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Case Report OMICS International

An Uncommon Case of Atypical Fibroxanthoma of the Bulbar Conjunctiva

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Abstract

An Atypical Fibroxanthoma (AFX) is a benign skin tumor of mesenchymal origin most commonly found on the sun-exposed or radiation-damaged skin of the elderly. AFX may share many common histologic and clinical characteristics with squamous cell carcinoma and melanoma; however, it is a benign process. Therefore, differentiating between these lesions is important. Differences in histologic features and patterns of immunohistochemical staining for CD68, CD10, cytokeratins, S-100 and Fi-67 may diagnose AFX. The presence of an AFX on the conjunctiva is rare with only five other reported cases. We report a case of AFX that developed in the bulbar conjunctiva near the limbus.

Keywords: Atypical fibroxanthoma; Malignant fibrous histiocytoma

Case Study

A 58 year old Caucasian male was referred to our ocular oncology clinic for suspected conjunctival intraepithelial neoplasia of the right eye. Over four months, he noted growth of a conjunctival mass. The tissue began clear but grew to become more "flesh-like" and lobulated. The mass was associated with conjunctival injection, irritation, and epiphora. The patient denied pain with extraocular movements, blurry vision and diplopia. His visual acuity was normal and color vision was intact.

On slit lamp examination a nodular papillomatous lesion was noted on the right nasal conjunctiva, abutting the limbus, with associated injection (Figure 1).

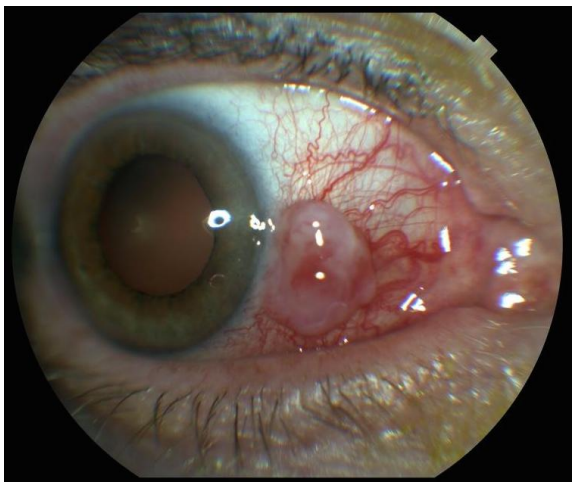

Figure 1: Gross image of an atypical fibroxanthoma of the bulbar conjunctiva.

The assessment was a malignant neoplasm of the right conjunctiva, likely squamous cell carcinoma. An excisional conjunctival biopsy was taken and histologic sections were made. Double freeze-thaw cryotherapy was applied to the base of the lesion immediately following excision. Tissue was then sent for permanent section to pathology. Haemotoxylin and Eosin staining was conducted to characterize the cell types present. Immunohistochemical staining with antibodies against CD68, CD10, Ki67, pan cytokeratin and S-100 was also completed using different sections of the biopsy.

Histopathology slides revealed a relatively well-circumscribed lesion within the conjunctiva (Figure 2A).

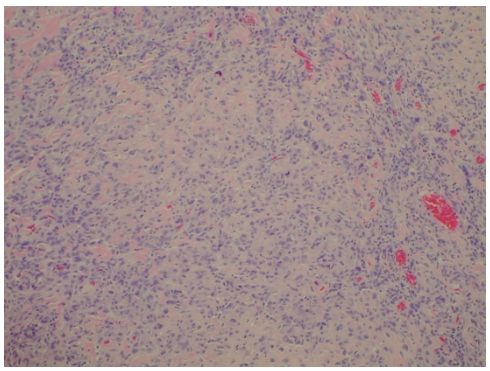

Figure 2A: H & E 4X.

The tumor was composed of highly atypical spindle and epithelioid cells focally surrounded by hyalinized stroma; thus, revealing many atypical mitotic figures (Figure 2B).

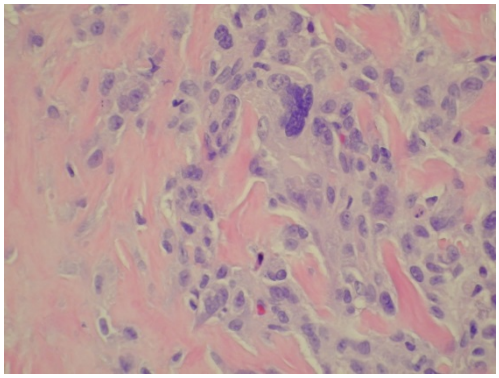

Figure 2B: H & E 40 X, spindle and epithelioid cells with interstitial hyaline fibrosis and scattered multinucleated cells and atypical cells with mitotic figures.

Immunohistochemical staining showed that the tumor cells were positive for CD68 (Figure 2C) and CD10 (Figure 2D) and had high mitotic activity by Ki67 (Figure 2E).

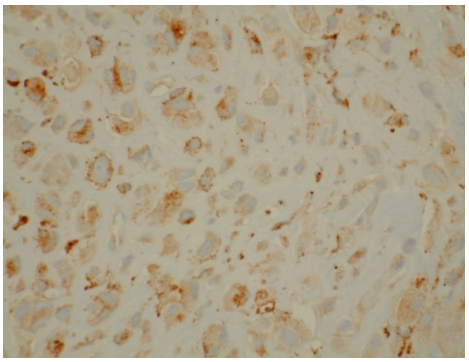

Figure 2C: CD68 40X, Majority of tumor cells are positive for CD68.

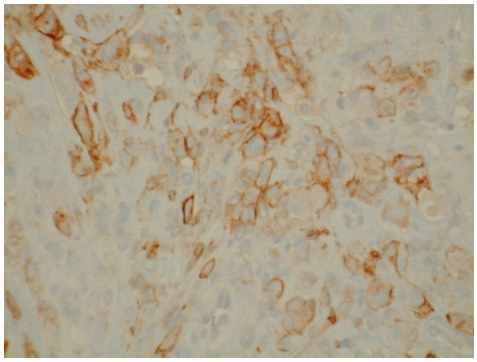

Figure 2D: CD10 40X, tumor cells show membranous and occasional cytoplasmic positive.

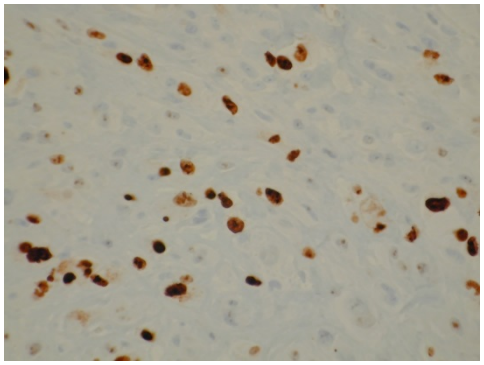

Figure 2E: Ki67 40X, tumor cells show high proliferation rate.

The cells were negative for Pan cytokeratin and S-100 (Figure 2F). A diagnosis of atypical fibroxanthoma was made.

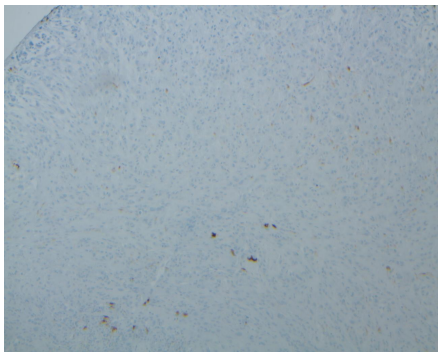

Figure 2F: S100 20X, scattered dendritic cells are stained by S100.

Discussion

This is the sixth reported case of AFX of the bulbar conjunctiva. Of the five other reported cases, four were found at the limbus, as in our patient [1-4]. The typical histologic presentation of AFX is spindle, plump, epithelioid and bizarre cells, in various proportions, arranged in haphazard, vaguely fascicular or storiform patterns [4]. Often, there is fibroblastic, myofibroblastic, and epithelioid cells with pleomorphism noted [5]. Some cells of the lesion may contain vacuolated and lipid-containing cytoplasm similar to a xanthoma [4]. Most of the reported cases of conjunctival AFX have been either spindle-cell predominant or pleomorphic with epithelioid and multinucleated giant cells [1]. Here we see some of these features including spindle-shaped cells, multinucleated giant cells and epithelioid cells, without a predominant cell type (Figure 2B).

Immunohistochemical staining is an important step in diagnosing AFX. The Ki-67 index is high in this tumor, relating to the rapid-growing nature of this tumor (Figure 2E). S-100 is positive in malignant melanoma. However, dendritic/Langerhans cells within AFX may express S-100 (Figure 2F) [4]. Cytokeratin is positive in squamous cell carcinoma but not AFX. This negative stain is not depicted. A histiocytic/macrophage marker, CD68, is positive in more than half of all AFX cases (Figure 2C). However, D68 is also detectable

in 86% of malignant melanoma cases [4]. Therefore, this marker can rule out squamous cell carcinoma but not melanoma. CD10 is positive in 95-100% of AFX (Figure 2D). It is reported that about one third of melanomas and half of squamous cell carcinomas are also positive for CD10. However, melanomas and squamous cell carcinomas stained weakly, with majority accumulating in the stroma [4]. When using immunohistochemical staining to diagnose AFX it is important to known these possible variations.

Many consider atypical fibroxanthoma as a superficial variant of Malignant Fibrous Histiocytoma (MFH), which is known to have an aggressive growth pattern with significant metastatic potential. The two are identical histologically featuring fibrohistiocytic lesions with myofibroblastic differentiation [4]. However, AFX is located in the dermis while MFH can be found within the subcutaneous fat, with perineural and lymphovascular invasion [5]. No case of metastatic conjunctival AFX has been reported. It is thought that any case of metastatic AFX of the skin was misdiagnosed MFH [6]. Likewise, any case of a metastatic soft tissue tumor of the conjunctiva has been reported as MFH [7,8].

This case of AFX had similarities and differences when compared to other reported cases in the literature. The location near the limbus was similar to four out of five cases of bulbar conjunctival AFX. While 33% of previously reported cases were associated with Xeroderma Pigmentosum (XP) [5], our case was not. Although AFX of the skin is associated with sun exposure or radiation damage in the elderly, now 42% of cases of AFX of the conjunctiva are found in patients under 60 years old without XP. The physical appearance, rapid growth period, and symptoms of irritation and epiphora were similar to previously reported cases [1]. The histologic appearance of our case was different from others in that there was no predominant cell type identified within the lesion.

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