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General Surgery

MALIGNANT PERIPHERAL NERVE SHEATH TUMOUR (MPNST) OF SCROTUM - A RARE CASE.

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ABSTRACT Malignant tumours arising from, or displaying differentiation along the lines of the various elements of the nerve sheath are collectively referred to as Malignant Peripheral Nerve Sheath Tumours(MPNST). We present a case of a 42 year gentleman with MPNST of the scrotum, which is in a rare location, making it unique.

KEYWORDS : Malignant peripheral nerve sheath tumour, malignant schwannoma, neurofibroma.

INTRODUCTION:

MPNST's also known as Malignant Schwannoma, constitute 5-10% of all soft tissue sarcomas[1]. These are malignant tumours arising from peripheral nerves or in extraneural soft tissues,or a pre-existing neurofibroma . Most common locations are buttock, thigh, brachial plexus, arm and paraspinal region. Sporadic tumours are seen in 2nd to 5th decades of life. Neurofibramatosis(NF1) related tumours are seen in 3rd to 6th decades of life [2]. MPNST of the scrotum has not bMalignant peripheral nerve sheath tumour, malignant schwannoma, neurofibroma.een reported in the literature so far.

CASE REPORT:

A 42 year gentleman was admitted to our hospital with complaints of gradually enlarging scrotal swelling of 2 years duration. There was no history of trauma and the swelling was painless. He had no significant medical or family history. General examination and systemic examination were unremarkable. Local examination revealed a globular 18x18 cm, firm, freely mobile swelling in the scrotum placed inferior to the testes with intervening lax scrotal skin. A healing ulcer of size 5x8mm seen on the base of the scrotum on the median raphe. Both the testes were normal. There was no significant inguinal lymphadenopathy. Routine laboratory investigations were within normal limits. USG showed a large mixed echogenic mass lesion inferior to both the testes in the scrotal sac with internal cystic components showing, lobular outline and internal vascularity. Metastatic work up was negative.

The patient was scheduled for wide local excision of the tumour. Intraoperative findings revealed, a fleshy, globular lesion with increased vascularity. Multiple vessels were traversing the loose areolar connective tissue between the lower poles of testes and the tumour. The mass lesion was excised, dissecting it free from the surrounding tissues including the excess scrotal skin ,maintaining wide margins. Primary closure of the dartos muscle and the skin was achieved. Both the testes were free from the lesion. Histopathological examination revealed ,oval to spindle shaped cells arranged in fascicles and storiform pattern. Cells have eosinophilic cytoplasm, large vesicular nucleus, lymphocytic collections around tumour bundles. Few places show cells with bizzare and hyperchromatic nucleus. Tumour giant cells were seen. Mitotic activity was increased 10/10 HPF. Few sections were exhibiting spindle shaped cells with wavy sinuous nuclei with hyperchromatism,and the tumour showed necrotic material. Immunohistochemistry revealed the tumour cells staining positive for S-100.

Patient had an uneventful recovery postoperatively. He was further advised to follow up with adjuvant radiotherapy of 60 Gy in 30 fractions, over 6 weeks, which he declined. However , one year post surgery, there was no recurrence of the tumour and the patient was healthy.

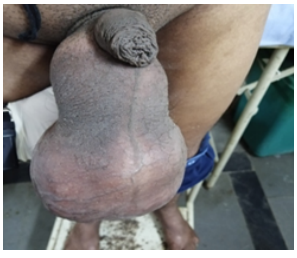

Fig.1 Preoperative photograph.

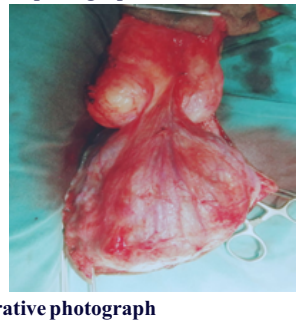

Fig.2 Intraoperative photograph

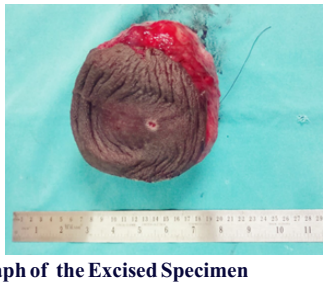

Fig.3 Photograph of the Excised Specimen

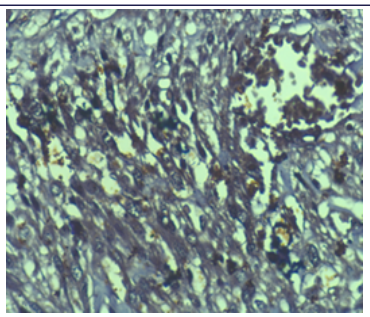

Fig.4 Photograph showing focally positive S-100 .

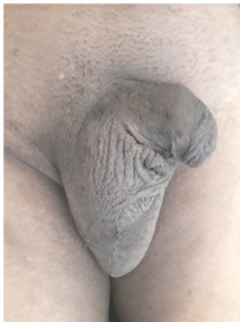

Fig.5. Follow up photograph after1 year of surger

DISCUSSION:

MPNST's arise from Schwann cells, perineural cells, or fibroblasts. They comprise approximately 5-10% of all soft tissue sarcomas[1]. The aetiology is unknown. 50% arise denovo and 50% of these tumours are associated with NF1[3]. There is an increased incidence in patients with radiation exposure [4]. It has also been related to genetic insults involving p53 and p16. They generally present as an enlarging palpable mass. Magnetic resonance imaging (MRI) is the imaging modality of choice [5]. They are often high-grade, demonstrating 4 or more mitotic figures per high powered field. S-100 is positive in 50 – 90% of all MPNST's. Wide local excision is the first line of therapy. Together with wide surgical excision, radiation therapy offers local and overall survival rates with fewer wound healing complications. In MPNST, it can be employed pre-operatively, intraoperatively and post-operatively[6]. Chemotherapy is intended for systemic disease, where the local treatment techniques are ineffective. It is also administered in high-grade disease, deeply placed tumours, large lesions more than 8 cm in maximum dimension, tumours in which metastatic disease is likely and in patients less than 65 years. Owing to a high risk of recurrence with incomplete resection, postoperative irradiation and chemotherapy are necessary. However, they are often used as adjuvant therapy, even if the tumor is completely resected . Even with aggressive therapy ,the local recurrence rate has been reported to range from 40-65% and the distant recurrence rate from 40-68%[7] .

In our case, the presentation was a large tumour of the scrotum inferior to testes, which was investigated and later excised with macroscopically tumour free wide margins. The HPE revealed , MPNST with high mitotic activity and IHC showed tumour cells positive for S100. Patient was advised adjuvant radiotherapy postoperatively. This case is presented, as MPNST is rare in the scrotum and has not been reported in literature.

CONCLUSION:

Malignant peripheral nerve sheath tumours constitute a small proportion of soft tissue sarcomas. In this case, the location of MPNST in the scrotum makes it a rare entity. Advanced imaging modalities, wide local excision of the tumour, immunohistochemistry , adjuvant radiotherapy and chemotherapy allows for accurate disease identification and appropriate management.

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