Journal Homepage: -www.journalijar.com
INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)
Article DOI:10.21474/IJAR01/4124
DOI URL: http://dx.doi.org/10.21474/IJAR01/4124
ISSN NO. 2320-5407

RESEARCH ARTICLE

LEIOMYOSARCOMA OF PENIS: A RARE CASE WITH UNUSUAL PRESENTATION.

Shankar A[1], Nagpal P[1], Mitra S[2], Kakkar N[2], Kumar N[1] andGhoshal S[1].

1. Department of Radiotherapy, PGIMER Chandigarh.
2. Department of Histopathology, PGIMER Chandigarh.

Manuscript Info

Manuscript History

Received: 01 March 2017
Final Accepted: 01 April 2017
Published: May 2017

Keywords:
Leiomyosarcoma, penis, rare

Abstract

Primary leiomyosarcoma of the penis is an extremely rare tumor. It presents in fifth to seventh decade of life. No effective treatment for metastatic disease has been found till date. We report a rare case of deep seated penile leiomyosarcoma with unusual natural disease course in a 51 year old male along with brief review of literature.

Introduction:-

Tumors of the penis comprises 0.4–0.6% of all malignant neoplasm[1]. Among them, sarcomas represent less than 5% of the total[2]. Primary leiomyosarcoma of the penis is an extremely rare tumor. There are very few cases reported in the medical literature till date. We report a rare case of leiomyosarcoma penis with unusual natural disease course.

Case Presentation:-

51 year old patient presented in 2010 with complaints of painless swelling over the shaft of penis for 3 years. On examination, there was a swelling in the mid penile shaft about 6*4 cm in size fixed to the corpora cavernosaalong with bilateral inguinal lymphadenopathy 1*1 cm.Besides the swelling, no other changes can be seen. Laboratory results were normal. Radiography of the chest showed normal condition of the lungs, without evidence of metastatic changes.PET-CT revealed heterogeneous enhancing soft tissue lesion involving corpora cavernosa on the left side of the penis (SUVmax-5.8) with mild FDG uptake in right inguinal, bilateral external iliac and left common iliac lymph nodes. Biopsy showed tumor arranged in short and long interlacing fascicle (Figure 1). The tumor cells were spindle in shape showing moderate pleomorphism with hyperchromatic nuclei and inconspicuous nucleoli with frequent mitosis(Figure 2,3). The tumor cells were positive for smooth muscle actin (Figure 4) suggestive of leiomyosarcoma. Then the patient was lost to follow up for 4 years and presented to the OPD in March 2014 with chest X ray showing multiple lung metastases.Patient was planned for systemic chemotherapy with vincristine, adriamycin and cyclophosphamide alternating with ifosfamide and etoposide for six cycles which completed in November 2014.PET-CT done two months later for response assessment was suggestive of complete resolution of the primary and FDG avid multiple bilateral lung nodules and mediastinal lymph nodes-metastsatic disease as it was before treatment. Patient being asymptomatic was kept on follow up. Another PET-CT done almost one year later showed FDG avid nodular lesion in penile shaft along with multiple lung, mediastinal and skeletal lesions-progressive disease. Subsequently patient was planned for second line gemcitabine based systemic chemotherapy but was again lost to follow up for 6 months after 3 cycles. Patient's general condition was very well preserved considering the amount of disease burden and was planned for another PET based evaluation of his disease status which would guide his further treatment. PET CT done on 23-09-16 showed FDG avid lesion in penis along with

Corresponding Author:-Shankar A.
Address:- Department of Radiotherapy, PGIMER, Chandigarh.

FDG avid lesion in retroperitoneum, pancreas, liver, bilateral lungs suggestive of disease progression. Gemcitabine based chemotherapy was continued for six cycles which completed on 6-12-16. Repeat PET CT done on 30-1-17 revealed FDG avid lesion in penis along with retroperitoneal, pancreas, bilateral lungs,and skeletal muscle metastasis suggestive of progressive metabolic disease. Patient last visited OPD on 7-2-17 where on examination he had multiple nodules in penis from root to tip and nodule in periumbilical region. Patient has been kept on best supportive care.

Discussion:-

The incidence of penile cancer is very low and varies from 0.5-5 /100 000 men. However, sarcomas are extremely rare and constitute less than 5% of all malignant tumors of the penis[3]. To date, less than 50 cases have been reported in the medical literature[4].Leiomyosarcomas of the penis occur most often in the fifth to seventh decades of life as has been in our case who presented at the age of 51.Symptoms are generally indolent.

Penile leiomyosarcomas can be classified into two types, superficial and deep-seated tumours[5].

Superficial tumors are usually present over the shaft distally or the prepuce. It most commonly occurs in middle-aged men. It is a slow growing tumor.

Deep tumors arise from the glans penis and from the corpora cavernosa or corpus spongiosum[6] as has been in our case. In comparison to superficial tumors deep tumors show greater propensity to develop metastasis[7]. The most common site of spread is the lungs as has been seen in our case who presented with diffuse lung metastasis.

Treatment modality has not yet been established because of few cases reported in the medical literature. Treatment often ranges from conservative surgery to total penectomy, and adjuvant radiotherapy or chemotherapy. Radical penectomy is the treatment choice for deep tumors. Lymph node involvement is very rare, and so elective lymphadenectomy is not recommended[8]. If lymph node is involved then there is a high chance of developing distant metastasis, primarily to the lung as has been in this case who presented with inguinal lymphadenopathy and lung metastasis subsequently.

External beam radiation has not proven to be of value in treating penile leiomyosarcomas[9]. Chemotherapy has provided poor results and generally comprises of anthracyclines or etoposide based regimens. Since our patient presented with metastatic disease at onset he was given palliative chemotherapy with anthracycline based regimen but as given in the literature showed disease progression on chemotherapy.

One rare presentation in our case which in itself is a rare entity is that the patient is doing well even after 6 years of diagnosis and since 2 years after being diagnosed of having such high systemic disease burden and showing disease progression after first line chemotherapy. Patient is well preserved till date. This presentation is in sharp contrast with the available data in the literature. A review with follow-up duration of 12 years showed that local recurrence occurred in 26% of the patients between three to 18 months from the first surgery, and among those patients that had deep lesions, eight died between the first month and 36 months from surgery.

Conclusion:-

Sarcomas of penis are rare tumors and deep-seated lesions are usually associated with poor prognosis. The radical tumor resection with negative surgical margins offers the best chance of cure. No effective treatment for metastatic disease has been developed till date.

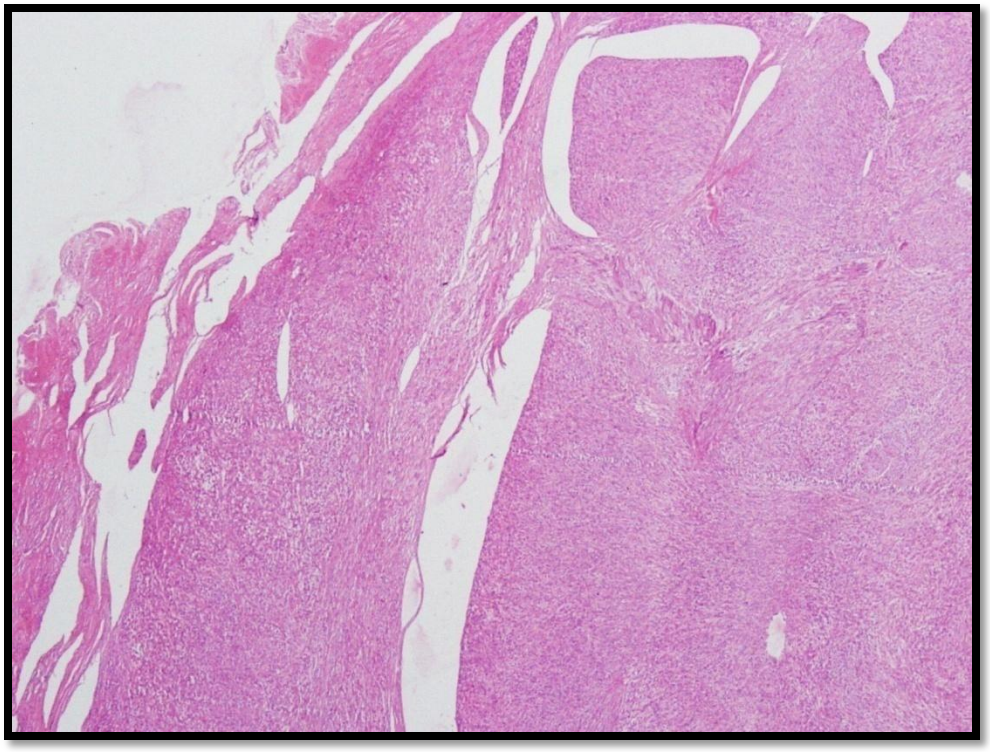

Figure 1:- Section showing a tumor with infiltrative margin.

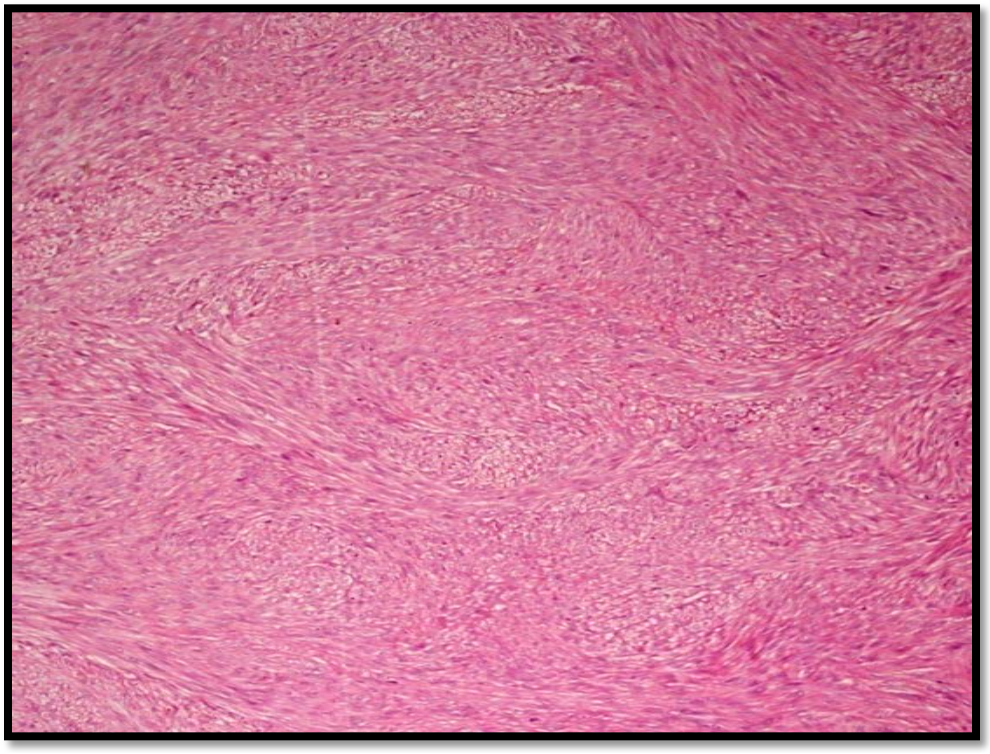

Figure 2:- The tumor showing fascicular pattern and moderate degree of pleomorphism

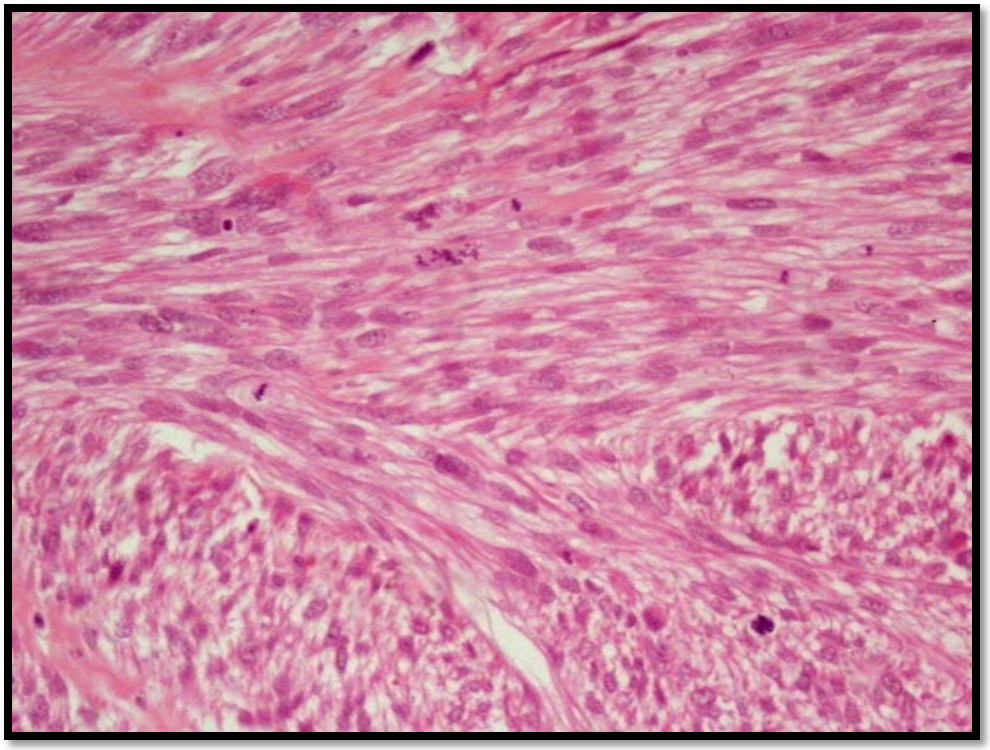

Figure 3:-Higher magnification showing multiple mitotic and apoptotic figures along with atypical mitoses

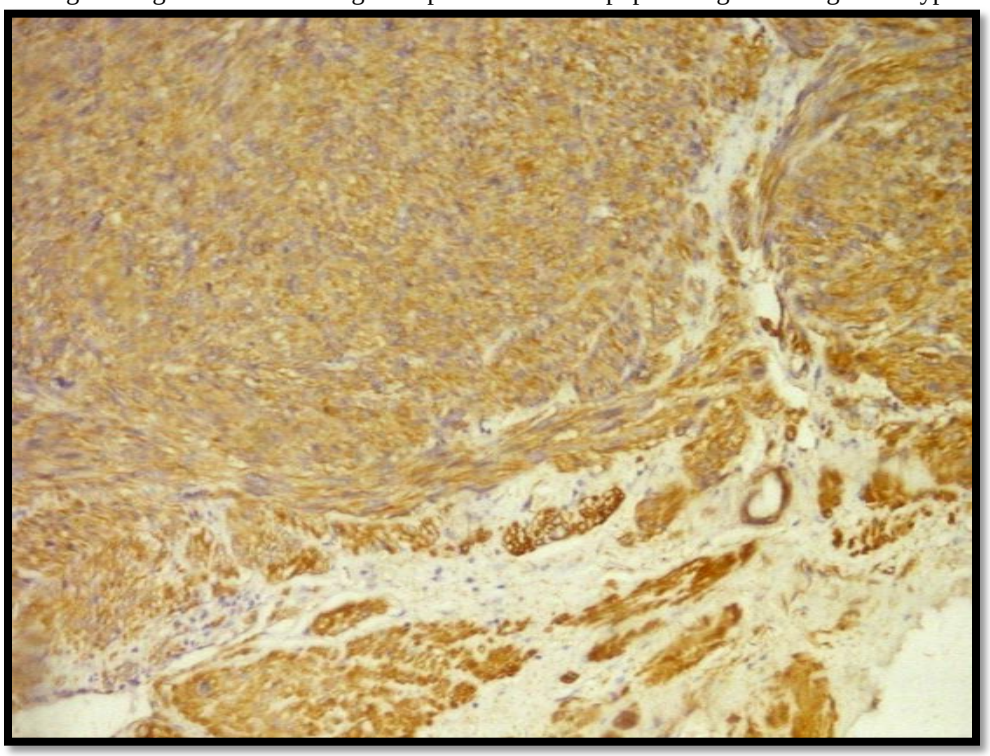

Figure 4:- Immunohistochemistry for SMA (Smooth muscle actin) showing diffuse strong cytoplasmic positivity

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