Saudi Journal of Ophthalmology (2018) 32, 33–39

Original Article

Inflammatory myofibroblastic tumor of the orbit: A clinico-pathological study of 25 cases

Diego Strianese [a,*]; Fausto Tranfa [b]; Marialuisa Finelli [b]; Adriana Iuliano [b]; Stefania Staibano [c]; Giuseppe Mariniello [b]

Abstract

Background: Inflammatory myofibroblastic tumor (IMT) is a rare entity characterized by the presence of myofibroblasts and inflammatory cells within a fibrous stroma, which typically occurs in children or young adults. The IMT is considered generally a benign lesion, although about 20% of cases may experience recurrence, and most rarely develop metastasis. Herein, we present the largest series of primary orbital IMT ever reported.
Patients and methods: The clinical records of 25 patients, collected between the 1995 and 2015, with biopsy-proven diagnosis of orbital IMT were retrospectively reviewed to determine demographic, clinical, radiologic and pathological features, management, and outcome.
Results: The study included 13 females and 12 male patients, age ranged from 5 to 76 years. Disease onset was in all cases unilateral (25/25), with posterior location (10/25) or extending anterior to posterior (7/25). The most common signs and symptoms were: proptosis (19/25), ptosis (18/25), diplopia (10/25), periocular swelling (9/25), pain (8/25), redness (7/25). All patients underwent to incisional biopsy which included total or subtotal tumor resection avoiding arming of the adjacent structure, followed by systemic steroid therapy (22/25) or radiotherapy (3/25). The disease recurred in 6 (24%) patients who responded to the subsequent therapy. No one developed metastasis or died because of the disease.
Conclusion: IMT is a distinct entity which may occur in the orbit primarily. It should be considered in differential diagnosis in all orbital masses, particularly with onset of acute or subcronic inflammation. Surgical biopsy associated to a partial debulcking of the tumor, avoiding to damage adjacent vital structure may contribute to improve the outcome. Steroid therapy, seems to be the suitable as first line medical therapy, although, as reported in literature, not all cases respond to this treatment regimen. Radiotherapy, may be considered as an alternative therapy. Recurrences occurred in 24% of patients and may be treated with additional surgical resection and a new course of steroid or radiotherapy. No specific pathological features which may correlate with the prognosis have been found in this series.

https://doi.org/10.1016/j.sjopt.2018.04.001

Introduction

Inflammatory myofibroblastic tumor (IMT) or inflammatory pseudotumor is a spindle cell proliferation of disputed nosology.[1] Although the lung is the best known and most common site, inflammatory myofibroblastic tumor occurs in diverse extra-pulmonary sites; the extra-pulmonary location concerns generally young patients between the first and the second life decades.[1–3] Although rare, there are case reports in the literature of IMT in the orbit.[4–11] Herein we report our series of 25 patients with the proven diagnosis of IMT collected in a

Available online 4 April 2018.

[a] King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia
[b] Department of Neuroscience and Reproductive and Odontostomatological Sciences, Napoli – University Federico II di Napoli, Napoli, Italy
[c] Department of Biomorphological and Functional Science, Napoli – University of Naples Federico II | UNINA, Italy

* Corresponding author at: King Khaled Eye Specialist Hospital, PO Box 7191, Riyadh 11462, Saudi Arabia.
e-mail address: dstrianese@kkesh.med.sa (D. Strianese).

Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

Access this article online: www.saudiophthaljournal.com www.sciencedirect.com

20 years period. To the best of our knowledge this is the largest ever reported in literature on IMT involving solely the orbit.

Patients and methods

The clinical records of 25 patients, collected between 1995 and 2015 at the Orbital Unit of the University of Naples '' Federico II'', with the proven diagnosis of IMT of the orbit have been reviewed retrospectively to obtain patient information, including age, sex, signs and symptoms at the onset, radiologic features, location, clinical course, outcome and management. Clinical data were correlated with histopathologic and immunopathologic findings.

Results

Demographic data, characteristics of disease, management and outcome of each case are presented in Table 1.

Patients ranged in age from 5 to 76 years (mean, 50 years), there was no significant gender predilection. Distribution of the age has been plotted with the VI decade which, turned out to be the most frequent one (Fig. 1).

Disease affected the left eye in 14 patients, the right eye in 11 patients. The superior-lateral quadrants was the most frequent site involved. The mass was considered to be anterior in 6 patients, posterior in 10 patients, diffuse in 9 patients. Data are summarized in Table 2.

Symptoms developed between 20 days and 13 years before initial examination. Duration of the symptoms was longer than 1 month in 16 patients, and acute few days in 2 patients (Table 1).

Proptosis and ptosis were the most common symptoms. Signs and symptoms of inflammation such as periocular swelling, chemosis, redness and pain occurred in 32% up to 40% of cases (Table 2).

Images or detailed reports of examination by computed tomograhy (CT) or magnetic resonance imaging (MRI) were

Table 1. Patients' characteristics.

Patient No	Sex	Age at Onset (y)	Onset Time (month)	Onset Course	Side	Clinical Signs and Symptoms	Visual Acuity (10/10)
1	F	22	6	CHR	Left	Swelling of left superior eyelid, moderate ptosis, moderate proptosis	20/20
2	M	66	2	CHR	Right	Eyelid edema, pain, moderate ptosis moderate proptosis	20/20
3	F	43	2	CHR	Left	Mild proptosis, conjunctiva edema, pain	20/20
4	F	69	4	CHR	Left	Moderate proptosis, diplopia, periorbital edema, diplopia	20/20
5	M	65	5	SA	Left	Moderate proptosis, tearing, diplopia, superior rectus muscle restriction.	20/25
6	M	63	24	CHR	Right	Moderate proptosis	20/200
7	F	68	2/3	SA	Right	Pain, conjunctival redness, moderate proptosis	20/25
8	M	49	1	SA	Left	Moderate proptosis, Severe ptosis, conjunctival redness	20/30
9	F	76	5	AC	Left	Orbital trauma, moderate proptosis, mild ptosis, pain, diplopia, hypofunction of lateral rectus muscle	20/20
10	F	40	4	CHR	Right	Swelling, conjunctival redness, mild proptosis	20/20
11	F	33	2/3	SA	Right	Pain, eyelid edema, mild proptosis, mild ptosis, diplopia	20/20
12	F	58	4	CHR	Left	Mild proptosis, tearing, diplopia	20/20
13	M	34	3	CHR	Right	Edema, pain, mild proptosis, severe ptosis, tearing, diplopia	20/20
14	M	48	3	CHR	Right	Moderate proptosis, mild ptosis	20/20
15	F	73	2	CHR	Left	Swelling of the lacrimal gland, severe ptosis, edema, conjunctival chemosis	20/20
16	M	73	8	CHR	Left	Swelling, pain	20/20
17	M	23	1	SA	Right	Swelling	20/20
18	M	23	2	SA	Left	Mass effect, proptosis, edema, conjunctiva chemosis, diplopia	20/60
19	M	63	1	CHR	Left	Pain, severe ptosis, swelling of lachrymal gland, proptosis, diplopia, hypofunction of the superior rectus muscle	20/20
20	F	58	36	CHR	Right	Swelling, diplopia, mild ptosis, mild proptosis	20/20
21	M	5	1	AC	Right	Eyelid edema, swelling, severe ptosis	20/20
22	F	21	156	CHR	Right	Moderate proptosis, diplopia	20/50
23	M	76	2	SA	Left	Eye displacement, moderate degree ptosis	20/100
24	F	32	4	CHR	Left	Swelling of the lacrimal gland	20/20
25	F	69	2	CHR	Left	Swelling, conjunctival redness and chemosis, edema, severe ptosis	20/20

M: male; F: female; CHR: chronic; SA: subacute; AC: acute.