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A Rare Case of Spindle Cell Sarcoma in a Pre-existing Neurofibromatosis- A Case Report

Authors

Sumegha Rana[1], Jitin Yadav[2], Babumani Baskey[3]

[1]JR, Department of Gen. Surgery, RIMS, Ranchi, Jharkhand, India
[3]SR, Department of General Surgery, RIMS, Ranchi
Email- *sumegharana@yahoo.in, jitin_86@rediffmail.com, babamanibaski@gmail.com*
Corresponding Author
Sumegha Rana
JR, Department of Gen. Surgery, RIMS, Ranchi, Jharkhand, India
Email: *sumegharana@yahoo.in*

ABSTRACT

Aims & Objectives- *A rare case report of a case of spindle cell sarcoma occurring in a known patient of neurofibromatosis.*

Methods- *A 35 year old female patient came with complaints of rapidly increasing pain and swelling of right upper limb below elbow, along with loss of movement in right hand. She had generalized multiple swellings in whole body for last 10 years suggestive of neurofibromatosis. On examination the swelling had involved whole of the forearm which was of variegated consistency, mostly hard, temperature was raised with no tenderness. FNAC was inconclusive as well as incisional biopsy. MRI of right upper limb was suggestive of neurofibroma/neurofibrosarcoma and excision was nearly impossible as tumour had involved whole circumference of the forearm.*

Results & Conclusion- *An above elbow amputation was done and tissue was sent for HPE which made the diagnosis of spindle cell sarcoma. This was curative treatment for the patient as she came for follow up after six months with no evidence of any recurrence and artificial limb was prescribed for a better quality of life.*

1 INTRODUCTION

Spindle cell sarcoma is a rare cancer and mostly begins from skin or soft tissue surrounding. Spindle cells are seen under the microscopes which are long and narrow cells.

1.1 HISTORY

A 35 year old female patient came in our hospital with the presenting complaints of swelling in right forearm for 6 months, pain over it for two months and loss of movement in the same hand for last one month. Patient had multiple swellings over whole

body for last 10 years suggestive of neurofibromatosis. The forearm swelling started initially as a small pea sized swelling over the front of forearm just below the elbow joint with no antecedent trauma, but grew rapidly over the last two months and attained present size as that of a coconut. She also had pain over the swelling for last two months. Pain was dull aching in nature with no radiation of pain. She also had loss of movement in the same hand for one month. There was no history of fever.

There was no family history of cancer. Patient's father, mother and all siblings were alive and well. The patient was never admitted to a hospital earlier for any other illness. She had turned up to a local health set-up around 10 years back for her generalized swellings, but was returned back with assurance and no treatment.

1.2 General Examination

On general appearance, the patient was an average built female with multiple generalized swellings all over the body. Pallor, lymphadenopathy or oedema were absent.

Her right forearm had a swelling involving whole of the circumference of the forearm extending from just below the elbow to one inch above the wrist joint. Skin over the swelling was stretched. Swelling was non-tender and temperature was raised over it. Surface was irregular with variegated consistency from firm to hard. It was non mobile, fixed to underlying structures as well as to skin. Power was 0/5 in wrist joint and 2/5 in elbow joint. Lesions are shown in fig.1 and 2.

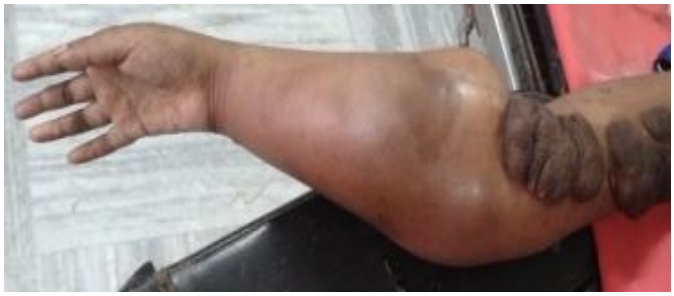

Fig.1-Swelling over the right forearm

All other systems of her body were within normal limits.

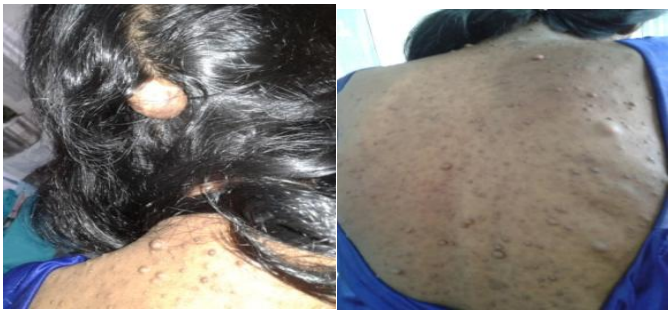

Fig. 2-Generalized swellings over scalp and back

1.3 Laboratory Investigations and Treatment

Various investigations, including all routine blood investigations as well as MRI, FNAC and incisional biopsy were done.

a. Blood Investigations

Routine blood investigations were done showing normal study as follows-

TLC - 8400/mm^3

DLC - N-64%, L-25%, M-8%, E-3%, B-0%

Haemoglobin - 12.6mg /dl

BT- 45 sec , CT- 2min.15sec.

ESR - 15mm/hr

Blood urea-20mg/dl, S.creatinine-0.9mg/dl

RBS- 86mg/dl

b.MRI

MRI of the affected limb was done which suggested the lesion as neurofibroma/neurofibrosarcoma. The MRI plates are shown in fig.-3.

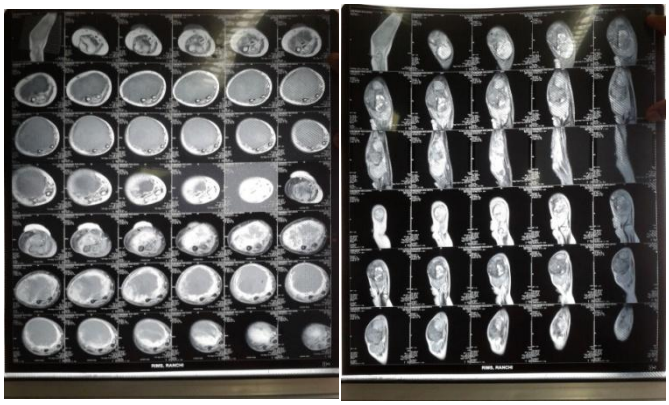

Fig.3-MRI plates of the forearm lesion

c. FNAC and Incisional Biopsy

Tissue diagnosis can be done only by FNAC and Incisional biopsy. FNAC was inconclusive so an incisional biopsy was taken from the margins of the tumour but it also showed only necrosed tissue with no viable tissue and hence it was also inconclusive.

d. TREATMENT

Since the patient had lost the functions of her limb and it was cumbersome for her to carry a useless, painful limb she gave her consent for amputation and an above elbow amputation was done and the tissue was sent for HPE which made the diagnosis of spindle cell sarcoma as shown in fig.4.

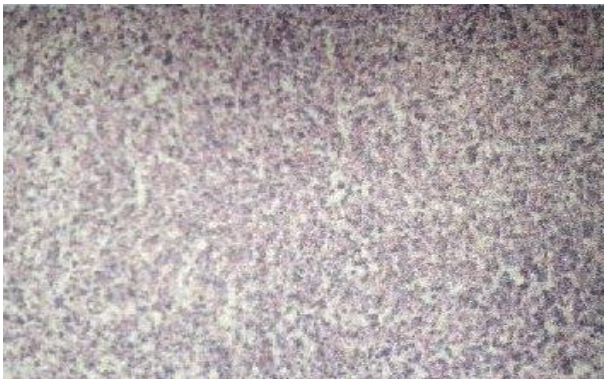

Fig.4- Histopathological picture of the lesion.

This was curative treatment for the patient as she came for follow up after six months with no evidence of any recurrence and artificial limb was prescribed for a better quality of life.

2. CONCLUSIONS

Spindle cell sarcoma is a variety of soft tissue sarcoma. It consists of nonspecific genetic alterations and typically complex unbalanced karyotypes, representing numerous genetic losses and gains. A genetic predisposition to soft tissue sarcoma has been associated with neurofibromatosis (NF) and familial adenomatous polyposis (FAP). Approximately 5% of patients with NF develop malignant peripheral nerve sheath tumours.

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