International Surgery Journal
Khatri UB et al. Int Surg J. 2024 Jul;11(7):1191-1193
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20241753

Case Report

A rare case of fascicular spindle cell sarcoma of neck: a malignant peripheral nerve sheath tumor

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Received: 05 April 2024
Revised: 06 May 2024
Accepted: 14 June 2024

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ABSTRACT

Malignant peripheral nerve sheath tumor-MPNST is rare type of sarcoma making 5% to 10% of sarcoma cases. It grows in the soft tissue of the body and MPNST is sarcoma of cells of sheath that making coverings of peripheral nerve. It is more common in people with genetic condition called neurofibromatosis type-1. Here we report a case of 14-year-old male with right sided neck swelling in the last 5 years without any inflammatory signs. On radiological investigation benign soft tissue tumor- nerve sheath origin was found. We posted patient for Excision of tumor and we found highly vascular friable mass which is excised completely. On histological examination fascicular spindle cell sarcoma found. On reference to oncologist PET-CT was done suggested of no residual growth or metastasis. There is no recurrence till date in our patient.

Keywords: MPNST, Neck swelling Neurofibromatosis type-1, Sarcoma

INTRODUCTION

Malignant peripheral nerve sheath tumor, or MPNST, is a cancer of the cells that form the sheath that covers and protects peripheral nerves. Peripheral nerves are those outside of the central nervous system (brain and spinal cord). MPNST is a type of sarcoma. Sarcoma cancer grows in the soft tissues of the body, such as muscle, fat, tendons, ligaments, lymph and blood vessels, nerves, and other tissue that connects and supports the body. MPNST grows quickly and can spread to other parts of the body. Sarcomas are rare cancers and MPNST is a rare type of sarcoma, making up 5% to 10% of sarcoma cases. MPNST is most common in young adults and middle-aged adults. MPNST is more common in people with a genetic condition called neurofibromatosis type 1 (NF1). About 25% to 50% of people with MPNST have NF1. And about 8% to 13% of people with NF1 will get MPNST in their life time.[1]

CASE REPORT

A 14 years old male presented to SVP hospital on 09/09/2022 with chief complaint of swelling over right side of upper neck in the last 5 years. There was no history of pain, fever, trauma.

General physical, cardio-vascular system, respiratory, central nervous system, and abdomen examination were normal.

On local examination a single approximately 10*5 cm sized elliptical, firm, non-warm, non-tender swelling with smooth surface and regular margin extending 1 finger below midpoint of chin to angle of mandible up to just behind pinna. No fixity to overlying skin or underlying structures.

Figure 1: Clinical photograph of neck swelling.

Investigation

All routine pre operative investigations were within normal limits.

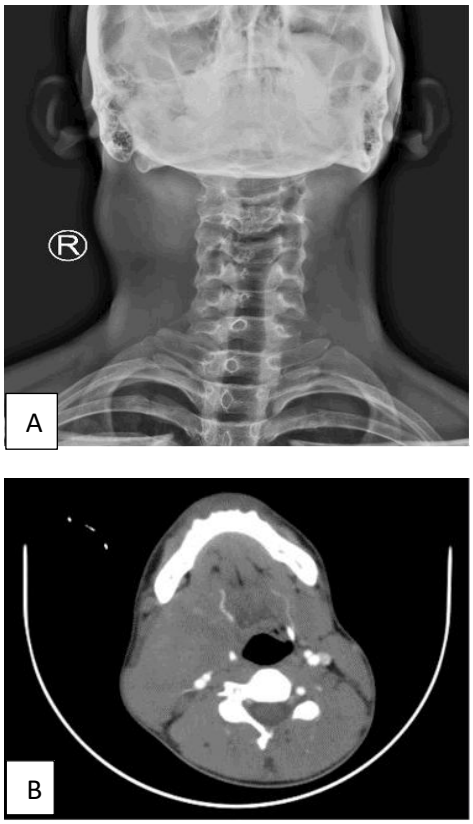

Figure 2 (A and B): Radiological images.

X-ray neck was showing soft tissue opacity. Ultrasonography NECK was suggestive of benign soft tissue tumor likely- nerve sheath origin. suggested HPE correlation. Computed Tomography neck contrast suggestive of well-defined mildly enhancing hypo dense lesion in right Para pharyngeal space.

Management

Patient was posted under general anaesthesia for right submandibular region swelling excision. Right submandibular region skin crease incision kept. Soft tissue dissected. Mass identified and dissected from all around. Neck mass was found to be inferior to mandibular gland which was highly vascular and friable. All parts of mass with cyst wall were removed preserving all vital structures and sent for histopathological examination. Haemostasis achieved and corrugated drain placed and fixed. Closure done in layers.

Histopathological examination

Sections reveal highly cellular soft tissue tumor having proliferation of spindle shaped cells with fascicular bundles arranged in herringbone pattern. Alternating hypo cellular and hyper cellular areas are seen. The spindle cells are having uniform bullet shaped nuclei and pointed wavy nuclei in hypo cellular areas. No evidence of necrosis is seen. Numerous mitoses (17-18/10 high power field) are seen. Adjacent striated muscle fibres show infiltration by the tumor.

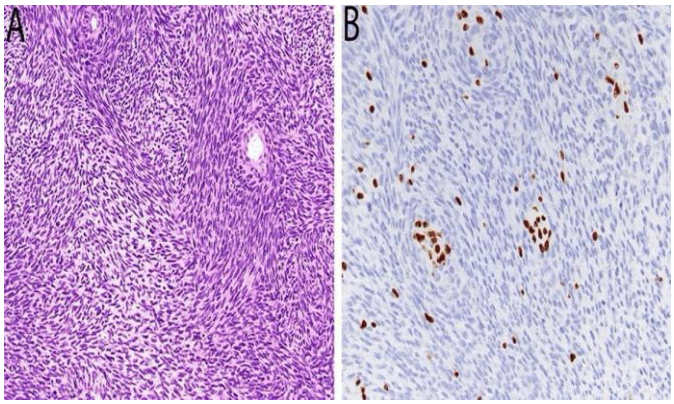

Figure 3 (A and B): Histopathological examination.

Commonest site for metastasis was regional lymph node. 8 patients had secondary deposits in liver, 2 were having deposit in anterior abdominal wall and two females were having secondary deposits in both ovaries.

Diagnosis

Fascicular spindle cell sarcoma, possibility of malignant peripheral nerve sheath tumor.

Follow up

Patient was referred to oncologist for further workup where PET-CT scan was done which suggested no residual growth or metastasis. Surveillance imaging was advised. No recurrence detected till date in patient.

DISCUSSION

Soft tissue sarcomas account for about 1% of malignant tumours and MPNST account for 5%-10% of all soft tissue sarcomas. Due to its uncommon nature, there is a paucity of data in the literature regarding prognostic factors and long-term outcomes.[2]

MPNST is more common in patients with NF-1, with an incidence of 4% to 13%, but are rare in the general population with an incidence of 0.001%. The most common age of presentation is between 20 and 50 years. Extremities, more upper than lower limb, are the usual locations (45%); the trunk (34%) and head and neck (19%) follow.[3]

Uncommon areas of presentation-like scalp, abdomen, including bladder have been reported, more often in NF1 patients. Our patient's age and presentation in the neck are consistent with the common presentation of MPNST but as he had no history of NF1; There was very little suspicion that he could present with sarcoma and the obvious diagnosis of benign soft tissue swelling was the initial, but mistaken.

CONCLUSION

Following resection, and histological confirmation of the diagnosis, treatment, according to oncology guidelines, was commenced. Despite appearing benign, at the initial presentation, it would be advisable to proceed with a complete preoperative radiological investigation of such masses, with vague clinical history. The possibility of a malignant soft tissue tumour, with a bad prognosis, must be kept in mind even in relatively straightforward cases, as a pre-operative staging could provide scope for better planning and management. Our study affirms that all resected tissues should be sent for histological confirmation of the suspected diagnosis. When intraoperative findings do not correlate with the initial presentation, the clinician should have a high index of suspicion for potential malignancy. Finally, early referral to a specialist regional soft tissue sarcoma unit is important. It increases the chances of survival.

ACKNOWLEDGEMENTS

Authors would like to thank the Head of Unit for the permission to publish this article.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: not required

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Cite this article as: Khatri UB, Parmar KD, Parikh SG, Patel PA**.** A rare case of fascicular spindle cell sarcoma of neck: a malignant peripheral nerve sheath tumor. Int Surg J 2024;11:1191-3.