

Malignant peripheral nerve sheath tumor of the vulva: A case report

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ABSTRACT

Malignant peripheral nerve sheath tumors (MPNST) constitute about 10% of soft tissue sarcomas and most MPNSTs arise in association with major nerve trunks, including the sciatic nerve, brachial plexus, and sacral plexus. The most common anatomical sites include the upper and lower extremities and trunk and less commonly the head and neck. Only few cases have been reported in the female genital tract. Our case is 63 years old and reported as epitheloid type MPNST in the left labium majus. The epitheloid type MPNST is seen extremely rare and displays some common features with carcinoma and melanoma. Cords of epitheloid cells grouped in a nodular fashion were seen in the hematoxylin-eosin stained sections of the well circumscribed, lobulated tumor. Immunohistochemically, the tumor showed positive reaction with S-100 protein, vimentin, EMA and high molecular weight keratin. There was also positive immune reaction with desmin and HMB-45 in the dedifferentiated areas of the tumor. As the tumor had no relation with the surface epithelium, it was distinguished from malignant melanoma. The case was reported as epitheloid type malignant peripheral nerve sheath tumor. [Turk J Cancer 2006;36(1):31-34].

KEY WORDS:

Malignant peripheral nerve sheath tumor, vulva

INTRODUCTION

Vulva cancers constitute approximately 5% of all gynecological malignancies and more than 90% of them are squamous cell carcinomas (1). Primary malignant mesenchymal tumors of the vulva are very rare. They are frequently located in the labium majus (2). There are only 5 cases of peripheral nerve sheath tumor of the vulva reported until now (3-7). Epitheloid type malignant peripheral nerve sheath tumors (MPNST) constitute less than 5% of all MPNSTs. They are commonly seen in patients between 20-50 years old. The cases reported in the literature mostly arise in association with the major nerves including the sciatic nerve, tibial nerve, peroneal nerve, fascial nerve and digital nerves. Epitheloid type MPNSTs share some common features with carcinomas and melanomas. The most characteristic appearance is short cords of large epitheloid cells arranged in a vague nodular pattern. The tumor cells usually have prominent nucleoli similar to melanoma cells. The tumors may appear myxoid or cellular depending on the accumulation of acid mucin. Immunostaining for the antigens like S-100 and Leu-7 aid in showing the nerve sheath origin of the tumor. Fifty to ninety percent of MPNSTs display focal positivity for S-100 protein. About 80% of epitheloid type MPNSTs are strongly and diffusely positive for S-100 protein (8).

CASE REPORT

Our case, a 63-year-old female patient, was admitted to Akdeniz University, Faculty of Medicine, Department of Pathology with consultation of 5 paraffin blocks and 5 Hematoxylin-eosin stained preparations. The serial sections of the paraffin blocks showed tumor cells in the entire tissue. The tumor was lobular in architecture and contained central necrosis in some regions. The tumor cells were large with eosinophilic cytoplasm and prominent nucleoli (Figure 1). The differential diagnosis included malignant melanoma, malignant epithelial tumor, epitheloid sarcoma and other malignant mesenchymal tumors. The histochemistry and immunohistochemistry aided in the differential diagnosis. The reticulin stain showed reticular fibers surrounding the tumor cells in groups (Figure 2).

There was positive immune reaction for S-100, vimentin and high molecular weight keratin in the cytoplasm of the tumor cells and also for desmin and HMB-45 in the dedif-

ferentiated areas of the tumor (Figures 3-8). The tumor cells did not show immunoreactivity for CD34 and Factor VIII. About 2 months later, the patient underwent an operation and we examined a specimen including the mass and the inguinal lymph nodes. The gross examination revealed a tumor of 6 cm in diameter with no relation with the epidermis over the specimen. The tumor was pink in color and lobulated in appearance. Cysts, hemorrhage and necrosis were detected in some areas.

The hematoxylin-eosin stained sections of this tumor revealed a tumor identical with the previous one. The immunohistochemical findings of both tumors were also common. The tumor had no connection with the epidermis, it was located in the soft tissue. With all these findings, the case was reported as epitheloid type MPNST. The patient underwent whole body scan for primary focus and no other focus could be identified. The patient was accepted as primary MPNST of the vulva.

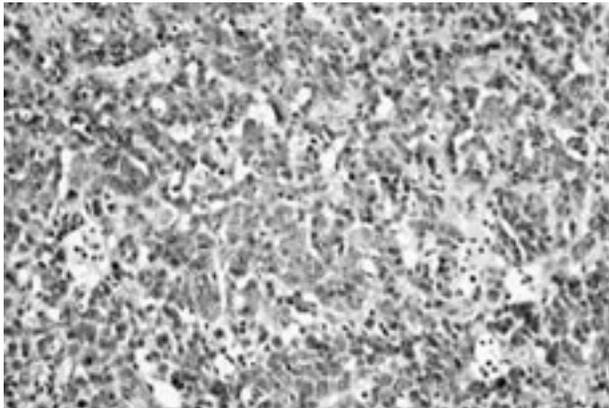


Fig 1. Epitheloid cells with large, eosinophilic cytoplasm and prominent nucleoli (H&E, x200)

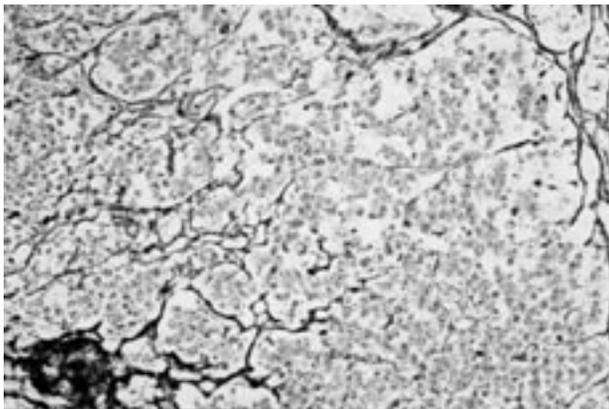


Fig 2. Reticular fibers surrounding tumor cells in groups (Reticulin, x200)

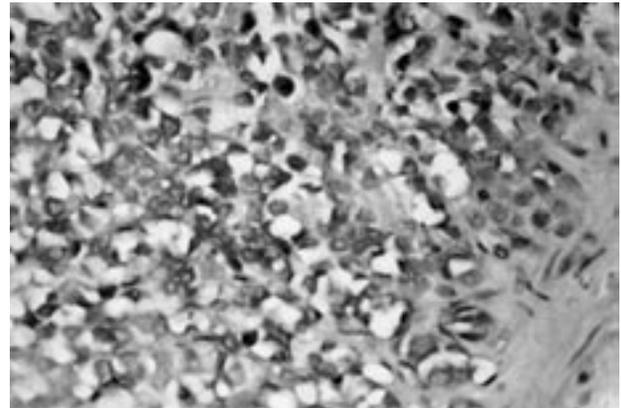


Fig 3. S-100 (DAB, x400)

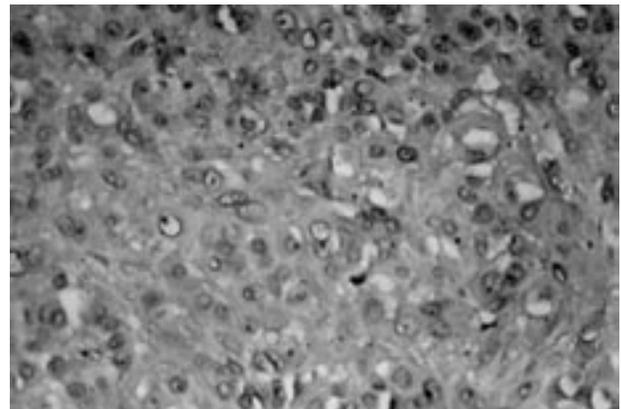


Fig 4. Vimentin (DAB, x400)

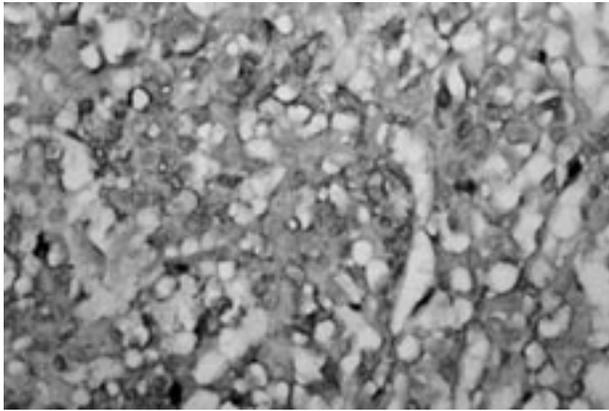


Fig 5. EMA (DAB, x400)

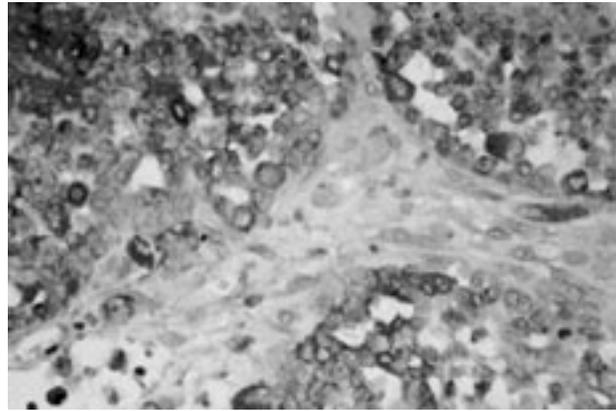


Fig 7. Desmin (DAB, x400)

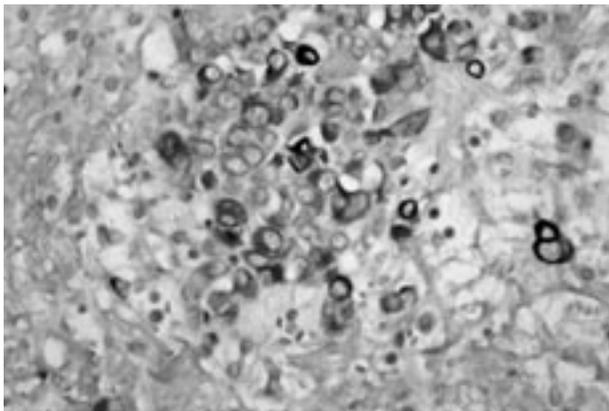


Fig 6. High molecular weight keratin (DAB, x400)

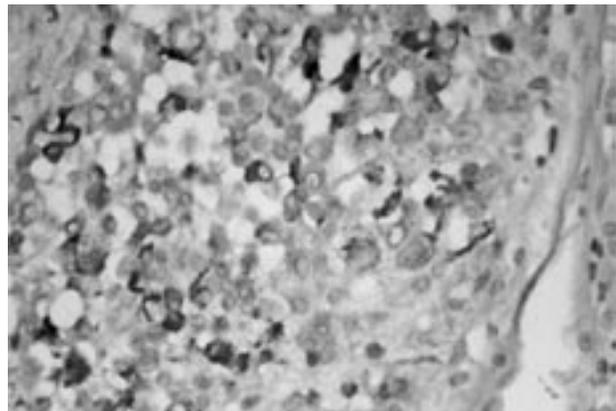


Fig 8. HMB-45 (DAB, x400)

DISCUSSION

MPNSTs constitute approximately 10% of all soft tissue sarcomas and arise from the major nerve trunks (8). The most common localizations are neck, arm, leg and hip. While they are usually seen in adults, there are some cases reported in the childhood (9). Less than 5% of all MPNSTs are of the epitheloid type and the reported cases frequently arise from the major nerve trunks (8). However, primary sarcomas of the vulva are extremely rare and epitheloid sarcoma is the most common in this region (1,8). There are also some MPNST cases reported in vulva (3-7). MPNSTs are commonly seen in patients 20-50 year-old. Although there is variation in age, the cervical MPNSTs reported in the literature are usually above 50 year-old (10-12). Our case was 63 year-old, either.

MPNSTs are usually more than 5 cm in diameter and the tumor in our case was also 6 cm in diameter with cysts, hemorrhage and necrosis on the cut surface (8).

The histology of the MPNSTs varies much but the most characteristic finding is short cords of large, epitheloid cells with prominent nucleoli. The nodular pattern is also an important feature. Though epitheloid sarcoma shows nearly the same nodular pattern, central degeneration and necrosis of tumor cells which is characteristic for this tumor, is not seen in MPNST with epitheloid type. Half of the cases show positive immune reaction for the schwann cell markers like S-100 and Leu-7 (8). The Hematoxylin-eosin stained sections of the tumor revealed cords of large tumor cells with prominent nucleoli similar to melanoma cells. Having no connection with the overlying epidermis, the tumor was differentiated from malignant melanoma. Though we found HMB45 positivity in the dedifferentiated cells, it was considered as an aberrant immunoreactivity. Positive immune reaction for S-100 protein, vimentin, EMA and high molecular weight keratin were detected in the tumor cells. The case was reported as epitheloid type MPNST with these histochemical and immunohistochemical findings.

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