

## Case Report

# Clinico-pathological Correlation of Inflammatory Myofibroblastic Tumor of the Eye Mimicking Nodular Scleritis: A Case Report

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*Inflammatory myofibroblastic tumor of the eye is extremely rare. The authors herein report the case of a 14-year-old Thai boy who presented with conjunctival nodule mimicking a nodular scleritis. Excisional biopsy was recommended after medical treatment failed, and histopathology of the mass revealed an inflammatory myofibroblastic tumor. There was no recurrence at 6 months following the surgery. This is the first report in Thailand of ocular IMT.*

**Keywords:** Inflammatory myofibroblastic tumor, IMT, ALK-1, Scleritis, Episcleral mass

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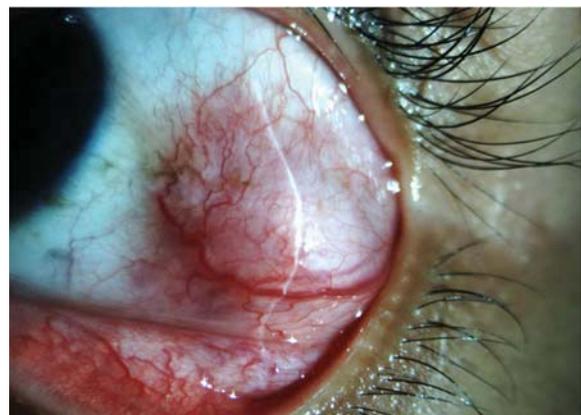
Inflammatory myofibroblastic tumor (IMT) is a benign inflammatory pseudotumor which consists of myofibroblastic spindle cells, plasma cells, lymphocytes, and eosinophils. It is found primarily in viscera and soft tissue and predominantly occurs in children and young adults. There are many synonyms for IMT including plasma cell granuloma, plasma cell pseudotumor, inflammatory myofibrohistiocytic proliferation, inflammatory pseudotumor, and inflammatory fibrosarcoma<sup>(1,2)</sup>.

The lung was the first organ in which IMT was originally described in 1990<sup>(3)</sup>. Later, it was also reported in extrapulmonary sites including the mesentery, retroperitoneum, genitourinary tract, and other organs. Ocular involvement has rarely been reported<sup>(4)</sup>, and surgical excision, which is recommended as the treatment of choice, has achieved favorable outcomes<sup>(1,2,8,9,11,12)</sup>. The authors herein report the case of a Thai boy who initially presented with conjunctival nodule mimicking a nodular scleritis, but in whom diagnosis of IMT was subsequently confirmed.

### Case Report

A 14-year-old boy was referred to our cornea clinic complaining of a slowly progressive, painless,

red-pink nodule at the inferotemporal bulbar conjunctiva of the left eye for 2 months. He had only occasional eye irritation with no eye pain, eye discharge, or red eye. He had no history of eye trauma, and he was not taking any medication. On ocular examination, the visual acuity was 20/20 in both eyes. On slit-lamp examination, a 1-cm nodular lesion was noted at the inferotemporal area of the left eye (Fig. 1). Extraocular muscle movement was normal, as was dilated fundus examination, and there was no palpable lymphadenopathy. It was initially diagnosed as a nodular scleritis, and treatment with systemic and topical steroids was initiated, but there was no response after



**Fig. 1** Pink nodular lesion located at inferotemporal area of the left eye.

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2 weeks. ANA was positive for fine speckle pattern (titer 1: 80), which was non-specific for autoimmune disease. All other laboratory results, including complete blood count, HbsAg, Anti HBs, Anti HCV, Anti-HIV, RPR, FTA-ABS IgG, rheumatoid factor, urinary analysis and chest x-ray, were within normal limits. Biopsy was then recommended.

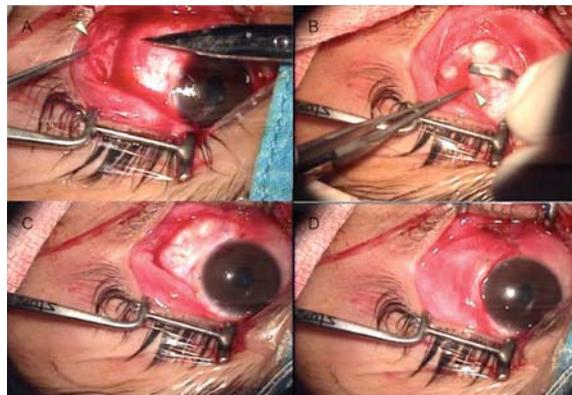
Excisional biopsy was performed under local anesthesia. Intra-operative findings revealed a relatively well-defined pink episcleral mass of about 1x2 cm attached to the sclera and muscle capsule (Fig. 2A, 3). The mass capsule was separated from the lateral rectus muscle capsule, and superficial sclerectomy was also performed to ensure complete removal (Fig. 2B). The gross tumor was totally excised, and no gross scleral invasion was noted (Fig. 2C). Simple conjunctival closure without graft was performed with cauterization and interrupted 10-0Vicryl sutures (Fig. 2D).

Histopathological examination revealed a spindle cell tumor in fibrous connective tissue (Fig. 4A). The tumor, composed of plump spindled and stellate-shaped cells arranged in fascicular and storiform patterns, appeared partly circumscribed and focally infiltrative. These cells exhibited variable amounts of pale eosinophilic cytoplasm with delicate cytoplasmic processes (Fig. 4B). They possessed plump, ovoid-shaped vesicular nuclei with occasional prominent nucleoli. Nuclear hyperchromasia or atypia was not appreciable. Mitotic figures were identified (2 per 10 high-power fields), and there was prominent inflammatory infiltrate, mainly of lymphocytes and plasma cells, throughout the tumor. Neutrophils rarely presented.

In immunohistochemistry, the spindle and stellate cells were labeled for smooth muscle actin ( $\alpha$ -SMA) and anaplastic lymphoma kinase (ALK or CD246) (Fig. 5). They were negative for Cytokeratin AE1/AE3, Desmin, Myogenin and S100. Infiltrating inflammatory cells comprised scattered IgG4-positive plasma cells (10 per high-power field with) with IgG4 to IgG ratio of less than 40.0%.

The morphologic and immunophenotypic findings were diagnosed as an inflammatory, myofibroblastic tumor.

Treatment with one percent prednisolone acetate eye drops and oral prednisolone 30 mg/day (0.5 mg/kg/day) was prescribed following the surgery; this was tapered down over 2 months. The patient was referred to the oncology clinic after pathological report, and no other systemic involvement of the tumor was



**Fig. 2** A) The arrow shows the episcleral mass of about 1x2 cm attached to the sclera and lateral rectus muscle capsule. B) Superficial sclerectomy was performed to ensure total tumor removal. C) Episcleral mass was completely removed. The sclera underneath the mass was normal without scleral invasion. D) Simple conjunctival closure was performed with cauterization and interrupted 10-0Vicryl sutures.



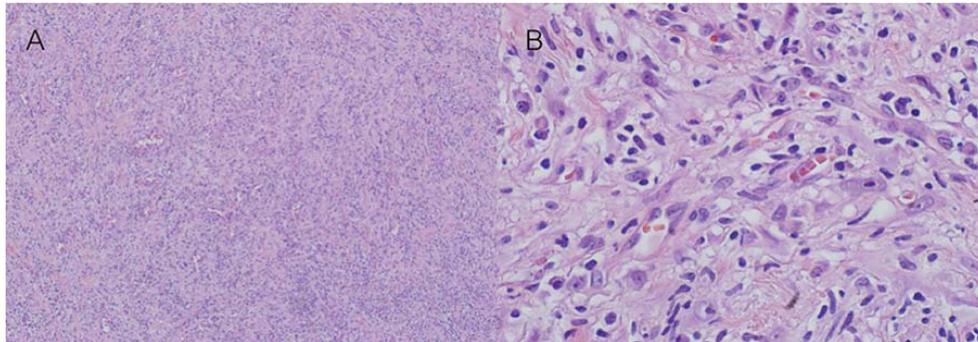
**Fig. 3** The red-pink episcleral mass with smooth surface and rubbery consistency.

detected. No adjunctive treatment was recommended, and there was no tumor recurrence 6 months following the surgery (Fig. 6).

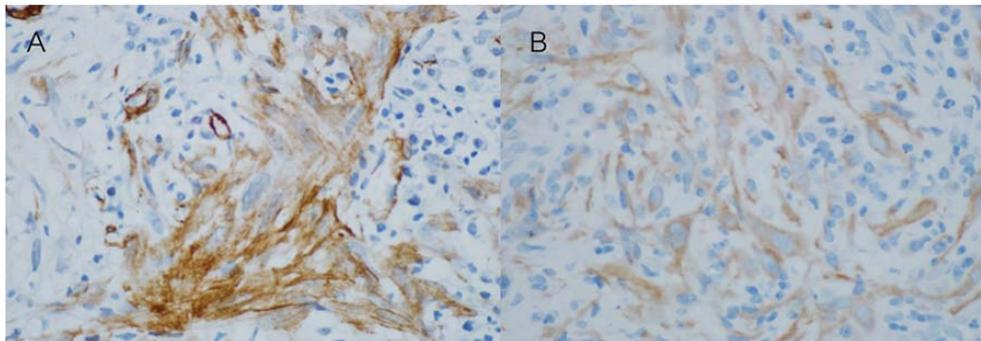
### Discussion

Inflammatory myofibroblastic tumors (IMT) were originally described in 1990<sup>(3)</sup>. The most common locations of IMT are the abdomen, pelvis, lung, and peritoneum. IMT of the eye and orbit is extremely rare. To the best of our knowledge, this is the first report of IMT of the eye in Thailand.

A literature review of 11 cases showed that



**Fig. 4** A) Intermediate magnification of mass revealed spindle cells, collagen fibrils, and inflammatory elements. Note the fascicular and storiform arrangement of spindle cells. (hematoxylin-eosin, original magnification 10x). B) High magnification revealed spindle cells with oval-shaped vesicular nuclei intermixed with lymphocytes and plasma cells (hematoxylin-eosin, original magnification 60x).



**Fig. 5** A) Staining for  $\alpha$ -SMA showed strong positivity in periphery of cytoplasm ( $\alpha$ -SMA immunohistochemistry, original magnification 60x). B) Staining for ALK protein showed diffuse, granular cytoplasmic staining (ALK immunohistochemistry, original magnification 60x).



**Fig. 6** Six months following surgery, there was no tumor recurrence.

IMT is predominantly found in males, and most commonly occurs in children and young adults, but

age range can vary from 8 months to 71 years. It can present with various characteristics such as subconjunctival mass<sup>(2)</sup>, limbal mass<sup>(4)</sup>, vitreous mass with episcleritis<sup>(7)</sup>, intraorbital mass with proptosis<sup>(10-15)</sup>, and episcleral mass. IMT has been reported involving the meninges, multiple cranial nerves<sup>(6)</sup>, and bone<sup>(8)</sup>.

Histopathological examination and immunohistochemical stain are essential in the diagnosis of IMT<sup>(2,9)</sup>, to reveal spindle cells, collagen fibrils, and inflammatory elements. The spindle cells in IMT are bland and uniform, and necrosis and calcification are rarely found<sup>(11)</sup>. The spindle and stellate cells are labeled for smooth muscle actin ( $\alpha$ -SMA).

Recent studies have reported that subsets of IMT are related to abnormalities of chromosome 2p23<sup>(2,7,8)</sup>. Deregulation of the anaplastic lymphoma kinase (ALK) gene located at chromosome 2p23 leads to the lack of ALK rearrangement suggesting ALK

overexpression<sup>(7)</sup>. IMT with ALK-1 immunoreactivity has been reported in 30% to 60% of cases and is more common in children and young adults<sup>(12)</sup>. ALK-1 positive indicates a poorer prognosis compared to those cases that are ALK-1 negative<sup>(9,15)</sup>.

Mudhar HS et al suggested that in the absence of phlebitis and lymphoid aggregates, the presence of an ALK-1 positive IMT with high numbers of IgG4 plasma cells probably represents a chronic inflammatory response to the IMT, not IgG disease<sup>(13)</sup>. In this case study, immunohistochemistry revealed IgG4-positive plasma with IgG4 to IgG ratio of less than 40%. The author suggests that the reaction responded to the IMT, not IgG disease, which can be explained by the low IgG4 to IgG ratio.

The treatment of choice is complete surgical tumor excision, which has favorable outcomes and a good prognosis. Systemic corticosteroids, chemotherapy, or radiation therapy are recommended for adjunctive therapy in cases with incomplete resection or aggressive tumor.

#### What this study adds?

This is the first report in Thailand of ocular IMT which can present mimicking a nodular scleritis in children. Total tumor excision and systemic steroid therapy following surgery had a favorable outcome with no recurrence.

#### Potential conflicts of interest

None.

#### References

1. Coffin CM, Fletcher JA. Inflammatory myofibroblastic tumour. In: Fletcher CDM, Unni K, Mertens F, editors. WHO Classification of tumours of soft tissue and bone. Lyon, France: IARC Press; 2002: 91-3.
2. Sa HS, Ji JY, Suh YL, Kim YD. Inflammatory myofibroblastic tumor of the orbit presenting as a subconjunctival mass. *Ophthalm Plast Reconstr Surg* 2005; 21: 211-5.
3. Pettinato G, Manivel JC, De Rosa N, Dehner LP. Inflammatory myofibroblastic tumor (plasma cell granuloma). Clinicopathologic study of 20 cases with immunohistochemical and ultrastructural observations. *Am J Clin Pathol* 1990; 94: 538-46.
4. Shah S, Badhu BP, Lavaju P, Pradhan A. Ocular inflammatory myofibroblastic tumor in the left eye with phthisis right eye: A rare occurrence in a child. *Case Rep Ophthalmol Med* 2015; 2015: 281528.
5. Coffin CM, Humphrey PA, Dehner LP. Extrapulmonary inflammatory myofibroblastic tumor: a clinical and pathological survey. *Semin Diagn Pathol* 1998; 15: 85-101.
6. McKinney AM, Short J, Lucato L, SantaCruz K, McKinney Z, Kim Y. Inflammatory myofibroblastic tumor of the orbit with associated enhancement of the meninges and multiple cranial nerves. *AJNR Am J Neuroradiol* 2006; 27: 2217-20.
7. O'Malley DP, Poulos C, Czader M, Sanger WG, Orazi A. Intraocular inflammatory myofibroblastic tumor with ALK overexpression. *Arch Pathol Lab Med* 2004; 128: e5-e7.
8. Tawfik HA, Raslan AO. Infantile inflammatory myofibroblastic tumor of the orbit with apical bone involvement. *Ophthalm Plast Reconstr Surg* 2013; 29: e44-e46.
9. Polito E, Pichierri P, Loffredo A, Moramarco A, Occhini R. Inflammatory myofibroblastic tumor of the orbit. *Ophthalmologica* 2007; 221: 353-5.
10. Leoncini G, Maio V, Puccioni M, Franchi A, De G, V, Ucci F, et al. Orbital solitary fibrous tumor: a case report and review of the literature. *Pathol Oncol Res* 2008; 14: 213-7.
11. Cramer SK, Skalet A, Mansoor A, Wilson DJ, Ng JD. Inflammatory myofibroblastic tumor of the orbit: a case report. *Ophthalm Plast Reconstr Surg* 2015; 31: e22-3.
12. Lauwers N, De G, V, Kenis C, Dhubhghaill SN, Claes J, De Keizer RJ. Atypical sino-orbital inflammatory myofibroblastic tumor with bone and cerebral invasion extending to the orbit. *Eur J Ophthalmol* 2014; 24: 608-10.
13. Mudhar HS, Nuruddin M. ALK-1 positive orbital inflammatory myofibroblastic tumour (IMT) associated with prominent numbers of IgG4 plasma cells - a case report. *Orbit* 2013; 32: 321-3.
14. Dutta V, Manoj MG, Malik A, Kumar P. ALK negative inflammatory myofibroblastic tumor of the orbit: a masquerading entity. *Indian J Ophthalmol* 2014; 62: 627-9.
15. Cook JR, Dehner LP, Collins MH, Ma Z, Morris SW, Coffin CM, et al. Anaplastic lymphoma kinase (ALK) expression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study. *Am J Surg Pathol* 2001; 25: 1364-71.