Malignant peripheral nerve sheath tumour (MPNST) – A rare case report with Skip Metastasis

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Abstract:

Soft tissue sarcomas (STS) as a whole account for approximately 1% of malignant tumors. They can occur anywhere in the body, but mostly originate in the extremities (60%), with the lower extremities involved three times as often as the upper extremities. Skip metastasis is a rare phenomenon in soft tissue sarcomas. Malignant peripheral nerve sheath tumors (MPNSTs) is a rare soft tissue sarcoma comprising 5% of all soft tissue sarcomas, they usually arise from peripheral nerves or somatic soft tissues. MPNSTs can develop in any anatomical region, but the sciatic nerve is affected most often. Around 50–70% of MPNST cases arise in the setting of NF1, the most common familial cancer-predisposing syndrome in humans–either from transformation of a plexiform neurofibroma or following radiation therapy. Metastasis occurs in 39% of patients and most commonly to lungs. The definitive diagnosis of MPNST is obtained by biopsy. Imaging modality of choice is MRI. Surgery is the most beneficial treatment but wide resection with negative margins are not just enough to declare complete removal of tumour because of Skip metastasis a rare phenomenon. so we suggest to keep in consideration about Skip nodule while doing soft tissue sarcoma surgery and should keep in mind the idea of whole compartmental resection (myomectomy) as maximum as possible in order to get rid of the parent nodule as well as the skip nodule along with it. Postoperative radio- and chemotherapy are part of adjuvant therapy. We are reporting a case of 36 yr old male with MPNST in the posterior aspect of left mid thigh with Skip metastasis. For the complete removal of tumour compartmental resection, not just free negative margins so as to get rid of the skip nodule a rare phenomenon in soft tissue sarcomas, forms the basis of this report.

Introduction:

Sarcomas are a heterogeneous group of rare tumors that arise predominantly from the embryonic mesoderm. The various sarcomas include bone sarcomas (osteosarcomas and chondrosarcomas), Ewing’s sarcomas, peripheral primitive neuroectodermal tumors, and soft tissue sarcomas. Soft tissue sarcomas can occur anywhere in the body, but most originate in an extremity (59%), the trunk (19%), the retroperitoneum (15%), or the head.
and neck (9%). Currently, more than 50 histologic types of soft tissue sarcoma have been identified but the most common are malignant fibrous histiocytoma (28%) (Now known as pleuromorphic undifferentiated sarcoma PUS), leiomyosarcoma (12%), liposarcoma (15%), synovial sarcoma (10%), and malignant peripheral nerve sheath tumors (6%). MPNST is least common among soft tissue sarcomas, these originate from peripheral nerves or from cells associated with the nerve sheath such as Schwann cells, perineural cells, or fibroblasts. Because they can arise from multiple cell types the overall appearance can vary greatly from one case to the next and this makes diagnosis and classification somewhat difficult, they can occur either spontaneously or in association with NF1 (50%). Though etiology is unknown but there is higher incidence in patients with radiation exposure, chemical exposure and genetic predisposition. MPNST generally occur in adulthood mainly between 20 – 50 years age group. MPNST usually present as an enlarging palpable mass with variable complain of pain, paraesthesias, and motor weakness. They are usually deep seated and often involve the proximal upper and lower extremities. MRI is the imaging modality of choice. Imaging study of chest is an important part of any sarcoma evaluation. MPNST are most likely to metastasize to the lungs followed by bone and finally the pleura, because of this CT chest is preferred. The definitive diagnosis is obtained by biopsy. On histology 80 – 85 % of MPNST are spindle cell tumours with fasciculating patterns. S-100 has been identified in 50 to 90 % of MPNSTs.

The mainstay of treatment is surgical resection. The goal of operation is to achieve complete surgical excision of the tumour with negative margins. Radiotherapy and chemotherapy can be used as an adjuvant postoperatively.

Skip metastasis is a rare phenomenon in soft tissue sarcomas and in MPNST its identity has not been reported that much so far. Because of this rare entity complete surgical removal of the tumour burden becomes nightmare so in order to get rid of this skip nodule it is better to go for complete muscle or compartmental resection. Reporting of such a rare phenomenon and the advocacy of complete muscle or compartmental resection as whole to get rid of the actual tumour burden forms the basis of this case report.

Case report:

36 yr old male presented with the complain of swelling over the left thigh posteriorly 6 months back. Swelling was sudden in onset, not associated with pain. The only complain was that, he felt numbness in the left leg on prolonged sitting, no history of any bone pain, no h/o chest symptoms, no h/o radiation exposure, no h/o any fracture treatment in the past. No family history. Regarding past history there is a h/o decreased vision in left eye 1 ½ yr back for which he was conservatively managed. Thereafter the vision improved. On examination hard nodular diffuse swelling of size approximately 7x3 cm. over posteromedial aspect of middle of left thigh, overlying skin was normal and intact, no signs of
inflammation, no other significant finding on local and systemic examination, for which he was investigated with routine investigations and MRI was done. Then the excisional biopsy of the swelling was done and histopathological report came out be Malignant peripheral nerve sheath tumour diffusely positive for s-100 and negative for SMA and myogenin.

Thereafter again swelling starts to appear and started to increase in size and leading to discomfort in sitting, symptoms of numbness on sitting increased in intensity. Then the patient was admitted and evaluated again clinically and radiologically by MRI and on MRI it was found out that, there is a residual lesion in relation to the left sided semitendinosus muscle at the level of mid thigh with adjoining inflammatory changes and areas of haemorrhage. Core needle biopsy was done and diagnosed as Malignant mesenchymal tumour, compatible with malignant peripheral nerve sheath tumour. CT chest was clear.

Patient was planned and prepared for the surgical excision of the swelling, in prone position under SA by an elliptical incision over posterior aspect of left thigh measuring about 14 x5 cm. given, swelling part included within the ellipse under hemostatic control layer by layer dissection done, muscle having the tumour mass exposed, sciatic nerve explored and secured, other vital structures coming in the field were explored and secured, separation of biceps femoris and semitendinosus from their bed was done, musculotendinous attachments of these two muscles were identified and after taking control of these two muscles they were resected from their musculotendinous attachments, after that the compartmental bed was explored for any tumour mass, it was found to be clear, then after achieving hemostasis the wound was closed in layers and antiseptic dressing done.

On histopathological examination of the excised tissue reveals a partially well circumscribed tumour measuring 4.5x3.5x3cm. serial slicing reveals another nodule (Skip Nodule) measuring 1x1 cm. which was situated 2 cm. proximal to main tumour, examination shows highly cellular spindle cell tumour. Tumour cells were arranged in sheets and interlacing bundles with moderate atypia and presence of scattered tumour giant cells, increased mitotic activity was there with focal necrosis and myxoid changes and nodular pattern. Tumour was staged as Enneking IIa and AJCC III. Margins of both main tumour and small nodule (Skip Nodule) were free. Diagnosed as Malignant peripheral nerve sheath tumour positive for s-100

Postoperatively hospital stay was uneventful, patient started the normal movements of left lower limb from 3rd postoperative day. Flexion, Extension, Abduction, Adduction, External rotation and Internal rotation at the left hip and Flexion, Extension at the left knee were in their good functional range. Patient was discharged with no complain other than mild pain at the wound site.

Discussion:
High grade sarcomas may break through the pseudocapsule to form “skip” metastasis within the same anatomical compartment. They are occasionally found with low grade sarcomas. Skip nodules are tumour foci that are not in continuity with the main tumour mass, and form outside the pseudocapsule. Multiple satellite nodules normally associated with a high grade malignant fibrous histiocytomas. This finding is preoperatively documented in less than 5% of patients.