CASE REPORT

SUBCUTANEOUS LEIOMYOSARCOMA OF SUPRA-PUBIC REGION MANAGED WITH WIDE LOCAL EXCISION AND TOTAL PENECTOMY

Syed Muhammad Nazim[1], Ayesha Nusrat[2], Ahmad Bashir[1]

[1]Department of Urology, [2]Department of Pathology, Aga Khan University, Karachi-Pakistan

Subcutaneous leiomyosarcomas (LMS) are rare soft tissue sarcomas arising from small-to-medium-sized blood vessels. Involvement of the anterior abdominal wall is extremely rare. We present a case of a 72-year-old gentleman who presented with 3 months history of a progressively increasing mass in the supra-pubic region reaching the root of the penis without any regional lymphadenopathy. Magnetic resonance imaging (MRI) pelvis showed a large heterogeneous mass in the lower abdomen indenting the penile corpora. Complete pathological clearance was achieved by wide excision of the mass with total penectomy and cutaneous urethrostomy. The patient received adjuvant radiation therapy and is free of local recurrence or distant metastasis two years after the surgery.

Keywords: Leiomyosarcoma; Subcutaneous; Abdominal wall; Surgery; Penectomy

Citation: Nazim SM, Nusrat A, Bashir A. Subcutaneous leiomyosarcoma of supra-pubic region managed with wide local excision and total penectomy. J Ayub Med Coll Abbottabad 2023;35(3):490–2.

DOI: 10.55519/JAMC-03-11083

CASE REPORT

A 72-year-old gentleman known case of chronic obstructive pulmonary disease (COPD) presented to the urology clinic with 3 months history of a mass in the lower abdomen associated with pain at the root of the penis. The mass was progressively increasing in size and did not show any change in posture or cough. There was no other lump elsewhere. The patient denied a history of lower urinary tract symptoms (LUTS), fever or weight loss and had no history of exposure to industrial chemicals. On examination, a mobile, firm hard irregular nodular lump was identified extending from the supra-pubic region to the root of the penis, especially on the left side with normal overlying skin. The penile shaft was normal and the cough impulse was negative. No regional lymphadenopathy was appreciated.

The haematological and biochemistry investigations were within normal limits. Magnetic resonance imaging (MRI) pelvis showed a heterogeneous exophytic midline mass of approximately 6.6×6.2×5.5 cm in the lower anterior abdominal wall reaching down and causing indentation of penile corpora. The tumour was not involving underlying bones or muscles. The rest of the abdomen and pelvis was normal with no inguinal or pelvic lymphadenopathy. (Figure-1) Computerized tomography (CT) scan chest showed lung changes consistent with COPD with no evidence of metastasis. A trucut needle biopsy of the mass was taken which showed spindle cell neoplasm. The case was discussed in a multidisciplinary tumour board meeting; a Sarcoma surgeon and plastic surgeon were also taken on board. The patient underwent wide-margin excision of the mass with total penectomy and cutaneous perineal urethrostomy under general anaesthesia. A Mercedes Benz incision was made extending about 2.5–3 cm around the mass. All subcutaneous tissue around and at the base of the tumour was excised reaching either side of the root of the penis. Tumour was dissected in toto and was found fixed to the corpus cavernosum on the left side; however, it was free from underlying bony symphysis. Urethra with corpus spongiosum mobilized from corpora cavernosa, neurovascular bundles tied. Corpora cavernosa were transfixed till the origin of crura at the pubic bone. Urethra was transacted, haemostasis was secured and margins from tumour base and urethra were sent for frozen section analysis which came out to be negative. A perineal urethrostomy was made on the dependent position in the perineum from the urethral stump and the wound was closed primarily in layers. Closed suction drainage was placed. (Figure-2) The excised mass measured 10×8.5×8 cm in size with an attached penis. On microscopy, it showed a neoplastic lesion with a fascicular growth pattern. The cells were round to spindle, having vesicular nuclei with inconspicuous nucleoli and moderate eosinophilic cytoplasm. Necrosis was seen & 40 mitoses/10 high power fields (HPF) were identified. The urethral margin and all resected tumour margins were tumour free. Immunohistochemistry (IHC) was strongly positive for Smooth muscle actin (ASMA). Other markers such as H-caldesmon, Desmin, Myogenin, CD 34, EMA and Sox-10 were found to be negative. Overall Features were consistent with Leiomyosarcoma, grade III, according to FNCLCC (Federation Nationale des Centers de Lutte Contre le Cancer) grading system for soft tissue sarcomas. (Figure-3) The patient's post-operative course was unremarkable. Owing to the large size (>5 cm) and high grade of neoplasm, the multidisciplinary team recommended external beam radiation therapy (EBRT) for the tumour bed. After two years of surgery, the patient is doing well with CT scan chest, abdomen & pelvis showed only bronchiectatic changes in lungs with no evidence of disease recurrence or metastasis.

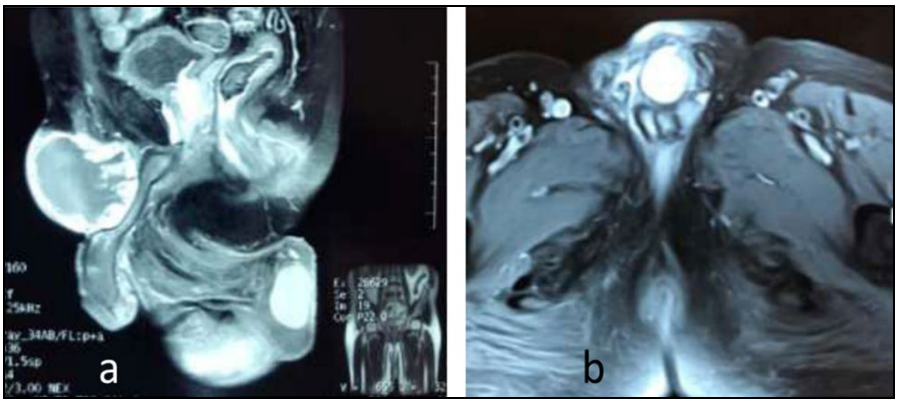

Figure-1: Magnetic resonance imaging.

A): T-1 weighted, sagittal section showing large heterogeneous lesion, protruding out from the lower anterior abdominal wall. Posteriorly the lesion is compressing the crura.
B) T-1 weighted, post-contrast axial section showing mass abutting the crura, more on the left side.

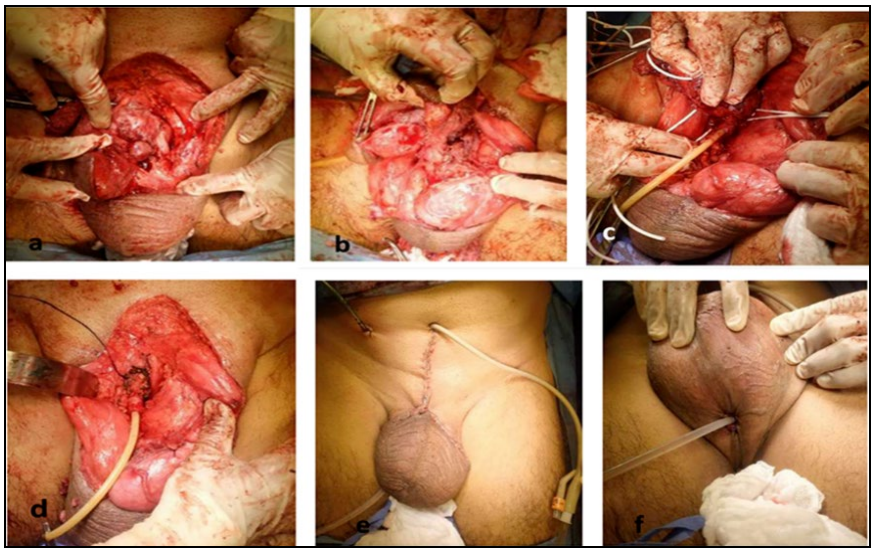

Figure-2: Operative pictures.

A): Infra-umbilical Mercedes Benz incision extending till rectus sheath and including all surrounding subcutaneous tissue. B): Exposed testes with spermatic cords; Penis degloved. C): Corpus spongiosum with urethra divided. Corpora seen to be involved by tumour. D): Crura at their origin at inferior public rami divided and ligated. Catheterized urethral stump for perineal Urethrostomy. E): Wound closed with supra-pubic catheter, and pelvic drain. F): Cutaneous perineal urethrostomy.

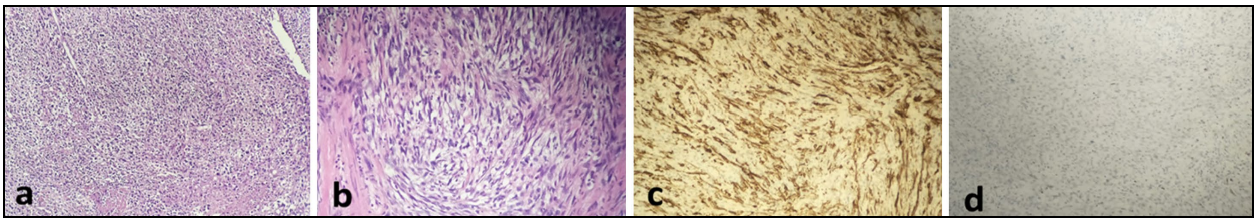

Figure-3: Histopathology

A): Low power magnification (10X) H&E: Neoplastic lesion showing fascicular arrangement and radiating from blood vessels. Necrosis is also seen. B): High power magnification (40X) H&E: The neoplastic cells are oval to the spindle, moderately to markedly pleomorphic, and have clear to eosinophilic cytoplasm.
C): Immunohistochemistry (20X): Immunostain Alpha- smooth muscle actin (ASMA) showing diffuse positive cytoplasmic staining. D): Immunohistochemistry (20X): Immunostain Myogenin shows negative staining.

DISCUSSION

Soft tissue sarcomas are a diverse group of malignant neoplasms which are mesenchymal in origin. They are more prevalent in the male population and account for 1% of all adult solid malignant neoplasms.[1,2] Majority is in the extremities, internal organs and retroperitoneal space and only 5–10% involve the chest/ abdominal wall.[3] The risk factors for soft tissue sarcoma include a history of exposure to radiation, chemicals, and carcinogens such as arsenic and vinyl chloride. Certain inherited conditions such as Li-Fraumeni syndrome (p53 gene mutation), hereditary leiomyomatosis and renal cell cancer syndrome (FH gene mutation) and Von Recklinghausen disease (NF1 gene mutation) are also associated with the development of soft tissue sarcomas. Similarly, the association with chronic lymphedema, burn and smallpox scars is also described.[4]

Subcutaneous Leiomyosarcoma (LMS) are extremely rare soft tissue sarcomas arising from small-to-medium-sized blood vessels in the subcutaneous tissues.[5] Due to rarity and misleading features, these tumours are being diagnosed late resulting in a delay in treatment. There are only a few reported cases of primary subcutaneous LMS derived from the abdominal wall.[1,3,5,6] These tumours can be symptomatic or asymptomatic and generally smaller at the time of diagnosis compared to their intra-abdominal counterparts. They are discovered on palpation either by the patient or as a coincidental finding during physical examination.[3] Cross-sectional imaging (CT, MRI scan) is required to provide an evaluation of the anatomical site of tumour origin, its vascularity, relationship to other organs and evidence of any metastasis.[7]

The treatment for soft tissue sarcoma is mainly driven by the disease stage.[8] A multidisciplinary approach is required with surgical resection being the standard primary treatment for localized abdominal wall LMS.[2] The surgical incision is planned according to the anatomical site of involvement. Wide local excision with an adequate (at least 1 cm) surgical margin of uninvolved tissue is recommended.[8,9] Inadequate excision, tumour rupture or a positive margin can lead to local recurrence or risk of metastasis.[9] Primary closure is the preferred approach; however, larger defects may need reconstruction requiring plastic surgery techniques such as myo-cutaneous flap or reinforcement by prosthetic mesh.[9]

Factors associated with poor prognosis include size (>5 cm), deep-seated tumour with fascial involvement, infiltrative growth pattern, tumour grade, presence of (% areas) necrosis, intra-tumoral vascular invasion, high mitotic rate (>20 per 10 HPF).[10] The role of adjuvant treatment such as chemo or radiation in subcutaneous LMS is not clear. Adjuvant radiotherapy following complete excision is shown to have excellent results, particularly in high-grade and larger (>5 cm) tumours.[10] Early surgical excision with wide local margins is the most important prognostic factor in patients with abdominal wall LMS. Being highly aggressive with a high risk of recurrence and metastases, these tumours, therefore, need close surveillance and follow-up.

Conflicts of interest: The authors declare no conflict of interest.

REFERENCES

1. Jena S, Bhattacharya S, Roy S. Giant subcutaneous leiomyosarcoma of anterior abdominal wall. Case Rep Surg 2014;2014:308916.
2. Gómez J, Tsagozis P. Multidisciplinary treatment of soft tissue sarcomas: An update. World J Clin Oncol 2020;11(4):180–9.
3. Eken H, Karagul S, Topgul K, Yoruker S, Ozen N, Gun S, *et al*. Giant cutaneous leiomyosarcoma originating from the anterior abdominal wall: A case report. Am J Case Rep 2016;17:35–8.
4. Thomas DM, Ballinger ML. Etiologic, environmental and inherited risk factors in sarcomas. J Surg Oncol 2015;111(5):490–5.
5. Matsuda-Mitake Y, Sawada N, Inoue C, Kira S, Mitysui T, Takeda M. A case of subcutaneous leiomyosarcoma of the scrotum presenting as a slowly growing mass in a 36-year-old male: A case report and literature review. Urol Case Rep 2019;23:90–1.
6. Kumar A, Paswan SS, Chumber S, Kumar B. Subcutaneous leiomyosarcoma of anterior abdominal wall in young girl: Rare case report. Saudi Surg J 2018;6:60–2.
7. Levy AD, Manning MA, Al-Refaie WB, Miettinen MM. Soft-tissue sarcomas of the abdomen and pelvis: Radiologic-pathologic features, Part 1-common sarcomas: From the radiologic pathology archives. Radiographics 2017;37(2):462–83.
8. von Mehren M, Randall RL, Benjamin RS, Boles S, Bui MM, Ganjoo KN, *et al*. Soft Tissue Sarcoma, Version 2.2018, NCCN Clinical Practice Guidelines in Oncology. J Natl Compr Canc Netw 2018;16(5):536–63.
9. Williams KJ, Hayes AJ. A guide to oncological management of soft tissue tumours of the abdominal wall. Hernia 2014;18(1):91–7.
10. Svarvar C, Böhling T, Berlin O, Gustafson P, Folleras G, Bjerkehagen B, *et al*. Clinical course of nonvisceral soft tissue leiomyosarcoma in 225 patients from the Scandinavian Sarcoma Group. Cancer 2007;109(2):282–91.

Submitted: June 16, 2022	Revised: December 17, 2022	Accepted: December 30, 2022

Address for Correspondence:

Dr. Syed Muhammad Nazim, Associate Professor, The Aga Khan University, P O Box 3500, Stadium Road - Karachi 74800-Pakistan
Tel: +92-21-34864761
Email: muhammad.nazim@aku.edu