Inflammatory myofibroblastic tumour of the paranasal sinuses

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ABSTRACT

Inflammatory myofibroblastic tumour (IMT) is a rare tumour most commonly found in the lungs and in general follows a benign clinical course. Sinonasal IMT however, is extremely aggressive, difficult to treat, prone to recurrence and often associated with fatal outcomes. Surgery is the gold-standard treatment and in aggressive cases, multiple resections with reconstruction maybe required. We report the case of a 56-year-old man with an aggressive sinonasal IMT who required extended medial maxillectomy, orbital decompression and subsequently an orbital exenteration followed by high dose corticosteroids therapy. This case report highlights the aggressiveness of the lesion which was extremely difficult to manage, with multiple surgeries and high dose long-term corticosteroid therapy as an adjunct treatment.

Keywords: Inflammatory tumour, inflammatory neoplasm, paranasal sinus

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT) has been classified as an intermediate soft tissue tumour due to its propensity for local recurrence and less than 5% risk of metastasis. [1, 3] In the head neck region, it is commonly found in the orbit. Sinonasal occurrence is extremely rare. [2, 3] In the sinonasal region, it may present as a nonspecific mass which grow slowly over weeks or months. [3] Radiologically, it has features suggestive of a malignant process. The diagnosis of IMT can only be achieved through histopathological and immunohistochemistry examinations. Surgery is the principle treatment of IMT. Regression with chemotherapy, radiotherapy, steroid therapy and non-steroidal anti-inflammatory drugs (NSAIDS) have been reported in literature.[1, 2, 4] Sinonasal IMT is extremely aggressive, difficult to treat, prone to recurrence and has fatal outcome. This case reports on a highly aggressive sinonasal IMT and its management.

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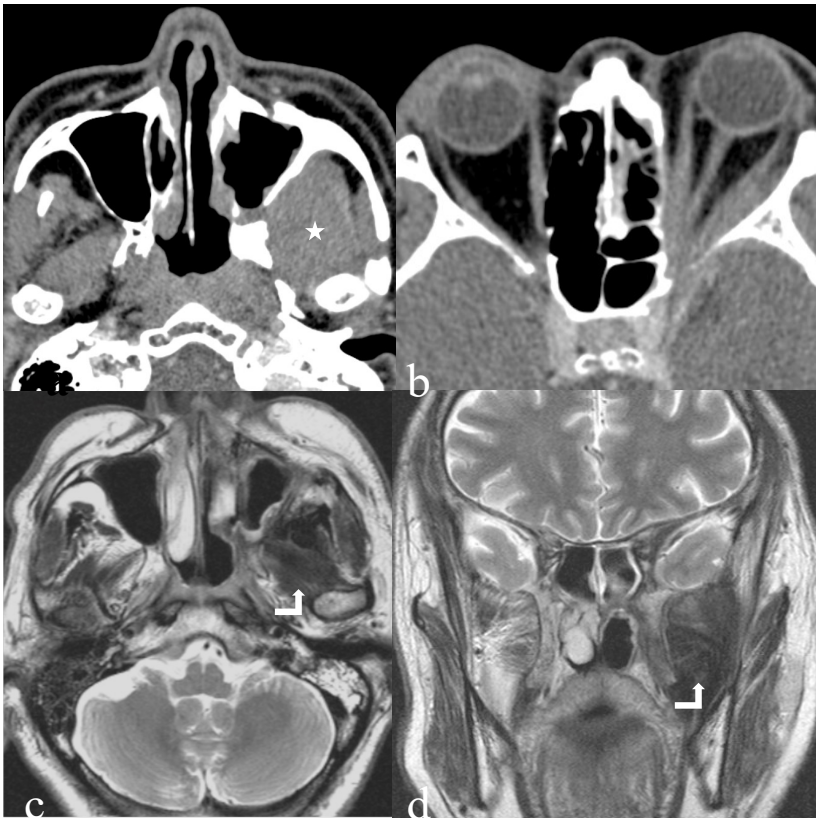
CASE REPORT

A 56-year-old man presented with an initial complaint of left upper toothache of eight months duration. He developed purulent nasal discharge with total blindness of his left eye five months later. He also had severe intermittent headaches but no facial numbness. From examination, there was left eye proptosis with the involvement of the II, III, IV, VI cranial nerves. Nasal endoscopy showed a left nasal cavity filled with copious pus and crusting. A tumour was not visualised endoscopically. Computed tomography and magnetic resonance imaging of paranasal sinuses and

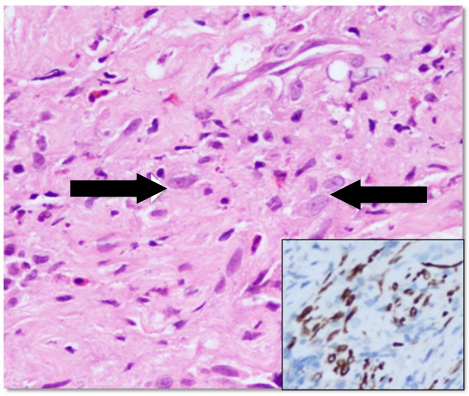


Figs. 1. Axial contrast enhanced CT of the orbit (a) and (b) and T2-weighted MRI of the brain and orbit in axial (c) and coronal (d). The enhancing mass (star) occupies the left masticator space involving the medial and lateral pterygoid muscles that extends into the left pterygopalatine fossa (short arrow). Superiorly, intraorbital extension to the retroorbital space is seen via the inferior orbital fissure (long arrow). The mass shows characteristic markedly low signal intensity on T2 compared to muscle (bent arrow).

orbits showed a poorly enhancing soft tissue mass originating from the left maxillary sinus with extension into left ethmoid sinuses, orbit, anterior temporal lobe and cavernous sinus (Figure 1). The mass extends posteriorly into masticator space. Left pterygoids and temporalis muscles were infiltrated by the tumour.

The patient underwent left endoscopic extended medial maxillectomy with removal of posterior maxillary wall which revealed tumour in the infratemporal fossa. The left lamina papyracea was then removed for orbital decompression. Histopathological examination revealed the diagnosis of inflammatory myofibroblastic tumour (Figure 2). The tumour was positive for vimentin, desmin and actin and negative for myogenin and S100 consistent with a diagnosis of IMT. .He was started on high dose prednisolone (75 mg divided dose daily) which was tapered down over one month period.

He was well until two months postoperatively when he complained of left eye pain. Prednisolone was restarted with the dose of 60mg daily that was tapered over two weeks. Unfortunately, despite the steroid



Figs. 2: Arrows indicate scattered myofibroblasts against a background of inflammatory cells, composed of lymphocytes, plasma cells and eosinophils. Insert shows myofibroblasts expressing strong desmin positivity.

therapy and radiological evidence response to treatment, the severe pain and blindness secondary tumour infiltration led to the exenteration of his left eye. He was then restarted on oral prednisolone 60 mg daily which was tapered over two months period. Nine months follow up showed that the outcome of the corticosteroid therapy given for a longer term was good and patient had no evidence of tumour recurrence.

DISCUSSION

In 2002, World Health Organisation (WHO) described inflammatory myofibroblastic tumour (IMT) as "a distinct lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils", synonymous with inflammatory pseudotumour. [1] It is presumed as an acute infective response, post inflammatory reparative process and low grade spindle cells malignancy. [2, 5] *Epstein-Barr Virus (EBV)*, *Human Herpes Virus*-8 (HHV8), Mycobacteria, Actinomycetes, *Escherichia coli*, Klebsiella species, Pseudomonas species, *Human Immunodeficiency Virus* (HIV), and *Helicobacter pylori* are among the pathogens associated with IMT. [3]

IMTs are more common in female and has a predilection in children and young adults in contrast to our elderly male patient. [1, 3, 5] Most commonly found in lungs, IMTs of head and neck are considered rare. [3] In general, IMT follows a benign clinically course but those of maxillary sinuses are found to be aggressive and cause extensive tissue destruction. [3, 6] It may present with nasal congestion, epistaxis, proptosis, headache, pain, paraesthesia, dysphagia and cranial nerve dysfunctions. [2, 7]

Endoscopic examinations will reveal a swelling covered by normal mucosa or oedematous mucosa, hypertrophic inferior conchae, grey-tanned exophytic nodular or polypoidal lesion that may extend deep into structures adjacent to its site of origin. [6]

Both CT and MRI of the head and neck commonly suggest granulomatous tissue or infiltrative growth with aggressive malignant potential. When aggressive, IMT may show erosion, remodelling or sclerotic bony changes which mimics malignant tumours. [2, 6] There was evidence of tumour extension in association with root resorption which may cause the initial complaint of toothache as described in our case. [3]

Microscopically, IMT is conformed into three basic patterns such as; 1) myxoid/ vascular pattern; 2) nodular fasciitis-like pattern and; 3) fibrous scar like pattern. [3, 6] For Immunohistochemistry, the cells are characteristically reactive to vimentin (99%), smooth muscle actin (92%), muscle specific actin (89%) and desmin (69%). [3] Myoglobin and S100 are typically negative in IMT. [1, 3] In our patient, vimentin are positive throughout the tissue with dual positivity of desmin and actin and negative for myogenin and S100 points to the diagnosis.

The gold standard treatment for head and neck IMT remains surgical resection with negative margins due to its erosive nature. [5] Response and regression to corticosteroids and NSAIDS have been reported. [1] In early lesions which includes high counts of lymphoid follicles, high doses of corticosteroids have often proved to be favourable in oppose to the mature lesions with more fibrous content. [2, 3] Thomas et al reported a case of maxillary sinus IMT successfully treated with surgery and oral prednisolone 60mg/day for six weeks. [8] In a case of laryngeal IMT, Zitsch *et al.* suggested that the best approach is local excision followed by high dose steroid for 6-12 weeks. [9] Radiotherapy has been given for recurrent and unresectable cases. [3]

Paranasal IMTs are very aggressive with poor response to treatment, with multi-

ple recurrence and has fatal outcome. [3] Extrapulmonary IMTs have a local recurrence rate of 25% in relations to location, resectability and multinodularity. [1, 3] In a study of 25 patients, 15 of 20 patients (75%) with follow-ups of between six and 120 months after diagnosis had tumour recurrence. [10] Five patients had one episode of recurrence, six patients had recurrence two to three times and four patients had ≥4 recurrence episodes. [10]

WHO has classified soft tissue tumours into benign, intermediate and malignant biological potential. [1] IMT has been categorised as having intermediate (rarely metastasising) biological potential. In this category, tumours are often locally aggressive but can give rise to distant metastases in infrequent cases. [1] Metastasis occurs in less than 5% of cases. [1] Metastasis sites includes the brain and lungs. [3] Gale *et al*. reported the first case of paranasal sinus IMT with a fatal outcome in a 22-year-old woman with extensive intracranial spread. [11] In case series of 59 patients with IMTs, mortality rate was reported 10% (n=6). [12] Five of the deceased had local recurrence and one had metastatic IMT. [12] Salehinejad *et al*. suggested strict follow up starting from 1-2 monthly for the first two years, 2 -3 monthly following year and six monthly in the 4th and 5th year. [3]

In conclusion, sinonasal IMTs are highly aggressive, prone to local recurrence and are associated with fatal outcome in 10%. Diagnosis may be difficult due to its multiple patterns and immunohistochemistry is required for confirmation. Surgical resection with clear margins remains the gold standard and it is noted to show response to long term steroids. We support the additional treatment of corticosteroids 60 mg/day for 6-12 weeks to help halt the disease's progression and possible recurrence. Metastases is rare occurring in less than 5%. Despite this, close follow -up is recommended for all patients.

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