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MALIGNAT PERIPHERAL NERVE SHEATH TUMOR – BIOLOGICALLY AGGRESSIVE SOFT TISSUE SARCOMAS: CASE REPORT

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Abstract

Primary retroperitoneal tumors are an exceedingly rare clinical problem. Masses in the retroperitoneum can be categorized as one of three entities: lymphomas, extragonadal germ cell tumors, and sarcomas. Malignant peripheral nerve sheath tumors are uncommon, biologically aggressive soft tissue sarcomas of neural origin that pose tremendous challenges to effective therapy

We present a young adult with retroperitoneal tumor. The patient presented a lumbar pain on the left side. Echotomography of the abdomen was madeand cystic tumor under the pancreas was detected. CT showed retroperitoneal tumor with hypodense features,with defined borders with size of 5,6x9,6 cm. Blood results and tumor markers showed no abnormalities. The patient had a surgery and whole extirpation was made without damaging the surrounding organs.

This case illustrate early diagnosis of MPNST is key to reduce mortality, with complete surgical extirpation with clear margins as treatment of choice.

Key words: Retroperitoneal tumor, soft tissue sarcomas, surgical extirpation

Introduction

Primary retroperitoneal tumors are an exceedingly rare clinical problem. Masses in the retroperitoneum can be categorized as one of three entities: lymphomas, extragonadal germ cell tumors and sarcomas.

Malignant peripheral nerve sheath tumors (MPNST) are uncommon, biologically aggressive soft tissue sarcomas of neural origin that pose tremendous challenges for effective therapy. (MPNST) are believed to derive from peripheral nerves or demonstrate peripheral nerve differentiation.

More specifically, they are defined as nerve sheath tumors arising from a peripheral nerve, from a pre-existing peripheralnerve sheath tumor, or in the setting of neurofibromatosis type 1 (NF1) syndrome[1].

MPNSTs comprise 2% of all sarcomas, a small fraction of a group of cancers that affect 5 people per one million per year [2].

Whereas MPNST may arise at any age with no gender predilection, it tends to present earlier in life than most other genomically complex sarcomas, which are generally more prevalent beyond the sixth decade. The median age for sporadic MPNST is between 30 and 60 years, and that for NF1-associated MPNST is between 20 and 40 years.

They tend towards early metastasis and often demonstrate resistance to chemotherapy [3].

We report an exceptional case of a young adult with a retroperitoneal tumor.

Clinical case

A 26-year-old patient was admitted in our clinic with retroperitoneal tumor.

The patient was diagnosed with the tumor 10 months ago. The patient had a lumbar pain on the left side. Echotomography of the abdomen was made at that time, and cystic tumor under the pancreas was detected with size of 6 cm and benign features. Others examinations were not made.

In the meantime,the patient got pregnant. After giving birth, she made echotomography to control her state, which showed that the tumor had increased, with a size of 6x9 cm. The patient was advised to make a CT with intravenous contrast.

The CT showed retroperitoneal tumor with hypodense features, with defined borders with size of 5,6 x 9,6 cm. MR of abdomen was made, and the same results were shown. Blood results and tumor markers showed abnormlities.

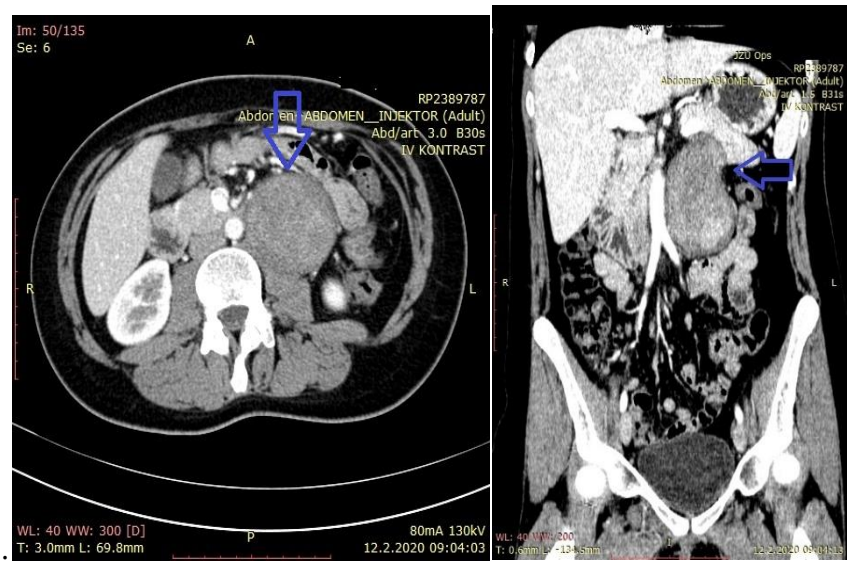


Fig. 1. CT results a) axial view b) coronal view (filled arrows-MPNST).

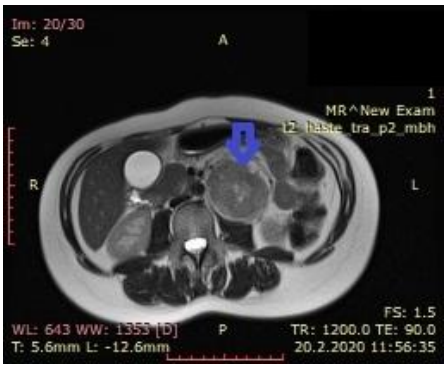


Fig. 2. MRI results a) axial view (filled arrows-MPNST).

The patient had a surgery under endotracheal anesthesia, with medial laparotomy. We found retroperitoneal tumor with a size of an orange. The tumor is spreading from pancreatic tail to aortic bifurcation. It is compressing the aorta in medial location. The lower mesenteric artery and vein presses the tumor. The tumor does not press the kidney; it does not infiltrate.We did tumor extirpation without damaging the surrounding organs. The tissue material was sent to pathology laboratory for ultimate histological analysis.

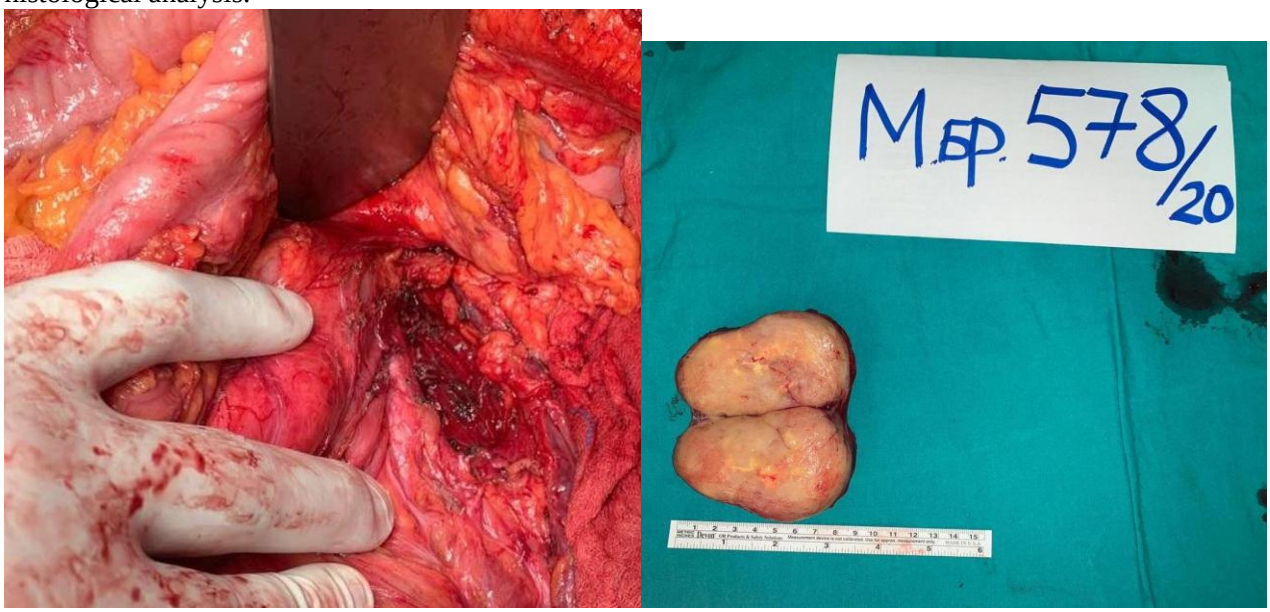
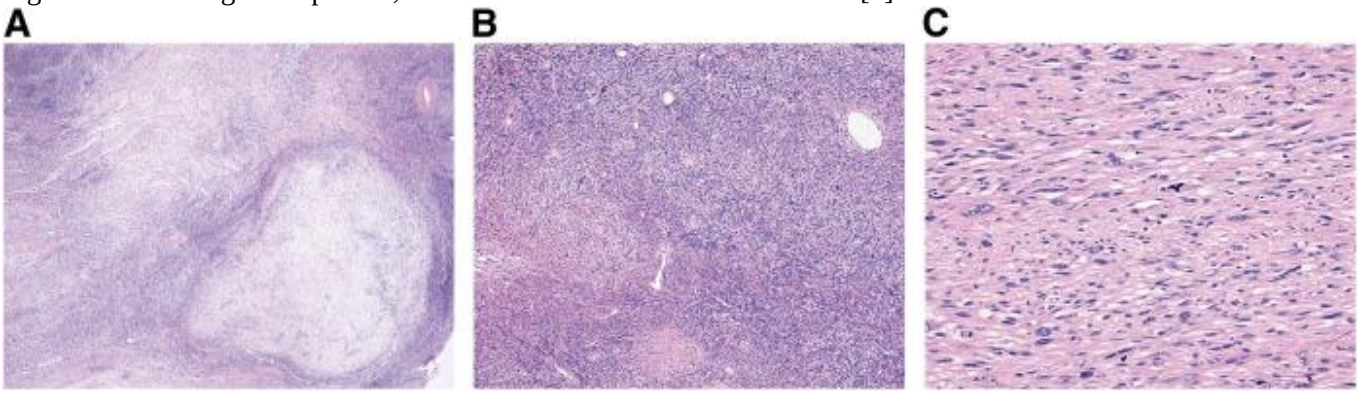


Fig.3. Intraoperative finding after removing the tumor and removed tumor

The result showed that the tumor is a low grade malignant peripheral sheath tumor. On the first postoperative day the patient was given painkillers and food per os. On the fifth postoperative day the patient was discharged from the hospital for home treatment.

Discussion

Reaching the diagnosis of malignant peripheral nerve sheaths tumorus is mainly based on imaging procedures and histological analysis, imaging procedures such as CT, MRI are needed to determine the localization and size of tumor size as well as monitor the effectiveness of therapy and relapse of illness. Histologic feature of MPNST are rather nonspecific [4].Generally, tumors are composed of monotonous spindle cells arranged in intersecting fascicles. Compared with benign neurofibromas, MPNST usually demonstrate a marked increase in tumor cellularity, pleomorphism and mitotic activity and show a more organized cellular growth pattern, with less extracellular matrix material [5].



Histopathologic features of **(A):** Low power view (×20) of MPNST demonstrating variable hypo- and hypercellular areas. **(B):** Low power view (×40) showing more cellular MPNST with scattered pleomorphic cells. **(C):** Moderate power view (×100) showing hypercellular area with intersecting fascicles of monotonous spindle cells.

In general, MPNST is known to have high metastatic potential and poor prognosis. Reported long-term outcomes vary widely across multiple series, with 5-year survival ranging between 15% and 50%. Most data on clinicopathologic factors are derived from several retrospective single institution studies analyzing between 100 and 200 patients each. Large tumor size at presentation (typically >5 cm) has been the most consistently determined adverse prognostic factor across all series. Other reported factors include tumor grade, truncal location, surgical margin status, local recurrence, and heterologous rhabdomyoblastic differentiation[6-8].

In the setting of localize disease, as is the case with all soft tissue sarcomas, complete surgical extirpation with clear margins is the treatment of choice, Multiple retrospective datasets have shown the negative prognostic impact of involved margins and local recurrence[9]. As in the case with most large (>5 cm) high-grade limb sarcomas, adjuvant radiation is advocated to reduce local recurrence[10].

The true prognostic impact of NF1 syndrome in MPNST remains somewhat in flux. Several large series report significantly worse outcomes for MPNST arising in the setting of NF1 compared with sporadic disease, with inferior responses to cytotoxic chemotherapy and 5-year survivals that are up to 50% worse [11-13].

A meta-analysis of several European studies, however, suggested that, whereas NF1 may have been negatively prognostic in studies before the year 2000, this effect was subsequently lost, possibly because of better overall surveillance and more rapid intervention at earlier stages of disease in patients with NF1 accruing from improvements in imaging and diagnostic techniques [14].

Conclusion

Early diagnosis of MPNST is of crucial importance to reduce mortality rate among affected patients. We are in need of a better understanding of the biology of, and refining clinical biomarkers for transformation of benign neurofibromas into MPNSTs in the setting of NF1. In this rare and diagnostically challenging disease, efforts already underway must aspire toward innovative trial designs that maximize

patient resources, buttressed by multicenter collaborations to improve the quality and quantity of meaningful translational and clinical data. With these efforts, the significant challenges to improve care for patients with this form of soft tissue sarcoma can be met.

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