(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 5, Issue 6 , Page No: 571-574
November-December 2022

IJMSCR

Malignant Peripheral Nerve Sheath Tumor : A Rare Case Report

[1]Dr. Chandrashekhar Singh, [2]Dr. Aditya Ranot, [3]Dr. Meghraj Kundan, [4]Dr. Chintamani, [5]Dr. Ravi Pratap Singh

[1,2,5]Post Graduate Student, [3]Associate Professor, [4]Consultant and Professor, ,
[1,2,3,4]Department Of General Surgery, [5]Department Of Pathogy
VMMC And Safdarjung Hospital, New Delhi-29, India.

***Corresponding Author:**
Dr. Chandrashekhar Singh
Post Graduate Student, Department Of General Surgery,
VMMC And Safdarjung Hospital, New Delhi-29, India

Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

MPNST is a rare disease, Up to 50% of MPNSTs occur in patients with NF1 . We present a rare case of MPNST which was not associated with NF1 . A 38 year old male presents with abdominal lump with pain abdomen and vomiting . Lump was non tender, intra-abdominal, intraperitoneal and was moving with respiration. There was no family history of cancer or co-morbidity ,CECT abdomen was done and patient underwent surgery. EXPLORATORY LAPAROTOMY WITH RESSECTION OF TUMOR WITH COLOCOLIC ANASTOMOSIS & LOOP ILEOSTOMY was done and adjuvant radiation was advocated .HPE report was suggestive of MPNST.

Keywords: Biopsy, MPNST, NF-1, Pain, Lump ,CECT abdomen ,Surgery

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas which originate from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells, perineural cells, or fibroblasts. The term MPNST does not include tumor arising from the epineurium or the vasculature of peripheral nerves , but has replaced previous less well defined terms of schwannoma , neurogenic sarcoma and neurofibrosarcoma.[1] Because MPNSTs can arise from multiple cell types, the overall appearance can vary greatly from one case to the next. This can make diagnosis and classification somewhat difficult. In general, a sarcoma arising from a peripheral nerve or a neurofibroma is considered to be a MPNST.

Case Presentation

A 38 year old male of New Delhi, India presented with pain abdomen 2.5 months and lump abdomen since last 1.5 months ,on and off vomiting which was non bilious since last 15 days . On examination There was single lump of size approx. 15 x 10 cm in Right lumbar region extending to right iliac and umbilical region, intra - abdominal , intraperitoneal , moving with respiration. There was no significant past medical or surgical history.There was no comorbidity & family history of cancer .Patient was admitted and work up was done. CECT of whole abdomen was done. Patient was operated , EXPLORATORY LAPAROTOMY WITH RESSECTION OF TUMOR WITH COLOCOLIC ANASTOMOSIS & LOOP ILEOSTOMY was done and adjuvant radiation was advocated .Tumor was arising from nerve supplying the transverse colon and tumor was involving the colon also. There was also enlarged lymph nodes. It was 19cm X17 cm in size and 3.5kgs in weight.Speciemen was sent for histopathological examination .

HPE report was showing malignant peripheral nerve sheath tumour, transverse colon.

Fig. 1.CECT abdomen showing abdominal mass in cross section

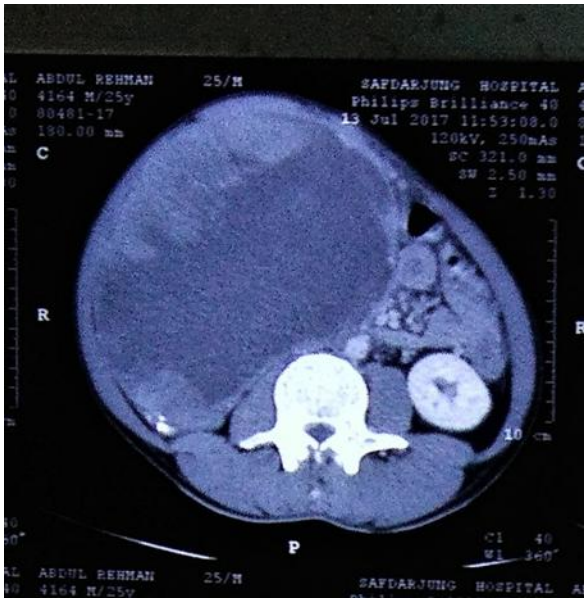
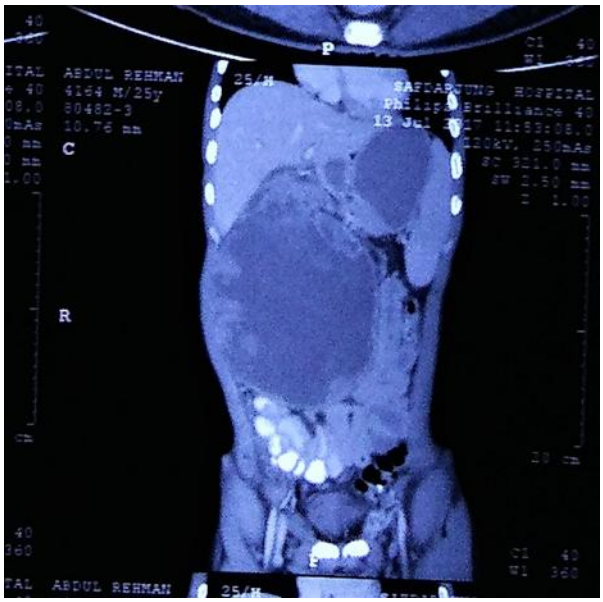


Fig. 2. CECT abdomen axial view showing tumor



Volume 5, Issue 6; November-December 2022; Page No 571-574

Fig 3

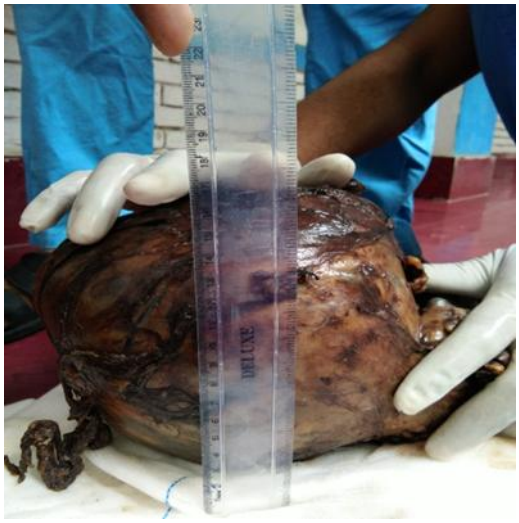


Fig 4

Measurements of tumor

FIG. 5 Tumour cells showing immunopositivity for S100.(Dab chromogen)

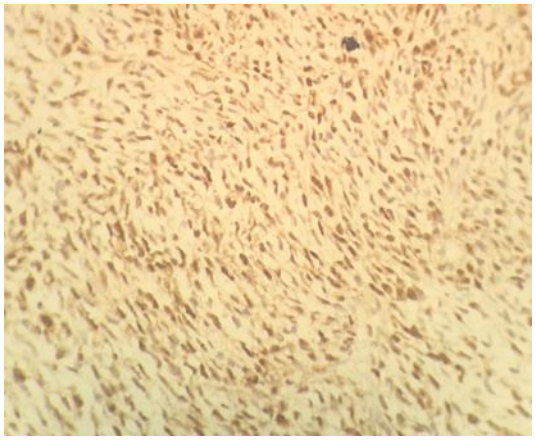
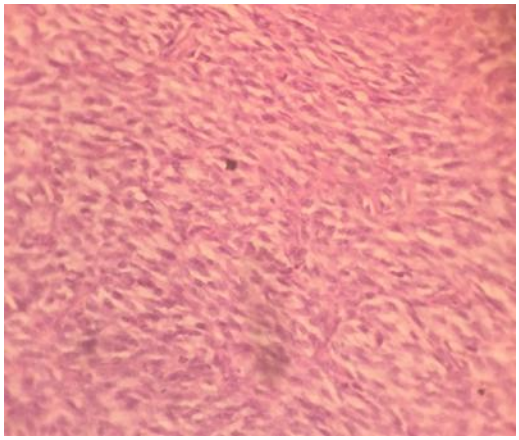


FIG. 6 MPNST (H & E)



Volume 5, Issue 6; November-December 2022; Page No 571-574

Discussion

MPNSTs comprise approximately 2% of all sarcomas, a small fractions of a group of cancers that affect 5 people per million per year.[2] They can occur either spontaneously or in association with neurofibromatosis-1 (NF1).

Up to 50% of MPNSTs occur in patients with NF1 , demonstrating the tendency for this tumor to arise from a preexisting neurofibroma. [3,4]Cross sectional studies have previously demonstrated a 1-2% prevalence of MPNST among NF1 patients although a recent study showed these patients have a 10% lifetime risk of ultimately developing an MPNST .[5,6] The etiology is unknown but there is a higher incidence in patients with a history of radiation exposure .[7,8] MPNSTs generally occur in adulthood, typically between the ages of 20 and 50 years of age as in our case . Treatment of disease is wide local excision with negative margins. Radiation therapy has become an integral part of local disease control in most soft tissue sarcomas **.**

Conclusion

MPNST is very rare disease and it progresses very rapidly .So early diagnosis of such patient is very important which could be done by radiologically and histopathology in

combination and recurrence is also very high so radiation and chemotherapy is also advisable.

References

1. Weiss SW, G. J.: Enzinger and Weiss's Soft Tissue Tumors. Edited, St. Louis, Mosby, Inc., 2001
2. Ng VY, Scharschmidt Tj, Mayerson JL,et al. Incidence and survival in sarcoma in the united states: A focus on musculoskeletal lesions. Anticancer Res. 2013;33:2597-2604.
3. .D'Agostino AN, S. E., Miller RH: Sarcomas of the peripheral nerves and somatic soft tissues associated with multiple neurofibromatosis (Von Recklinghausen's disease). Cancer, 16: 1015-27, 1963.
4. King AA, D. M., Riccardi VM, Gutmann DH: Malignant peripheral nerve sheath tumors in neurofibromatosis 1. Am J Med Genet, 93((5)): 388-92, 2000.
5. Huson SM, C. D., Harper PS: A genetic study of von Recklinghausen neurofibromatosis in south east Wales. II. Guidelines for genetic counselling. J Med Genet, 26((11)): 712-21, 1989.
6. Evans DG, B. M., McGaughran J, Sharif S, Howard E, Moran A: Malignant peripheral nerve sheath tumours in neurofibromatosis 1. J Med Genet, 39((5)): 311-4, 2002.
7. Adamson DC, C. T., Friedman AH: Malignant peripheral nerve sheath tumor of the spine after radiation therapy for Hodgkin's lymphoma. Clin Neuropathol, 23((5)): 245-55, 2004.
8. Amin A, S. A., Flanagan A, Patterson D, Lehovsky J: Radiotherapy-induced malignant peripheral nerve sheath tumor of the cauda equina. Spine, 29((21)): E506-9, 2004.

Volume 5, Issue 6; November-December 2022; Page No 571-574