Ophthalmology Practice

Benign Fibrous Histiocytoma

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Fibrous histiocytomas (FHs) are mesenchymal tumors that may be benign or malignant. Ocular involvement by FHs is infrequent and primarily limited to the orbit. Rarely, FHs can also involve the conjunctiva and perilimbal area. We report the case of a 38-year-old male with lid, conjunctival, and neck FHs. The diagnosis was confirmed by histopathology.

Key words: Benign, excision, lids and caruncle, no recurrence, ocular FH

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Fibrous histiocytomas (FHs) are mesenchymal tumors that may be benign or malignant. Ocular involvement by FHs is infrequent and primarily limited to the orbit. In a series of 1264 consecutive orbital tumors from an Ocular Oncology Center, FH represented 0.5% (6/1264 tumors) of all orbital tumors and 6% (5/810 tumors) of all benign orbital tumors.[1] Rarely can FH be found in the conjunctiva. In a review of 1643 conjunctival tumors, FH represented only 0.2% (4/1643) of all tumors.[2] This report describes the clinical and histopathological features of lid and conjunctival FH in a 38-year-old male patient.

Case Report

A 38-year-old male was seen in August 2004, with a one-year history of mass lesions in the right lower and left upper lid, and redness and foreign body sensation in the left eye since one month. The patient was seen previously by an ophthalmologist, who recommended a trial of hot compresses and erythromycin ointment for a presumed chalazion, as also some topical medications for red eye, but there was no response; the size and character of the lid and conjunctival lesion remained stable. On clinical examination, visual acuity in both eyes (OU) was 20/20. Yellowish brown nodules that were firm, nontender, well-circumscribed, not fixed to the overlying skin or underlying bone, were present in the right lower lid (four in number, all less than 20 mm in diameter) and left upper lid (two in number, both about 10 mm in diameter). A firm, pinkish nodule (4 mm) with surrounding engorged conjunctival vessels involving the left caruncle was also seen. The remainder of the ocular examination of both eyes was unremarkable. On systemic examination, five nodules (less than 20 mm in size), yellowish brown, firm, nontender, well-circumscribed, not fixed to the overlying skin or underlying bone, were also seen on the neck [Figs. 1-3].

An excisional biopsy of one of the neck nodules was done and a histopathological diagnosis of benign FH was made [Fig. 4]. Keeping this in mind we went for excision of the lid nodules and the excision was accompanied by cryopexy of the conjunctival nodule. The histopathology reports of all excised nodules were similar to those of the neck nodules (i.e. consistent with findings of benign fibrous histiocytomas). The patient was followed up for seven years and there was no evidence of recurrence.

Discussion

O'Brien and Stout[3] first described fibrous histiocytoma with orbital involvement. Russman[4] reported another case with primary orbital involvement. Zimmerman[5] commented on the histopathology of orbital fibrous histiocytoma, but did not include any case protocols. Fibrous histiocytoma of the conjunctiva is rare and can be classified as benign or malignant. Only 23 previous cases of conjunctival FH have been reported since 1968 [Table 1]. The mean age of these 23 patients was 39 years with no gender predilection. The mean duration of the symptoms before diagnosis was eight months. Two cases involved both eyes,[6,7] and the rest of the cases were unilateral (nine OD, eleven OS, one unspecified). Tumor location was at the corneoscleral limbus in 74% (17 cases) and on the cornea in 22% (five cases). Our case was of a 38-year-old male patient, with a one-year history of nodules on the lids, left caruncle, and neck.

Histopathologically, benign FH typically shows a biphasic cell population of histiocytes and fibroblasts. In some cases the cells resemble myofibroblasts, primitive mesenchymal cells, and cells having intermediate or mixed features. The presence of a homogeneous population of fibroblast-like cells has also been described. Other histological features frequently described in benign FH are the presence of multinucleated giant

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Table 1: Summary of previously reported cases of conjunctival fibrous histiocytoma: Clinical features, management, and clinical outcome

Study	Age (y)	Gender	Duration of Lesion (mo)	Eye	Conjunctival Location	Quadrant Location	Basal Diameter (mm)	Thickness (mm)	Malignant	Total Excision (*n*)	Additional treatment	Recurrence (*n*)	Follow-up (mo)
Kiratli (2003)	39	M	1	OS	Bulbar	Superonasal	4	3	N	1	None	0	48
Allaire (1999)	60	M	9	OD	Limbus	Nasal	4		Y	4	None	3	12
Mietz (1997)	27	F	5	OS	Limbus	Inferior	9		N	2	None	0	Lost
Brodovsky (1996)	11	M	<1	OD	Limbus	Superotemporal	5		N	2	None	1	18
Balestrazzi (1991)	53	M		OS	Limbus	Temporal			Y	4	Exenteration	3	18
Pe'er (1990)	4	F		OU	Bulbar		12	7	Y	1	None		Lost
Pe'er (1990)	58	M	12	OD	Limbus	Superonasal	15	4	Y	2	Exenteration	1	50
Margo (1989)	59	M	18	OD	Bulbar		12		Y	1	Exenteration parotidectomy	1	12†
Nores (1989)	49	F	2	OS	Limbus	Nasal	5		N	1	None	0	48
Schellini (1989)	75	F	3	OS	Limbus	Nasal	10		N	1	None	0	1
Lahoud (1988)	11	M	3	OS	Limbus	Inferior	9		N	1	None	0	20
Lahoud (1988)	28	F	4	OS	Limbus	Inferior	2		N	1	None	0	2
Jakobiec (1988)	<1	M	<1	OS	Caruncle	Nasal	18	8	N	1	None	0	10
Kantelip (1988)	49	F			Limbus				N	1	None	0	48
Lew (1985)	66	M		OS	Limbus	Temporal			N	2	Immunotherapy‡	2	24
Urdiales-Viedma (1983)	72	F	10	OD	Limbus		19	9	Y	1	None	0	18
Litricin (1983)	65	F	36	OD	Limbus	Inferotemporal			N	4	Enucleation	1	48
Iwamoto (1981)	31	M	5	OS	Limbus	Interonasal	4		N	1	None	0	48
Paglen (1980)	14	M	1	OD	Canthus	Temporal canthal	6	4	N	0	Radiation		0.25
Faludi (1975)	21	F	2	OS	Limbus	Inferotemporal	5		N	2	None	1	5
Jakobiec (1974)	3	F	3	OD	Limbus	Temporal	4		N	1	None	0	36
Delgado-Partida (1972)	50	F	36	OD	Limbus	Nasal	7	3	Y	3	Enucleation	2	19
Albert (1968)	44	F	6	OU	Bulbar	Inferior conjunctiva			N	1	None		

F: Female, M: Male; N: No, OD: Right eye, OS: Left eye, OU: Both eyes, Y: Yes, Mo: Months, †Patient showed metastasis to the parotid four months after exenteration and was found to have multiple lung metastasis 10 months after parotidectomy with radical neck dissection; patient died one year after.‡Dinitrochlorobenzene was administered subconjunctivally onto the lesion, which helped achieve resolution and prevention of the recurrence. ‡Dinitrochlorobenzene was administered subconjunctivally onto the lesion, which helped achieve resolution and prevention of the recurrence

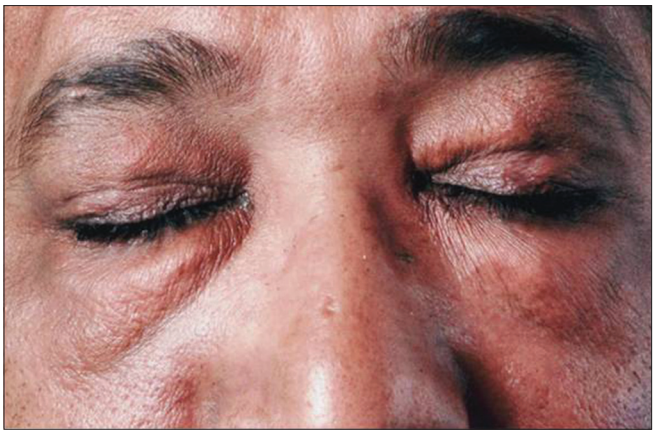

Figure 1: Nodules on the right and left lids

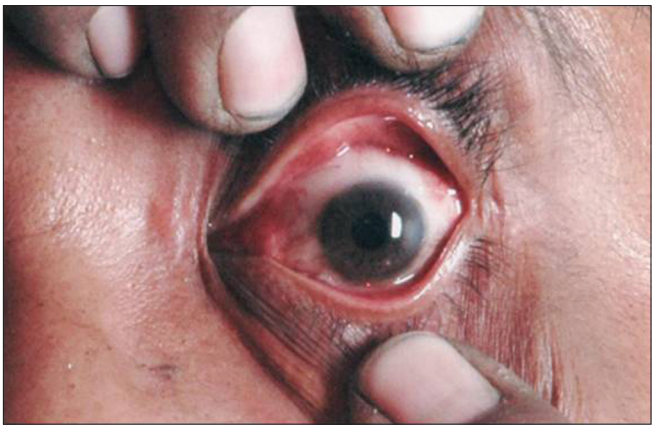

Figure 2: Nodule in the left caruncle

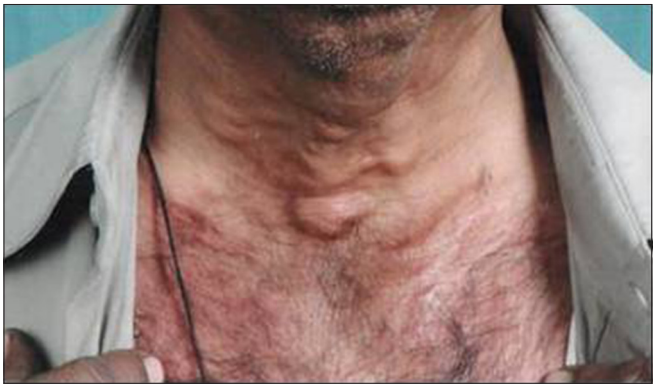

Figure 3: Nodules on the neck

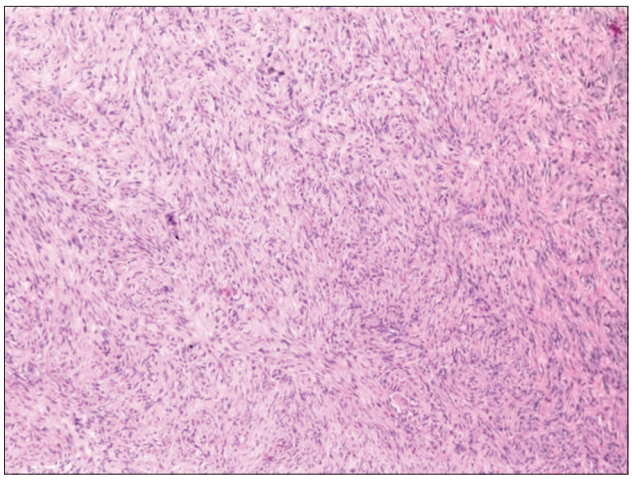

Figure 4: Histopathology slide of a nodule showing spindle cells, multinucleated cells, foam cells, and few mitotic figures. All excised nodules revealed similar findings on histopathology

cells, abundant vascularity, and an inflammatory infiltrate. Benign FH must be differentiated from soft tissue lesions of Langerhans cell disease (histiocytosis X) and routine xanthoma, and from reactive histiocyte proliferations as seen after trauma or in persons with hyperlipidemia or hypercholesterolemia. The histiocytic cells of a xanthoma typically express S-100 protein, while the lesional cells of the fibrous histiocytoma do not. Cholesterol clefts, moreover, are commonly seen in the xanthoma. The multinucleated giant cells of Langerhans disease do not typically have their nuclei pushed to the cell periphery. Our case had fibroblast-like spindle cells, multinucleated cells, foam cells, and few mitotic figures.

The most appropriate management of FH at any site, particularly the conjunctiva, is complete surgical excision with tumor-free margins. Local recurrence, orbital invasion, and metastasis typically follow incomplete initial excision. Of the 23 reported benign conjunctival FH cases, 20% (four cases) recurred within an average time of seven months (median, seven months; range, two to eleven months), but no metastasis was observed. Repeat excision successfully controlled the recurrence in two cases[8,9] and immunotherapy controlled the recurrence in one case. One case required enucleation for intraocular involvement after four excisions.[10] In our case there was no recurrence in a follow-up of seven years.

To conclude, orbital fibrous histiocytomas are rare periocular tumors that can manifest multiple ocular signs and symptoms. A careful histological examination is necessary for diagnosis, as these tumors have a wide range of morphology. Complete resection is advised, with careful evaluation of the surgical margins. Both benign and malignant FH can show local recurrence and must be treated aggressively.

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