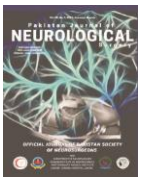

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Case Report

Recurrent Cervical Neurofibrosarcoma: A Rare Case of Malignant Peripheral Nerve Sheath Tumor of Head and Neck Region

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ABSTRACT

Neurofibrosarcoma is a malignant peripheral nerve sheath tumor (MPNST). The cervical location of the neurofibrosarcoma is very rare and is less than 1% in the literature. MPNSTs are often associated with neurofibromatosis type 1 (NF1). We are presenting a case report of 31 years old female, with huge recurrent cervical neurofibrosarcoma on the right side of the neck. To date, surgical excision followed by chemotherapy and radiotherapy is the treatment of choice which requires a multidisciplinary approach. We excised the above-mentioned cervical neurofibrosarcoma in a piecemeal fashion and discharged the patient on follow-up with the oncology department.

Keywords: Neurofibrosarcoma, Cervical, Neurofibromatosis type 1.

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INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are the lesions that arise from the peripheral nerves as the name indicates. Neurofibrosarcoma which is also MPNST arise from the cells in the sheath of the peripheral nerve and is a rare entity.[1,4]

These are highly aggressive lesions and can present at any part of the body in the location of peripheral nerves.[5] Cervical neurofibrosarcoma is very rare and less than 1% of cases of MPNST are reported in the literature, in the head and neck region.[3]

Neurofibrosarcomas of the head and neck is 10% of the malignant soft tissue tumors arising in this region and are often associated with neurofibromatosis type 1 (NF1).[4]

25% – 70% of neurofibrosarcomas are associated with NF1 and the lifetime risk of developing these lesions is 8%-13% in the NF1 population in comparison to 0.001% in the general population.[6,7]

CASE REPORT

History and Examination

We are presenting a case report of 31 years old female, presented in a Neurosurgery outdoor clinic, with complaints of a huge mass lesion about 15 cm in length and 10 cm in width on the right side of the neck. Her lesion was operated on twice in 2007 and 2012, at that time lesion was about 6cm in size and the diagnosis was a malignant peripheral nerve sheath tumor on histopathology. Now, she presented with sudden enlargement of a mass at the previously operated site from the last 4 months which was 15 cm × 10 cm in size, and pain in the neck radiating to the right arm. She also had multiple cutaneous nodules over the neck and trunk region.

Investigations

We performed an MRI neck plain and with intravenous contrast to see the extent and vascularity of the lesion which showed a large 15 cm × 10 cm large multilobulated mass in the right posterior triangle of neck and extending in C3 – C4 spinal neural foramina with post-contrast non-homogenous enhancement. Other baseline investigations for general anesthesia fitness were also done.

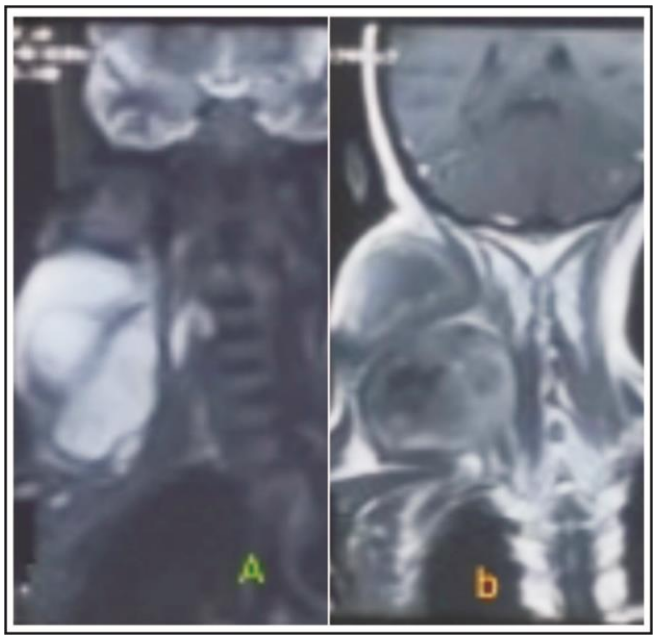

Figure 1: A = T2WI coronal section showing a large multi-lobulated mass in the right posterior triangle of the neck. B = T1WI with contrast showing non-homogenous contrast enhancement and central necrosis in the lesion.

Management

A surgical excision plan was devised to remove the lesion with the goal of maximum possible resection.

SURGERY

Surgical Technique

Recurrent neurofibrosarcoma in the cervical region having a large size with the intra-foraminal extension is challenging and a rare case from regarding surgical excision point of view. Such challenging surgeries require proper pre-operative surgical planning.

Procedure

Under general anesthesia, the patient is positioned laterally with the lesion facing upward. An elliptical incision was made over the swelling, meticulous dissection was done to expose the tumor, devascularization of the tumor was done with bipolar cautery all over the lesion. Biopsy was taken and the tumor was excised in a piecemeal manner.

Intra-foraminal extension of tumor excised with unilateral laminectomy of involved vertebrae with help of punch and biopsy forceps. Hemostasis secured. Primary closure of the wound was done, with *Redivac* suction drain in situ.

Discharge

Patient was discharged on 5th post-operative day in satisfactory condition.

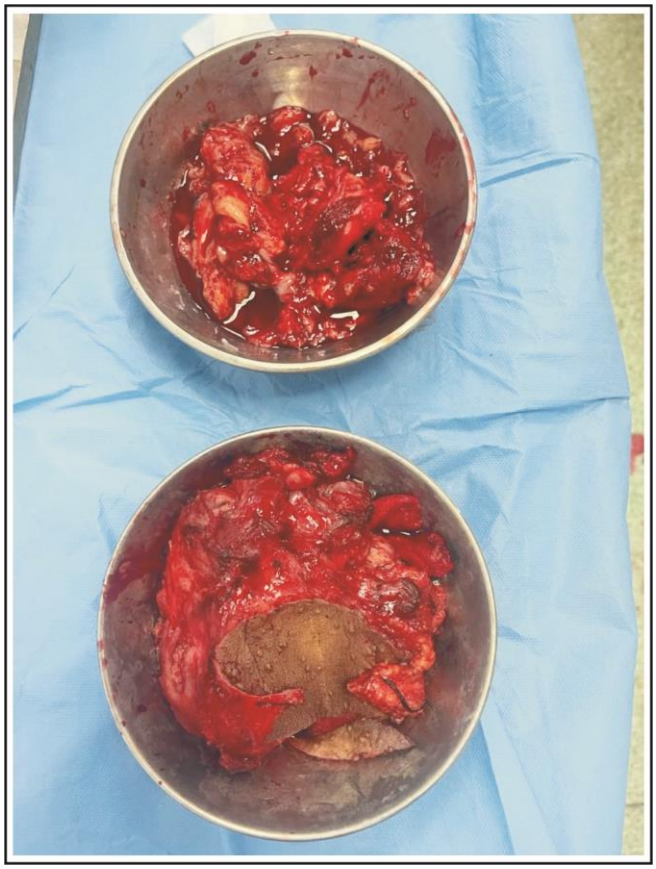

Figure 2: Piecemeal excision specimen of tumor.

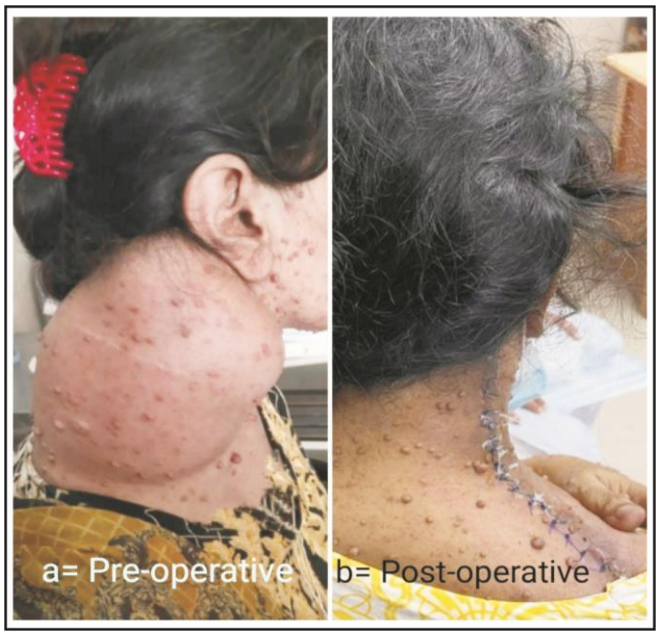

Figure 3: A = Pre-operative, B = post-operative. (images used with patient's permission)

Histopathology

Malignant peripheral nerve sheath tumor, Neurofibrosarcoma, Spindle cell type.

Micro: High-grade spindle cell neoplasm, showing marked nuclear pleomorphism and mitosis.

Immunohistochemical Stains

SO × 10	**.....**	**Focal Positive**
Desmin:	**.....**	**Negative**
CD34:	**.....**	**Negative**
EMA:	**.....**	**Negative**
TLE1:	**.....**	**Negative**
S100:	**.....**	**Negative**
INT1:	**.....**	**Negative**

Follow-up

Patient was referred to the oncology department for the chemotherapy and radiotherapy.

DISCUSSION

Neurofibrosarcoma is a type of malignant peripheral nerve sheath tumor. It is a highly aggressive tumor with a high recurrence rate.[1,2] To date best treatment is the maximum surgical excision of the tumor followed by chemotherapy. Neurofibrosarcoma of the cervical region is a rare entity and reported less than 1% in the literature.[3,4]

Due to high recurrence and aggressive nature, these tumors require a multidisciplinary approach for the treatment which involves surgery followed by chemotherapy or radiotherapy.[8,9] Cervical location of neurofibrosarcoma is challenging due to the involvement of important neural structures like the brachial plexus, spinal nerves, and spinal cord, which warrants an expert neurosurgical excision plan.

We excised above mentioned recurrent cervical neurofibrosarcoma which was extending into the

neural foramina as well. Post-operatively patient remained fine and was discharged on follow-up with the oncology department for the chemotherapy and radiotherapy.

CONCLUSION

Cervical Neurofibrosarcomas are MPNSTs, very aggressive and highly recurrent lesions. Such nature of these tumors requires a multidisciplinary approach for the treatment which includes surgery followed by chemotherapy and radiotherapy.

Informed Consent

This case report was written after the informed consent of the patient which includes the sharing of pictures as well.

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Additional Information

Disclosures: Authors report no conflict of interest.

Ethical Review Board Approval: The study was conformed to the ethical requirements.

Human Subjects: Consent was obtained by the patient in this study.

Conflicts of Interest:

In compliance with the ICMJE uniform disclosure form, all authors declare the following:

Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Mubashir Malik	1. Study design and methodology
2.	Zubair Ahmad Khan	2. Paper writing and data calculations
3.	Umer Farooq	3. Data collection and calculations
4.	Shehzad Safdar	4. Analysis of data and interpretation of results etc.
5.	Asad-ur-Rehman	5. Literature review and referencing
6.	Noman Saleem	6. Analysis of data and quality insurer