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MALIGNANT PERIPHERAL NERVE SHEATH TUMOR (MPNST) – CASE REPORT AND REVIEW OF LITERATURE

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Summary
Malignant peripheral nerve sheath tumors (MPNSTs) are locally invasive soft tissue sarcomas. These tumors constitute up to 10% of all soft tissue sarcomas. They arise from minor or major peripheral nerve branches or sheath of peripheral nerve fibers. They occur with equal frequency in males and females. The majority of malignant peripheral nerve sheath tumors are observed in the extremities. Because of its rarity, this neoplasm is often misdiagnosed. It is characterized by an aggressive behavior and rapid growth. The incidence of malignant peripheral nerve sheath tumor is low with a lifetime risk of 0,001%.
The author of this article presented a case of a 76-year-old Caucasian woman who was admitted to the Department of Surgical Oncology because of malignant peripheral nerve sheath tumor (MPNST) which was located on the right side of inguinal and pubis. The progression of disease was very fast. The author performed a literature review on malignant peripheral nerve sheath tumor (MPNST) diagnosis and treatment.

Key words: malignant peripheral nerve sheath tumor, diagnosis, treatment

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are locally invasive soft tissue sarcomas (1, 2). These tumors constitute up to 10% of all soft tissue sarcomas (1). Because of its rarity, this neoplasm is often misdiagnosed (3, 4). It is characterized by an aggressive behavior and rapid growth (2, 4, 5). The incidence of malignant peripheral nerve sheath tumor is low with a lifetime risk of 0.001% (1).

CASE REPORT

A 76-year-old Caucasian woman was referred to the Department of Surgical Oncology due to tumor which was located on the right side of inguinal and pubis (fig. 1, 2). For a three months she reported rapidly growth of tumor. Pathological examination which was made from open surgical biopsy of the tumor in outpatient room showed: malignant peripheral nerve sheath tumor (MPNST). She reported general weakness and malaise. In the interview, the patient reported no fever.

She had no any other symptoms, there was no history of weight loss and loss of appetite. The patient was treated chronically for hypertension disease. She had no surgeries and there was no history of carcinoma in patient family. Blood test and other routine hematological examinations and biochemical tests were within normal limits.

In the Department of Surgical Oncology the patient was prepare for surgery. Considering the malignant tumor was located in the right region of inguinal and pubis, an wide excision with inguinal lymphadenectomy was performed (fig. 3). During a surgical procedure, there was no bleeding. Patient after surgery felt good and did not complain of pain. The postoperative period was uncomplicated and the patient left the ward in the 4th day after surgery.

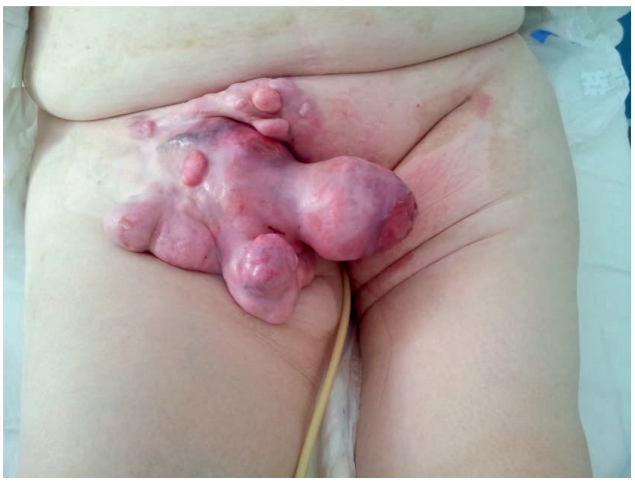

Fig. 1. A 76-year-old Caucasian woman with tumor which was located on the right side of inguinal and pubis.

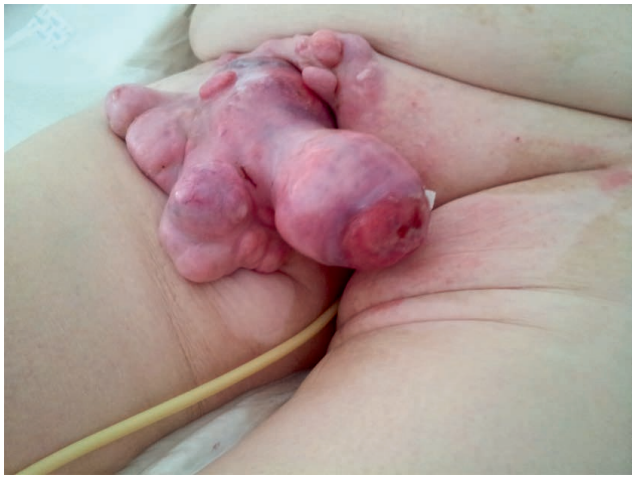

Fig. 2. Malignant peripheral nerve sheath tumor (MPNST) on the right side of inguinal and pubis.

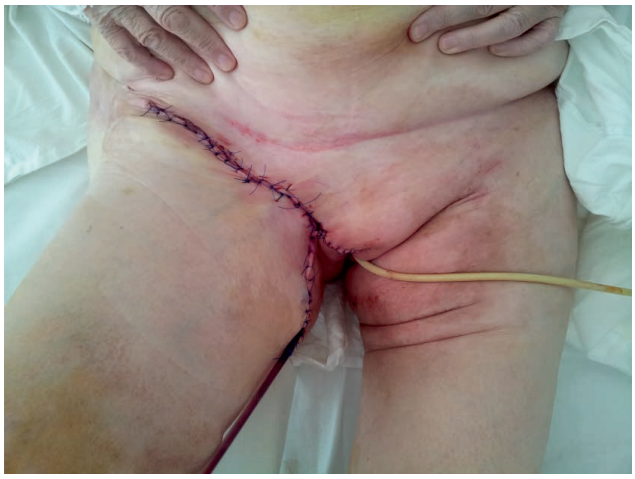

Fig. 3. The scar after wide excision with inguinal lymphadenectomy due to malignant peripheral nerve sheath tumor (MPNST).

Histopathology examination of material showed malignant peripheral nerve sheath tumor (MPNST). Tumor cells were S100 (+ in some cells), desmina (-), CD34 (++), caldesmon (-), Ki67 (+ in 40% cells), HMB45 (-), melan A (-), SMA (-/+), pan CK (-), CD99 (++), vimentyna (+++), CD117 (-). Histopathologial examination did not show metastases in removed lymph nodes.

The patient is now under the care of oncological outpatient at our hospital. After two months of surgery performed, there was no recurrence or metastasis.

DISCUSSION

Malignant peripheral nerve sheath tumor (MPNST) is also known as “malignant schwannoma”, “neurosarcoma” and “neurofibrosarcoma” (1-3). These tumors are locally invasive soft tissue sarcomas which are derived from Schwann cells or pluripotent cells of the neural crest (6-8). They arise from minor or major peripheral nerve branches or sheath of peripheral nerve fibers (9, 10). These tumors account only 5% of malignant soft tissue tumors (6). They occur with equal frequency in males and females (9). The majority of malignant peripheral nerve sheath tumors are observed in the extremities, but also they are seen in unusual sites such as: the pelvic retroperitoneum and infratemporal fossa.

Malignant peripheral nerve sheath tumors (MPNST) create diagnostic problems because of cellular origin and pathological similarities with sarcomas cells such as leiomyosarcoma, fibrosarcoma and monophasic synovial sarcoma (5).

Imaging is routinely performed to plan surgical resection. MRI is investigation of choice and it can reveal the nerve of origin and its relationship to other important structures (11).

Surgery plays important role in multimodal therapy in malignant peripheral nerve sheath tumors (MPNST). Radical wide surgical resection is the treatment of choice. Radical lymph node dissection is not indicated. Amputations are not indicated but if wide resection is not possible or patient has compromised limb function amputation is permitted.

Malignant peripheral nerve sheath tumor (MPNST) are considered as a chemotherapy and radiotherapy resistant tumors but some authors recommend their using postoperative. The oncology consensus group recommend postoperative radiotherapy as a part of treatment policy for MPNSTs (12).

Malignant peripheral nerve sheath tumors characterized the highest recurrence rate of any sarcomas (13). Adequate surgical treatment gives the best chance of survival (14). The treatment of malignant peripheral nerve sheath tumors (MPNSTs) very often need multimodal therapy, so that it is important to treat patients in high specialized oncological centers.

CONCLUSIONS

1. Malignant peripheral nerve sheath tumors (MPNSTs) are locally invasive soft tissue sarcomas.
2. These tumors characterized by an aggressive behavior and rapid growth.
3. The majority of malignant peripheral nerve sheath tumors are observed in the extremities.
4. MRI is investigation of choice and it can reveal the nerve of origin and its relationship to other important structures.
5. Radical wide surgical resection is the treatment of choice.

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