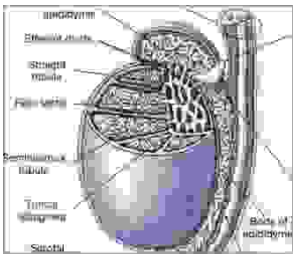
CASE REPORT PROF-847

PARATESTICULAR LEIOMYOSARCOMA



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ABSTRACT ... We are reporting the case of a 44-year old patient with paratesticular leiomyosarcoma, which was initially reported to be leiomyoma on testicular biopsy. Due to persistence of symptoms, his orchidectomy was done. On histopathological examination the tumour proved to be leiomyosarcoma arising from tunica albuginea. Operation was followed by adjuvant chemotherapy and the patient is presently asymptomatic.

Key Words: Paratesticular Tumor, Leiomyosarcoma

INTRODUCTION

Leiomyosarcoma arising from the paratesticular tissues is a rare malignant tumour[1]. Paratesticular sarcoma can arise from tunica albuginea or tunica vaginalis[2] in addition to other fascial layers. It should be differentiated from a pure intra-testicular leiomyosarcoma, which arises from the stroma of testis and is exceedingly rare[3]. Clinical behaviour of the tumour is difficult to predict. It may run an indolent course or may metastasize early[1]. Diagnosis is usually histopathological and immunohistochemical. This tumour, though rare should be included in the differential diagnosis when managing a case of testicular swelling.

CASE REPORT

A 44 years old male reported to the surgical outpatient, with history of pain in the testis of 02 years duration. The pain was dull, localized, and got aggravated by exertion or prolonged standing and relieved by rest. There was no history of weight loss. Clinical examination revealed a male of weak build, whose pulse rate was 80/min and BP was 130/90mmHg. There was no cervical or inguinal lymph-adenopathy. Abdominal examination was unremarkable. A 10 x 8 x 7 cms intra-scrotal, ovoid swelling was present, which was firm, non-tender and non-translucent.

Laboratory investigations revealed his hemoglobin to be 10.4 grams/dl. Blood chemistry was within normal limits. Ultrasonography of the abdomen revealed no para-aortic lymphadenopathy. FNA of the mass was carried out under ultrasound guidance. It showed a hypocellular aspirate consisting of a few

scattered fragments and isolated spindle shaped cells of benign morphology. The cytologist gave an opinion of "suspicious for a smooth muscle tumor" and an advice for open biopsy to reach a definitive diagnosis.

Scrotal exploration revealed a firm growth arising from the epididymo-testicular junction. An excision biopsy was performed and wound was closed in layers. Histopathological examination of the specimen revealed it to be leiomyoma of the testis. Patient made an uneventful recovery and was discharged. He remained asymptomatic for about 06 months when he started having dull testicular pain. Examination revealed a firm, non-tender swelling, 2 x 1cm in size, at epididymo-testicular junction. He was treated with analgesics and was reassured. However he continued having symptoms and after 06 months of initial admission he was readmitted and right orchidectomy was performed through inguinal approach. Histopathological examination of the testis revealed it to be leiomyosarcoma grade II arising from the tunica albuginea of testis. There was no evidence of involvement of spermatic cord. CT scan and ultrasonography of abdomen was unremarkable. Patient was seen by oncologist and had six courses of adjuvant chemotherapy including Mesna, Adriamycin, I-phosphamide and Decarbazine (MAID regime). At present he is under regular follow-up by the oncologist and is symptom-free.

DISCUSSION

Paratesticular leiomyosarcoma is a very rare tumor. The mean age of all reported cases is 51 years. It is widely accepted that paratesticular leiomyosarcomas represent neoplastic proliferations of primitive mesenchymal cells with committed smooth muscle differentiation.[4] Leiomyosarcomas are the most common soft-tissue sarcomas of the gastrointestinal (GI) tract and uterus. Leiomyosarcomas of the retroperitoneum and the vena cava occur most commonly in women. Tumors originating in other large veins affect men and women equally. Cutaneous and subcutaneous leiomyosarcomas generally affect men. Other infrequent sites are epididymis, testis, urinary bladder, vagina, meninges, and orbital cavity.[5] Because these neoplasms can arise from the walls of arteries or veins, they can occur anywhere in the body.

Leiomyosarcomas usually present as firm masses. Their growth is apt to be insidious because they seldom cause pain or interfere with function. Therefore, although patients may notice a swelling or a definite lump, they may pay no attention to it until the tumor has increased in size over a period of months. In the absence of explosive growth, both the patient and the clinician may be slow to react, and the physician may misdiagnose the lesion as a cyst, benign tumor, hematoma, or abscess. This lack of concern is reflected in an average delay of 6 to 12 months from the onset of symptoms to an accurate diagnosis.

The differentiating features between leiomyoma and leiomyosarcoma include frequency of mitosis and nuclear atypia[5,6]. The histological findings of our case were typical for a smooth muscle tumor. The degree of pleomorphism and mitotic rate indicated a leiomyosarcoma. These sarcomas usually spread via the blood stream and less frequently through lymphatics. They metastasize preferentially to the lungs, except those arising in the GI tract, which often metastasize to the liver. The American Joint Committee on Cancer (AJCC) has developed a clinicopathologic staging system that depends primarily on the grade and size of soft-tissue sarcomas[7]

Myogenic tumors of testis are very infrequent. These days they are considered under the heading of stromal tumors, which is a group of neoplasms encompassing leiomyoma, leiomyosarcoma and epithelioid tumours[8]. Sex-cord-stromal tumors (SCSTs) of the testis comprise predominantly of spindle cells and can be difficult to classify. Ultra structural features of SCSTs must be considered while making a diagnosis. Taken together, the histologic, histochemical, immunohistochemical, and ultrastructural features of these tumours help in establishing a diagnosis[9].

Treatment is radical resection followed by adjuvant chemotherapy. The single most important prognostic variable for soft-tissue sarcomas is the histopathologic grade. The 5-year survival rate for soft-tissue sarcomas arising in various anatomical sites is similar when corrected for grade, except for intra-abdominal and retroperitoneal tumors, which tend to be large and to invade vital organs[10].

In conclusion, it should be emphasized that although paratesticular soft tissue malignancy is a rarity, it should be kept in mind while managing an intrascrotal lump. It is also necessary to obtain maximum representative slices for histopathology in order to avoid missing the diagnosis. The number of mitoses, the degree of pleomorphism, and presence or absence of invasion to adjacent structures are the key to clinching the diagnosis as well as for grading of the tumor.

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