A paratesticular inflammatory myofibroblastic tumor and review of the literature

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1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a pseudosarcomatous inflammatory lesion originating from soft tissue or visera. Although it was initially reported in thoracic cavity, it has been documented in a variety of extrapulmonary sites, ranging from brain to bladder.1 Genitourinary IMTs have been reported, accounting 9% of extrapulmonary IMTs. In addition, paratesticular IMTs have been rarely reported in the literature.2–6 We present a young male patient who presented with paratesticular IMT and review of the literature.

2. Case report

A thirty eight year old male patient without any comorbidities, was presented with scrotal pain and lump. There was no trauma, fever, symptoms of infection. On physical examination a 4 × 3 cm non–tender, firm mass was palpated at the posterior of right testis. An inguinal hernia was detected. The skin of scrotum was normal and there were no pathological inguinal lymph nodes. Ultrasonography revealed a 4.5 × 3.5 cm ovoid heterogeneous mass related with epididymis. Abdominal imaging was normal. AFP, BCG ve LDH levels were normal. Patient was treated with right orchiectomy, resection of mass and and repair of inguinal hernia. Pathological examination showed a myofibroblastic proliferation related with epididymis (Fig. 1). There were inflammatory cells, predominantly plasma cells and lymphocytes. There was a sclerotic fibrous stroma consisting of vascular structures and mononuclear cells. Immunohistochemistry was positive for actin, desmin, vimentin and negative for cytokeratins (AE1/AE3) and S100. Pulmonary and abdominal computed tomography was normal. Patient was followed without any further treatment. Patient has been followed with 3 monthly intervals and a workup with physical examination, scrotal/abdominal ultrasonography and chest x-ray. After 1 year of follow up, patient is free of recurrence.

3. Discussion

Inflammatory Myofibroblastic tumor (IMT) is a rare spindle cell proliferation which has been reported in the literature with different designations, such as inflammatory pseudotumor, plasma cell pseudotumor, xanthomatous pseudotumor, atypical myofibroblastic tumor, atypical fibromyxoid tumor, pseudosarcoma, plasma cell granuloma, and fibrous pseudotumor.4 It is composed of myofibroblastic mesenchymal spindle cells with an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils.7 The pathology has been reported in different organs, such as lung, skin, soft tissues, breast, gastrointestinal tract, pancreas, oral cavity, bone
and central nervous system.\textsuperscript{3}

Genitourinary system is a rare location for this rare pathology. In the genitourinary tract, IMT has been reported in the kidney, urethra, prostate, ureter, and testis but is most frequently observed in the bladder. IMT has been reported in paratesticular structures in fewer than 10 cases in literature. The exact etiology is not known, but ischemia, infection and chronic irritation have been postulated as etiological factors.\textsuperscript{8} Painless mass in scrotum is the usual presentation of paratesticular IMT. It has been most commonly reported in children or young adults. However, it has also been described in an elderly male patient.\textsuperscript{9} Ultrasonography of the scrotum should be the initial imaging modality to differentiate testicular masses from paratesticular ones. During evaluation of paratesticular mass, more common pathologies should be considered. Varicocele, spermatocele, infections and primary malignant diseases should be excluded.\textsuperscript{5}

According to cases, paratesticular IMT is considered as a benign pathology. Although IMT series in different organs have a 25% recurrence rate, aggressive and locally invasive paratesticular IMT has been rarely reported.\textsuperscript{10} Thus, lesions occurring in the retroperitoneum, mesentery or mediastinum are considered potentially malignant and could recur after treatment. Optimal treatment strategy is wide resection of the mass with conserving testis. Due to benign nature of the disease and rare recurrences, adjuvant chemotherapy or radiotherapy are not recommended. In recurrent cases, resection of the mass is recommended. Radiotherapy or chemotherapy with methotrexate or steroids could be used.\textsuperscript{10} Rearrangements involving the ALK locus on chromosome 2p23 have been documented in approximately 50% of IMTs. Distant metastases occur primarily in ALK-negative IMTs, but local recurrence occurs regardless of ALK expression.\textsuperscript{11} In recurrent ALK positive cases, Crizotinib is an effective treatment modality.

In conclusion, our case had similar clinical and pathological characteristics with other cases in the literature. The patient was free of recurrence after 1 year and still under close follow up. Paratesticular IMT is a rare pathology and should be considered in the differential diagnosis of paratesticular masses. Although it has a benign nature, due to inadequate experience and recurrence potential in other organs, patients should be followed after surgical resection.

References