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Case Report

A case report on a sporadic retroperitoneal malignant peripheral nerve sheath tumour mimicking a pelvic abscess

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ABSTRACT

Soft tissue sarcomas (STS) are a diverse group of neoplasms with about 60 subtypes arising from a variety of anatomical structures across all age groups. The diagnosis of STS can prove to be challenging as it mimics a variety of non malignant conditions. Here we present one such case of a sporadic MPNST which was operated on with the main differential diagnosis of a Pelvic Abscess. Hence though STS is rare in general population it should always be kept in mind in cases of doubtful and confusing presentations.

Keywords: Malignant peripheral nerve sheath tumour, Malignant schwannoma, Pelvic abscess retroperitoneal tumour, Soft tissue sarcomas

INTRODUCTION

Soft tissue sarcomas have an incidence of about 1% among the population while accounting for 2% of all cancer deaths.[1] Trunk and Extremity sarcomas are more common than the intra-peritoneal and retroperitoneal types. Most are of the sporadic type but there are well known association with germline mutations, radiation exposure and environmental factors.[1]

MPNSTs constitute 2% of all sarcomas. 50% of which are a part of Von Recklinghausen's disease, 40% are sporadic and 10% are associated with Radiation exposure. Here we'd like to present a case of STS that was diagnosed on table in a 43yr old female who was operated on with a provisional diagnosis of Pelvic abscess – which turned out to be a Retroperitoneal Soft tissue Sarcoma and later on was proven to be a MPNST.

CASE REPORT

A 43-year-old pre-menopausal female presented to the outpatient clinics of our department with the chief complaints of constipation, lower abdominal pain – of a dull aching type, constant, and aggravated by defaecation, bleeding per rectum - on and off on straining at stools for a duration of 6 months. She also gave a History of Weight and Appetite loss. She is a known seizure disorder patient on regular medication for the past 7 years. She has no other co-morbid illnesses.

History of previous surgeries was as follows:

- Peurperal Sterilisation done 22 years back
- Open Haemorrhoidectomy done 3 years back

She has regular menstrual cycles with the last menstrual period being 2 weeks back. She has 3 live children all of which were full term normal vaginal institutional

deliveries, with the last childbirth being 22 years back after which she underwent Peurperal Sterilisation.

There are no records of malignancy in her family members.

On examination she was thin built and pale. Systemic examination reveals no obvious abnor-malities.

On examination of external genitalia, inspection revealed a fullness of the left labia majora, left side of perineum and left gluteal region. On palpation, there was no warmth or tenderness and no variability in consistency from surrounding tissues.

On per vaginal exam cervix was healthy, uterus was retroverted and fornices were free. On per rectal exam – rectal mucosa was normal and no masses were palpable through the walls.

On further investigation, blood work up showed a normal WBC count.

USG local region showed a 9×7 cm collection in the left perianal region – subsequently needle aspiration was attempted and showed no yield.

CT Brain showed no significant abnormality, MRI Pelvis, showed a predominantly hypointense collection with internal T2 hypointense shaggy strands, a few cystic regions and incomplete surrounding wall 17×8×9.3 cm in the perineum and pelvis on the left side extending from the labia majora upto the recto sigmoid junction exerting mass effect on the uterus and rectum pushing them to the right - F/S/0 – large perineal, inter sphincteric and pelvic abscess – left side.

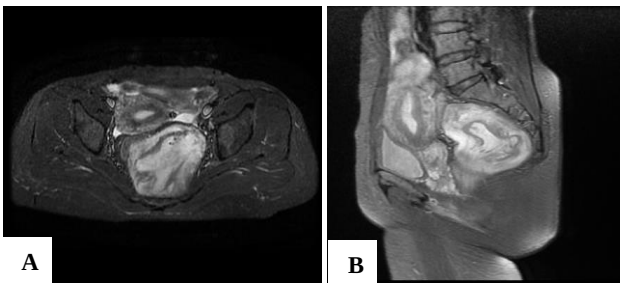

Figure 1: MRI image of the lesion (A) sagittal section of the pelvis showing the lesion behind the uterus; (B) coronal section depicting the same.

Colonoscopy revealed normal colonic mucosa and vascularity upto a distance of 90 cm from anal verge.

Gynaecological clearance was obtained.

Patient was posted for an Elective Explorative laparotomy.

Intra-operative findings were as follows:

- A ~20×8×10 cm retroperitoneal soft tissue tumour found to be occupying the recto-uterine pouch on the left side pushing the uterus and recto sigmoid junction to the right.
- Tumour is found to be separate from both the genitourinary and alimentary tracts
- Peritoneum opened and the upper two thirds of the tumour freed from surrounding structures as was feasible.
- Lower third of the tumour approached via a left perineal incision with the patient in lithotomy position – dissected from the external sphincter and pelvic floor muscles using electrocautery
- Tumour was then delivered out through the perineal incision after ligating the feeder vessels and freeing the final attachments from the anterolateral and medial pelvic walls.

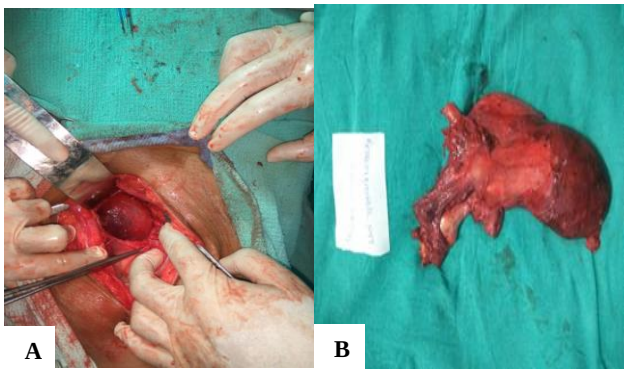

Figure 2: Per-operative pictures of the tumour (A) intra-operative image of the tumour; (B) excised specimen with frayed lower edges.

Histopathological examination shows

Microscopic

Section studied shows moderately cellular neoplasm containing short fascicles and singly dispersed cells in abundant stroma. Tumour cells are spindle shaped, buckled nuclei, have hyperchromatic dispersed chromatin and less mitotic activity. Increased vascularity. Some vessels show invasion by tumour cells.

Impression - low grade malignant periipheral nerve sheath tumour.

Immuno histochemistry

- CD 99 - Focal membrane positivity
- Ki 67- occasional cells show positive- low proliferation index
- Vimentin, desmin - negative

Post-operative period was uneventful and patient was referred to oncology department for adjuvant radiotherapy.

Repeat MRI pelvis post radiation shows no residual lesion. Patient is now on regular follow up with us.

DISCUSSION

Soft tissue sarcomas encompass a wide variety of tumours that may arise from any of the following structures – skeletal muscles, adipose tissue, blood vessels, lymphatics peripheral nerves and connective tissues with a common mesodermal origin.[1] They affect individuals across all age groups. Most are sporadic but known associations exist with:

Germline mutations- Li fraumeni syndrome, neurofibromatosis 1, familial adenoma-tous polyposis etc. Radiation exposure and environmental exposure to carcinogens like thorotrast and polyvinyl chloride (with hepatic angiosarcoma).[1]

Various non-neoplastic condition may mimic STS like:

Hypertrophic scar, retroperitoneal lymphadenopathy: lymphoma, germ cell tumour, or metastasis from gastrointestinal primary, hematoma, myositis ossificans,benign lipoma, cyst, abscess, cutaneous malignant neoplasms - including melanoma.[1]

This was the case in our patient. Hence a diagnosis of STS should be kept in mind in cases of suspicious presentations.

Malignant peripheral nerve sheath tumours are malignant counterparts of benign schwanno-mas arising from peripheral nerves or the nerve sheaths. Previously called malignant schwan-noma, Neurofibrosarcoma and neurogenic sarcoma. Usually painless at presentation. Commonly affecting those in the 20 to 50 years age bracket. About 40% of tumours are sporadic, 50% are a part of neurofibromatosis 1 and 10% are due to radiation exposure. These tumours are highly aggressive with poor prognosis and high rates of relapse.

Preoperative imaging includes MRI and CT.

A heterogenous lesion due to necrosis and haemorrhage and patchy contrast enhancement in MRI is an indication of malignant MPNST.[2,3] MRI is the investigation of choice because it can reveal the nerve of origin and its relationship to adjacent structures.[4]

A CT chest is mandatory to rule out lung metastasis in all cases.

As for biopsy - FNAC is unsatisfactory, image guided core needle biopsy or incisional biopsy is preferable. Care should to be taken to ensure that the needle trajectory is included in the future resection margins however. Treatment options are Margin free resection with adjuvant Radiotherapy. Radical surgical re-section with a good three-dimensional clearance is mandatory for a successful outcome. Am-putations are indicated when wide excision is not feasible and in those patients with severely compromised limb function. Routine nodal dissection is not indicated. When a major nerve is identified, the cut end should be sent for frozen section to assess the tumour free margin of the resection.

In most instances, the tumours display fascicles of spindle cells woven into herringbone pat-tern with varying degrees of mitosis and necrosis. However, it is not always possible to demonstrate the origin from a nerve, especially when it arises from a small peripheral branch. This point was proven by Nambisan et al and Bilge et al.[5,6] Still, there are several other dis-tinct features, such as proliferation of tumour in the sub-endothelial zones of vessels with nepotistic cells herniation into vessel lumen and proliferation of small vessels in the walls of the large vessels, which are very characteristic features of MPNST, as demonstrated by the tumour showing vascular invasion in the histopathological report of our case.[7]

Postoperative radiotherapy is recommended by oncology consensus group as part of a uni-form treatment policy for MPNSTs, much like other high-grade soft tissue sarcomas even in the presence of tumour free margins.[8-11] Basso-Ricci demonstrated 56% disease free surviv-al using combined surgery and radiation therapy for MPNST.[11]

Postoperative surveillance by physical examination and imaging is mandatory.

CONCLUSION

Soft tissue sarcomas are best treated by the Multimodality approach. Same is applicable for the MPNSTs. In all cases margin free resection is the goal followed by Radiotherapy. Proper follow-up for relapses are mandatory. A variety of non-neoplastic conditions may mimic STS and proper vigilance is therefore required in such suspicious presentations.

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