Letter to the Editor

Leiomyosarcoma of penis

Sir,

Sarcomas represent about 1% of all malignant tumors. Primary mesenchymal tumors of penis are rare. Most of them are of vascular origin. In the present study, the authors report two cases of primary leiomyosarcoma of penis.

A 26-year-old male patient presented with progressively increasing painless swelling at the base of penis since 3 months. On examination, it was a 3cm × 3cm firm swelling at the penoscrotal junction, free from underlying pubic bone and no groin lymphadenopathy. Magnetic resonance imaging revealed a 4.7 cm × 3.7 cm × 5.4 cm lobulated soft-tissue mass involving left corpus cavernosum without lymphadenopathy. Fine needle aspiration cytology suggested a spindle cell tumor. Mass was excised through an inguinoscrotal incision taking margin of left corpus cavernosum. No lymphadenectomy was done due to N-0 status. The histopathology report was high grade leiomyosarcoma with immunohistochemistry positive for (Desmin, smooth muscle antibody [SMA]), Calponin and negative for myoglobin and Myo-D1 [Figure 1]. Margins of the resections were free, closest being 2 cm. No adjuvant therapy was given. Patient is disease free at present after 2 years of follow-up.

A 38-year-old male patient presented with multiple lesions over penis since 8 months. On examination, he had fungating mass over the glans of 3 cm × 4 cm size involving urethral meatus with three separate mobile nodules over proximal shaft of penis [Figure 2]. There were no groin nodes. Punch biopsy revealed high grade leiomyosarcoma with immunohistochemistry positive for SMA and desmin and negative for S-100, CD34, Myogenin. Patient underwent total penectomy with perineal urethrosthomy. No lymph node dissection was done in view of N-0 status. Patient is disease free at 9 months of follow-up.

Soft-tissue sarcomas (STS) of the genitourinary (GU) tract are relatively rare, accounting for 2.1% of STS’s and 1-2% of all malignant GU tumors. Leiomyosarcomas occur more frequently as compared with liposarcomas and rhabdomyosarcoma.

Penile sarcomas represent less than 5% of penile malignancies. They comprise of endothelial cell sarcomas (Kaposi’s sarcoma, epithelioid hemangioendothelioma, angiosarcoma) and less frequently rhabdomyosarcoma and leiomyosarcoma. Leiomyosarcoma represents approximately 5-6% of penile sarcomas.

First case was described by Levi in 1930. The age range at diagnosis for penile leiomyosarcoma is from 6 years to the late 80s with the highest incidence in fourth and fifth decade.

Pratt and Ross classified penile leiomyosarcomas as superficial and deep depending upon invasion of the tunica albuginea. Histologically the two types are identical.

Superficial lesions arise from the dartos muscle layer, the piloerector complex and the muscular walls of superficial vessels situated outside the albuginea. They present as a small tumor in the distal shaft or the penile prepuce, often in middle-aged men. These are slow growing tumors that are likely to recur locally with fewer propensities for metastases.

Deep lesions arise from the proximal portions of the corpora cavernosa or corpus spongiosum and occur at a relatively later age. Clinically they are poorly circumscribed, firm, non-tender masses that infiltrate surrounding tissues and can also cause urinary obstruction. Deep lesions show a greater propensity to metastasize and have a poorer prognosis.

Sarcomas usually spread by hematogenous route. Lymph node metastases is rare and seen in disseminated disease.

On gross section leiomyosarcomas are well-circumscribed with rubbery consistency. Microscopically, spindle shaped
smooth muscle bundles arranged into interlacing fascicles are seen. On electron microscopy, myofibrils, dense bodies and abundant pinocytic vesicles are noted with a continuous basal lamina. The literature shows that mitotic rate and degree of differentiation are reliable in order to predict the tumor propensity to infiltrate the adjacent structures or to metastasize.

Treatment of this rare malignancy is predominantly surgical, either wide excision or amputation; however, the approach should be individualized. Complete resection is associated with better survival. Additional histological parameters such as tumor growth pattern (circumscribed and uninodular vs. infiltrative and/or multinodular), mitotic count (>10 mitotic Figure/10 high power fields) and grade three histology can be studied in the future to better prognosticate these tumors.

Penile leiomyosarcomas are rare tumors and have a poor prognosis when deep-seated. The analysis of prognostic factors can help to identify patients at higher risk for disease progression. Surgical treatment provides the best chance of cure, with additional therapies required in few patients.

Regional lymph node dissection is usually not indicated in the absence of clinicoradiologically apparent lymph-node metastases as nodal involvement is uncommon.

Adjuvant radiation and chemotherapy have no proven value in treatment of primary. Pre- or post-operative radiation has not proved its value in reducing loco-regional recurrences or in increasing survival rates. Radiotherapy has been used for palliation, with chemotherapy being reserved for cases of disseminated disease. In the series by Russo et al. of adult urological sarcomas, no patient with disseminated disease was fully responsive to the use of several chemotherapy regimens.[2]

Local recurrence seems to be a frequent phenomenon and leiomyosarcomas become more undifferentiated with each recurrence. The recurrence rate is similar for superficial and deep lesions, 23% and 29% respectively, but the metastatic potential is higher in deep seated lesions (50%).[8]

The metastatic potential also varies according to the size of lesions, 29% for <5 cm and 50% for >5 cm. The most frequent sites of distant metastases are the lungs, liver and brain. Large tumors especially those located at the root of the penis, often have a poor prognosis despite aggressive surgical intervention.[8]

References


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Figure 1: Leiomyosarcoma comprised of pleomorphic spindle cells with mitotic activity (encircled)

Figure 2: Clinical photograph