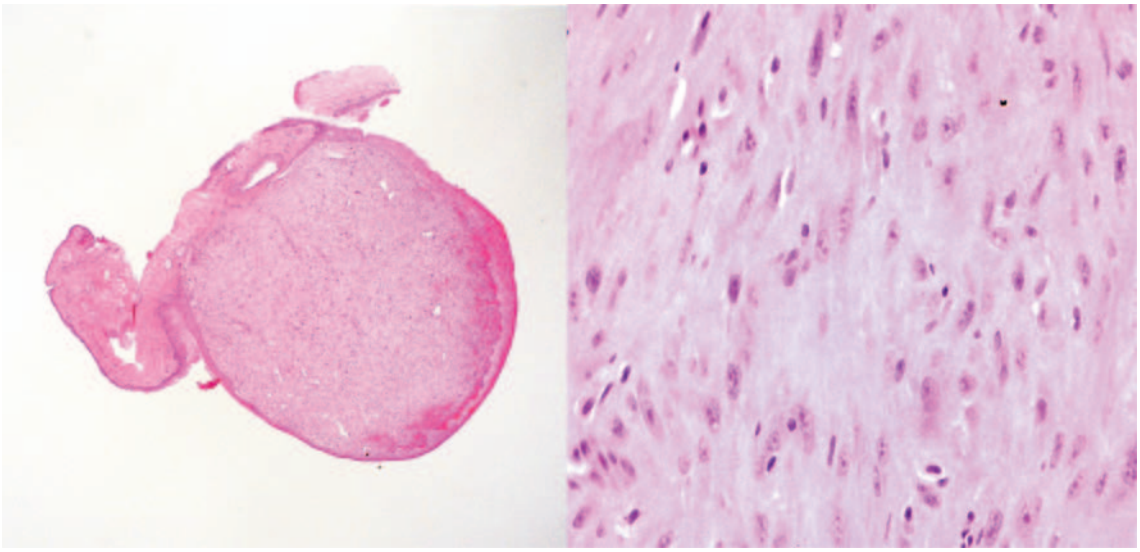

Figure 2