Successful treatment of inflammatory myofibroblastic tumor by surgical resection and radiotherapy: A case report

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► ***Case report***

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ABSTRACT

Background: Inflammatory myofibroblastic tumor (IMT) is a tumor composed of myofibroblasts and a mixed inflammatory infiltrate that rarely undergoes malignant transformation. The most common sites of involvement include the lung, liver and orbit, but it has been reported to occur in nearly every site of the body, including oral cavity and soft tissue. Although common in other sites, such a tumor in rarer location including inguinal region is likely to be missed/misdiagnosed and hence need to be reported. ***Case Report:*** We present an unusual case of a 50 year old male with bilateral inguinal swelling attended our institute few days back with history of local excision twice, but still showing recurrence. He was treated by wide local excision with bilateral orchiectomy this time. Histopathological report revealed inflammatory myofibroblastic tumor with malignant transformation. Adjuvant radiotherapy was also given and patient taken in followup. At one year of serial follow-ups, the patient is disease-free both clinically and on investigations, indicating successful combined surgery and radiotherapy in the treatment of malignant IMT. ***Conclusion:*** IMT of the inguinal region is very rare. Its clinical presentation may resemble malignant tumor, so IMT should be kept as differential diagnosis. The recommended treatment is complete surgical resection with adjuvant corticosteroid. Adjuvant radiotherapy may be considered depending upon the location of the tumor.

Keywords: *Inflammatory myofibroblastic tumor (IMT), surgery, adjuvant radiotherapy.*

INTRODUCTION

Inflammatory Myofibroblastic tumor (IMT) is a rare neoplasm consisting of variable numbers of inflammatory cells and myofibroblastic spindle cells. It was first observed in the lung and described by Brunn in 1939 and was so named by Umiker *et al.* in 1954 because of its clinical and radiological behavior that mimics a malignant process [(1)]. The most common sites of involvement include the lung, liver and orbit, but it has been reported to occur in nearly every site of the body, including oral cavity and soft tissue [(1-4)]. The tumor is rare at other sites including inguinal region likely to be missed or misdiagnosed, both of which are detrimental to the patient and hence need to be reported. The etiology and pathogenesis of IMT remains unclear. Signs and symptoms depend on specific location of the tumor.

CASE REPORT

A 50 years old male presented to our department with bilateral inguinal swelling. Initially the patient noticed a painless swelling in right inguinal region two years back. The swelling gradually increased in size. Then the swelling was excised and after three months of surgery, patient was giving the history of recurrence and re-excision was done twice at the same site at the peripheral hospital. Nearly six months back patient again developed recurrence in right inguinal region and this time it was associated with a nodular swelling in left inguinal region. Gradually both the swellings increased in size and converted into fungated mass with bleeding ulcer. The patient denied any history of local trauma or infection.

On investigations, laboratory tests like complete blood count, renal function tests and liver function tests were unremarkable. Serum markers α-fetoprotein, β-human chorionic gonadotropin, and lactate dehydrogenase levels were also within normal limits. Preoperative sonography was suggestive of multiple small to large hypoechoic masses noted in bilateral l inguinal region which were matted together largest measuring 5.6 × 5.3 × 6.2 cms suggestive of possibility of nodal mass. As the tumor recurred thrice after attempts at resection, wide local excision and bilateral orchiectomy was planned.

Histopathological report of specimen revealed a spindle cell neoplasm with prominent eosinophilic nucleolus. The tumor was arranged in fasciles and showed a prominent lymphocytic infiltrate (figure 1; figure 2) and was suggestive of malignant inflammatory myofibroblastic tumor. The tumor cells were immunonegative for desmin, CD 34, CD 31, HMB 45, S-100 protein, CD 21, CD35, c-kit, cytokeratin, EMA, CD 30 and ALK-1.

In view of this, fractionated conformal radiotherapy (FRT) was planned; 60 Gy in 30 fractions of 2 Gy over 6 weeks were delivered. The follow up of the patient was done for 1 year after surgery with adjuvant radiotherapy. On serial follow up visits, on the basis of clinical examination and investigations no evidence of recurrence was documented.

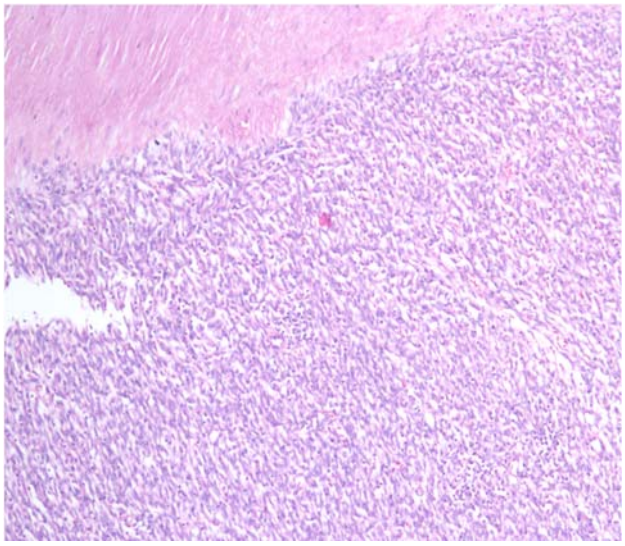

Figure 1. 10x; H & E stain.

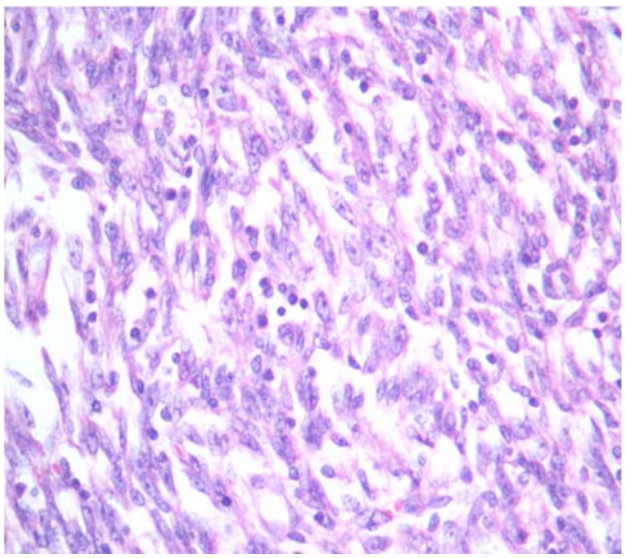

Figure 2. 40x ; H & E stain.

H & E stain (figure 1 & 2) showing a spindle cell neoplasm with prominent eosinophilic nucleolus. The tumour is arranged in fasciles and shows a prominent lymphocytic infiltrate.

DISCUSSION

IMTs are tumors with unpredictable clinical behavior, requiring complete surgical excision and continuous monitoring of clinical

consequences [1]. According to the World Health Organization IMTs are classified as tumors of intermediate biological potential due to a tendency of local recurrence and small risk of distance metastasis [5]. Inflammatory myofibroblastic tumor is a soft tissue lesion that may be confused with a sarcoma. The symptoms depend on the specific location of the tumor, which can be anywhere in the body.

This tumor has been primarily described in the soft tissues and viscera of children and young adults [6], with equal incidence in male and female patients [7]. Despite an apparently benign morphological nature, they have been reported to have locally aggressive growth. Locally recurrent and metastatic forms have been reported [8].

Most IMTs are grossly poorly defined gray-white nodules, which may be gelatinous or firm. Occasionally, hemorrhage and cystic changes may be seen. They are composed of dominant spindle cells, which are proliferated with a variable inflammatory component. These spindle cells are myofibroblasts, and this is the reason for the current designation of this disease. High cellularity showing a preponderance of inflammatory components is often seen in the early phases of the disease. The mature lesions are less cellular and contain collagen bundles.

In many cases, IMTs seem to represent a reactive condition, although there is evidence that a subset is clonal. It has recently been established that many IMTs have abnormalities of chromosome 2p23 and express ALK1 (anaplastic lymphoma kinase 1) [9].

An IMT may be a locally aggressive and destructive neoplasm. Tumor recurrence is unusual after complete surgical resection or organ-preserving combined modality therapy [10]. When surgery fails, radiotherapy, methotrexate, and corticosteroids have been used to treat recurrence [11].

After extensive search, we did not find any literature on IMT of inguinal region. Series of D K Lee et al showed aggressive clinical behavior of IMT involving the skull base and its poor response to steroid therapy and low-dose radiation therapy. For improving control, more aggressive initial efforts including high-dose radiation therapy with or without concurrent steroid therapy might play a role [12]. In a recent report by Jean-Philippe Maire *et al.*, fractionated conformal radiotherapy (FRT) was indicated at a low dose; 20 Gy in 10 fractions of 2 Gy over 12 days were delivered in a case of IMT of skull base. In this report, clinical response was complete 3 months after FRT, radiological response was subtotal 6 months after FRT and after two years, the patient was well and symptom-free [13].

IMT in the inguinal region may be confused with other malignant tumors on clinical, radiographic, and histologic appearance. Therefore, it is important to recognize the distinction among them in order to provide better guidelines for treatment and outcome. So after reviewing the articles on IMT of other sites, adjuvant radiotherapy was planned in our case.

CONCLUSION

IMT of the inguinal region is very rare. Its clinical presentation may resemble malignant tumor, so IMT should be kept as differential diagnosis. The recommended treatment is complete surgical resection with adjuvant corticosteroid. Adjuvant radiotherapy may be considered depending upon the location of the tumor. Although we did not find any literature on IMT of inguinal region but considering the tumor location and reviewing the articles on IMT of other sites, we have delivered radiotherapy in our case. Even though the efficacy of such treatment is debatable, further case reports are needed to ascertain the optimal therapeutic regimen.

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Conflict of interest: *Declared none.*

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