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Case Report

Malignant peripheral nerve sheath tumour over the posterior triangle of neck: a rare case report

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ABSTRACT

Malignant peripheral nerve sheath tumour (MPNST) is an extremely rare soft tissue sarcoma which usually arises from peripheral nerves or somatic soft tissue with an incidence of 0.001%. It's most common anatomical sites are the proximal portions of the upper and lower extremities and the trunk and it's extremely rare for such a tumour to occur elsewhere in the body. We report a rare case of such a tumour over the left posterior aspect of neck in a 28-year-old female patient. We have reviewed this case in terms of clinical presentation, investigations, surgical treatment and adjuvant therapy and have shortly described our experience. MRI and CT neck supported the diagnosis of this tumour. Fine needle aspiration cytology taken from the swelling revealed a low-grade spindle cell tumour with a possibility of MPNST. Excision of the tumour was done and the excised specimen was sent for histopathological examination which revealed MPNST. Adjuvant radiotherapy was given postoperatively. At a 6-month follow-up, patient is doing well with no evidence of recurrence. Suspicion of this tumour should be raised in a rapidly growing painless tumour in and around a nerve tissue. Diagnosis is made by assessing a combination of clinical, pathological and immunohistochemistry features. Complete surgical removal should be the goal of treatment with definitive histological diagnosis. A regular follow up is recommended to confirm any recurrence or metastasis.

Keywords: Malignant peripheral nerve sheath tumour, Rare sarcoma, Soft tissue sarcoma

INTRODUCTION

Malignant peripheral nerve sheath tumour is an extremely rare soft tissue sarcoma in the neck with an incidence of 0.001%. It's most common anatomical sites are the proximal portions of the upper and lower extremities and the trunk. It is extremely rare for such a tumour to occur elsewhere in the body.[2] It is of diagnostic importance since it is one of the most aggressive and rare malignant soft tissue sarcomas. Also, these tumours pose difficulties in their diagnosis due to nonspecific, indistinct clinical diagnostic criteria and histopathological resemblance with other spindle cell sarcomas like monophasic synovial sarcoma, leiomyosarcoma and fibrosarcoma.[3]

We report a rare case of MPNST in the posterior triangle of left side of neck in a 28-year-old female patient.

CASE REPORT

A 28-year-old female patient presented with a swelling over the left side of neck for 7 months (Figure1).

History of pain on and off was present for 7 months. History of weight loss was present. There was no history of weakness/paraesthesia in the left upper extremity. No other positive history was present. There was no family history of tuberculosis. On general physical examination, there were no positive findings.

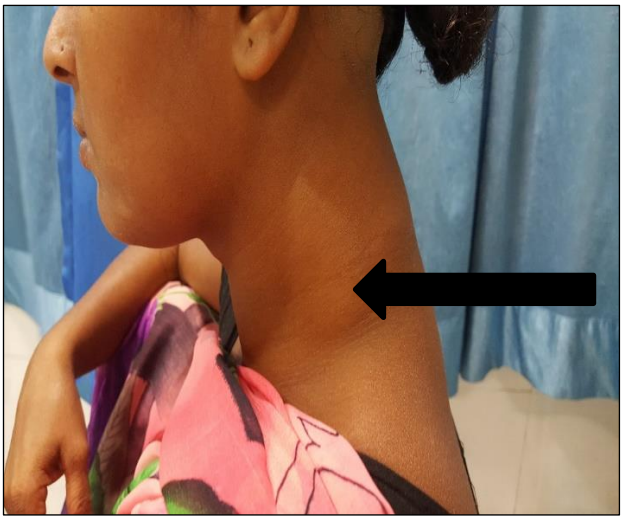

Figure 1: Depicting the swelling over the left posterolateral aspect of neck.

No other lymphadenopathy was noted. On local examination, an 8 x 6 cm swelling was present in the lower half of the left posterior triangle of the neck. There were well defined margins. Skin over the swelling was normal. No dilated veins were seen. There was no local rise of temperature. Swelling was non-tender, firm in consistency and was immobile.

No neurological deficits in the left upper extremity were noted on systemic examination. For clinically differential diagnosis we thought in terms of lymphoma, tubercular lymphadenitis, nonspecific cervical lymphadenopathy and secondaries in neck.

Routine haematological investigations were normal. Chest X-ray was normal. Tuberculin test was negative. Ultrasonography (USG) neck revealed a well-defined, heterogeneous, solid lesion of size 9 x 4 x 5 cm, suggestive of a solid tumour.

Fine Needle Aspiration Cytology (FNAC) revealed a low-grade spindle cell tumour with a possibility of malignant peripheral nerve sheath tumour.

Computed tomography (CT) scan and Magnetic Resonance Imaging (MRI) scan both revealed a solitary well circumscribed, hypodense lesion in the left posterolateral compartment of neck extending superiorly up to C-3 vertebra, anteriorly displacing semispinalis and posteriorly displacing Trapezius, consistent with neoplastic aetiology (Figures 2a, 2b, 2c, 3a and 3b).

Excision of the tumour was done and the excised specimen was sent for histopathological examination. A 10 x 6 cm tumour was excised in toto. Tumour was not adherent to any major nerve.

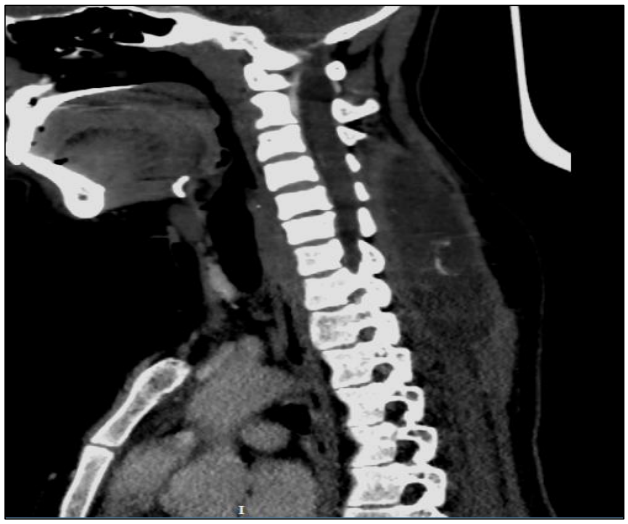

Figure 2a: CT neck sagittal view shows an evidence of a hypo dense lesion in left posterolateral compartment of neck extending from C-3 to C-9 vertebral level with small specks of calcification, likely to be a neurogenic neoplastic tumour.

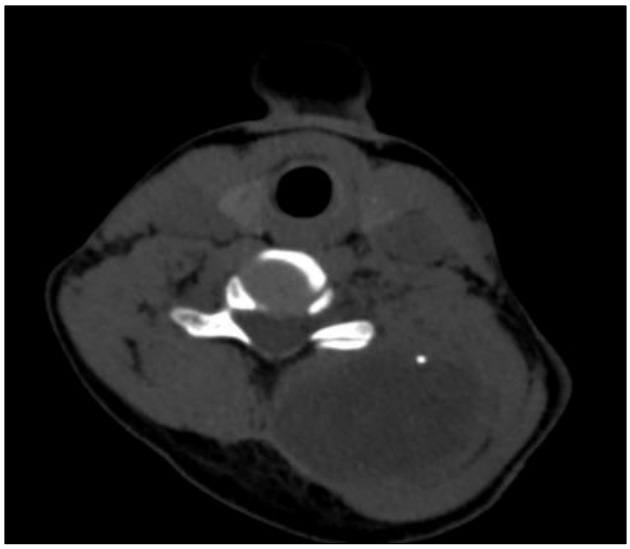

Figure 2b: Plain CT neck shows a solitary well circumscribed, hypo dense lesion in the left posterolateral compartment of neck extending superiorly up to C-3 vertebra, anteriorly displacing Semispinalis and posteriorly displacing Trapezius, consistent with neoplastic aetiology. Lesion shows small specks of calcification.

Histopathology of the excised tumour revealed tumour cells arranged in fascicles having a palisading and whorling appearance. Nuclei were plump and hyperchromatic and abundant mitotic figures were seen. Many nuclei showed serpentine appearance. Spindle cells showed immunoreactivity to S-100.

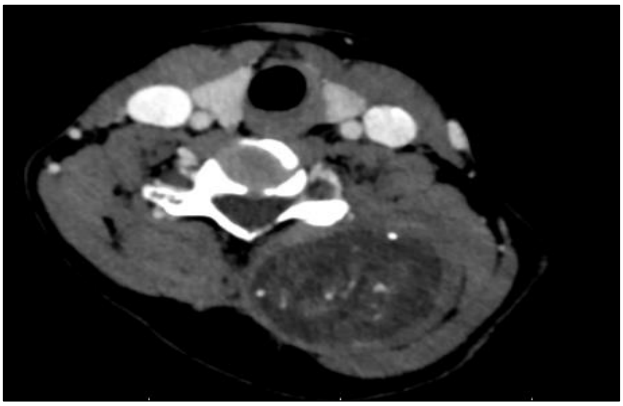

Figure 2c: Post contrast CT neck shows an evidence of peripheral type of enhancement with some central inhomogeneous enhancement with small specks of calcification. No evidence of underlying bone destruction or obvious intraspinal extension noted.

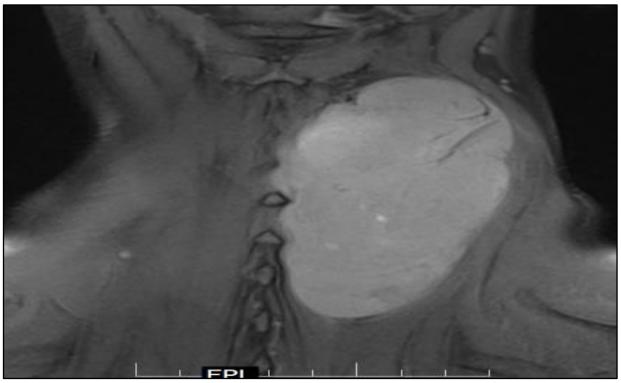

Figure 3a: Stir coronal MRI neck shows a large hyper dense lesion in the midpart of neck posteriorly, extending from C-3 to C-7 level.

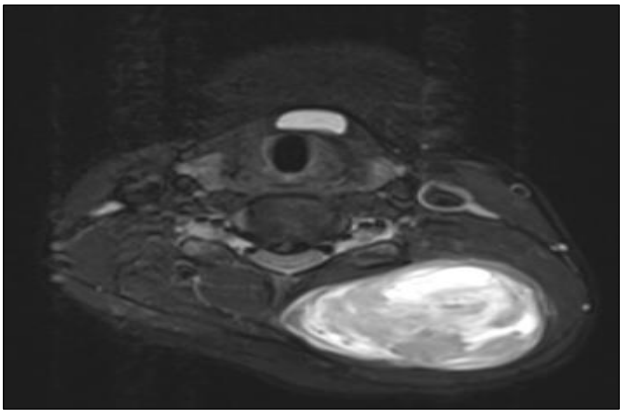

Figure 3b: T2W axial view of MRI neck shows a hyper intense lesion in the left posterolateral compartment of neck extending superiorly up to C-3 vertebra, anteriorly displacing semispinalis and posteriorly displacing Trapezius. No intraspinal extension noted. The lesion shows in between a cystic component suggestive of necrosis.

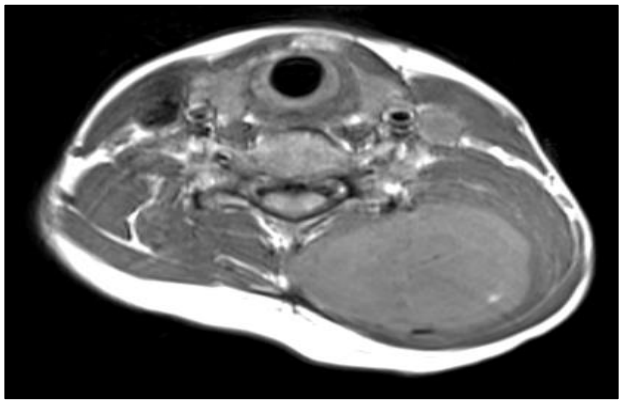

Figure 3c: T1W axial view of MRI neck shows evidence of an iso intense lesion in the left posterolateral compartment of neck extending superiorly up to C-3 vertebra, anteriorly displacing Semispinalis and posteriorly displacing Trapezius. No intraspinal extension noted.

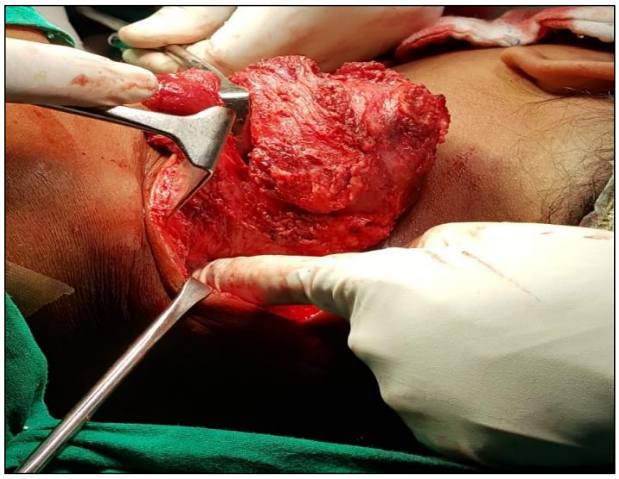

Figure 4: The excision of tumour.

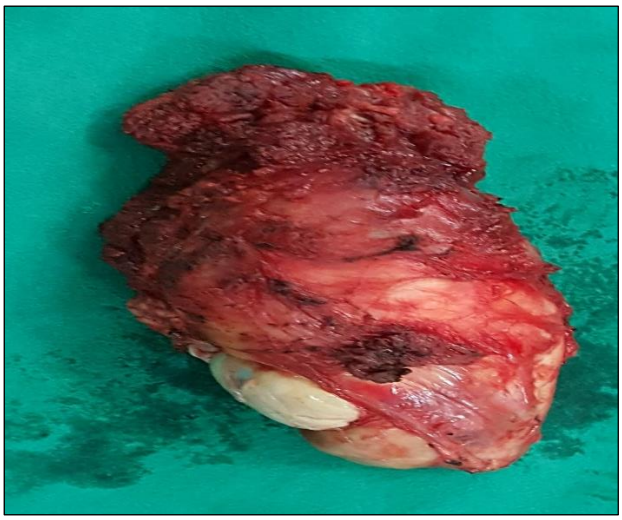

Figure 5: The excised specimen of the tumour.

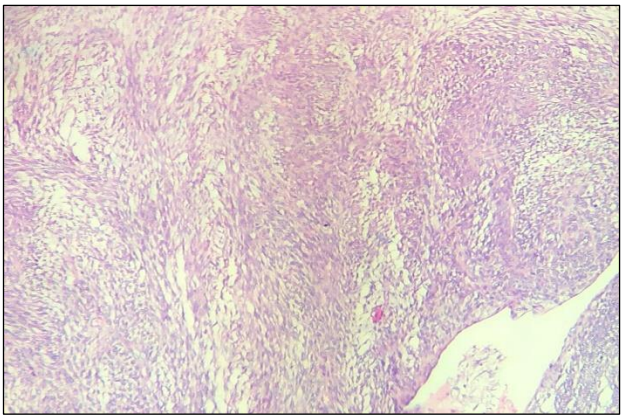

Figure 6a: Spindle cells arranged in fascicles showed palisading and whorling.

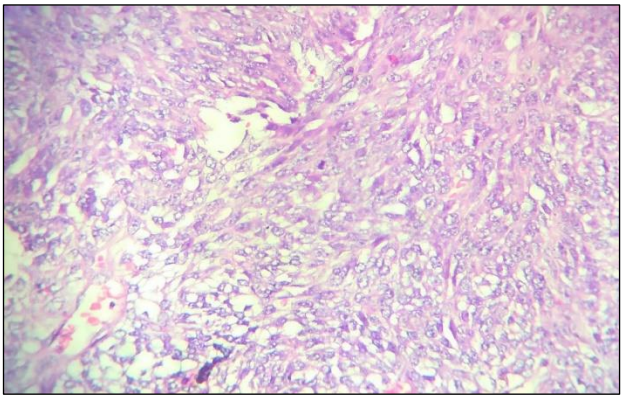

Figure 6b: Plump and hyperchromatic nuclei with prominent nucleoli and numerous mitotic figures were seen.

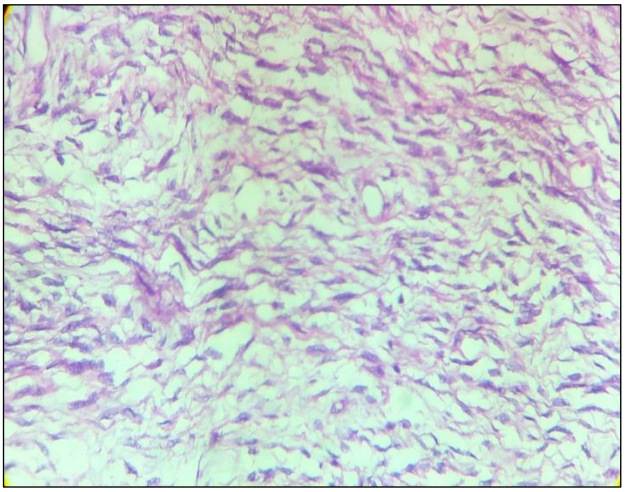

Figure 6c: Many nuclei showed serpentine appearance.

Post operatively, the patient recovered well and was discharged on postoperative day 7. She was given local radiotherapy. On a 6-month follow-up, the patient is doing well with no local recurrence or any distant metastasis.

DISCUSSION

Malignant peripheral nerve sheath tumour is an extremely rare, aggressive soft tissue sarcoma of ectomesenchymal origin with an incidence <0.001%. It arises from peripheral nerve cells like Schwann cells or perineural cells.[1]

A higher incidence of 4.6% is noted in presence of neurofibromatosis type 1.[3] These tumours more commonly occur in upper extremity (63%) and lower extremities (29.2%), followed by trunk (25%). It very rarely occurs in the head and neck region (4%).[4]

They are usually seen in adults of 20-50 years of age.[5] There is no gender preponderance.[6-8] These tumours present with pain, palpable mass, hemiparesis, or paraesthesia.[9] Histological features are those of a highly cellular spindle cell neoplasm resembling a soft tissue sarcoma but with differentiation towards elements of nerve sheath, Schwann cells and perineural cells. Frequent mitosis and focal necrosis are typical.[10]

CT scan is useful to assess the tumour extension and eventual metastasis (the more frequent are bone and lung metastasis). MRI can reveal the nerve of origin and it's more accurate to evaluate the topographical relationship of the tumour with neighboring structures especially vascular, muscular structures and fat planes involvement.[11]

Metastases occur in 39% of patients.[12,13] The most common metastatic sites are the lungs. They are also seen in soft tissue, bone, liver.[13]

Treatment is predominantly surgical. The goal of treatment is to obtain complete surgical excision with negative margins.[14]

Postoperative radiotherapy has been reported to provide benefit in high grade MPNST. Adjuvant radiotherapy is recommended for all intermediate- to high-grade lesions and for low-grade tumors after a marginal excision.[15]

Role of chemotherapy is controversial.[16] However, Moretti et al reported a 2-year overall survival rate of 80% and disease-free survival of 57% when using doxorubicin and ifosfamide in combination with surgery and radiation therapy in 10 patients with MPNSTs.[17]

Studies have shown the average 5-year survival rate to be 16-52%.[18]

CONCLUSION

A suspicion of malignant peripheral nerve sheath tumour should also be kept in mind for a swelling over the neck. Diagnosis is made by assessing a combination of clinical, radiological and pathological features. Early surgical resection followed by radiotherapy is the preferred mode

of treatment. A regular 3 monthly follow up is recommended to confirm any recurrence or metastasis.

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