Malignant peripheral nerve sheath tumor of the vagina

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ABSTRACT

عادتاً ما تتطور أورام غلاف العصب المحيطي الخبيثة (MPNSTs) في العصب الرئيسي للجذع، وتزيد من الأورام التي في الأقسام القريبة للأطراف العلوية، السفلية والجذع. يعد ظهور أورام غلاف العصب المحيطي الخبيثة (MPNSTs) بشكل آلي في الجهاز التناسلي الأنثوي أمر نادر. نستعرض في هذا التقرير حال لسيدة مصابة بورم غلاف العصب المحيطي الخبيثة (MPNST) في المهبل، ومناقشة خطوات التشخيص والعلاج. تمت معالجة المريضة بشكل نهائي عن طريق استئصال موضعي جذري. كانت النتيجة جدية حيث لم يكن هناك أي بوادر لظهور الورم بعد عامين من عملية الاستئصال.

Malignant peripheral nerve sheath tumors (MPNSTs) usually develop in major nerve trunks, giving rise to tumors in the proximal portions of the upper and lower extremities and trunk. The MPNSTs primarily occurring in the female genital tract are quite rare. We describe a case of MPNST of the vagina and discuss its diagnosis and treatment. This patient was treated definitively with radical local excision and is well with no signs of recurrence 2 years postoperatively.

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Malignant peripheral nerve sheath tumors (MPNSTs), which are also known as neurofibrosarcomas and malignant schwannomas, are defined as any malignant tumor arising from or differentiating toward cells restricted to the nerve sheath, including schwann cells or perineural cells. The MPNSTs are rare tumors with an expected incidence of one per 10^6 people per year.[1] The MPNSTs often occur in patients with neurofibromatosis type 1 (NF-1), an autosomal-dominant disorder that involves the NF-1 tumor suppressor gene, which is located on chromosome 17.[2] Most MPNSTs usually arise in association with major nerve trunks, including the sciatic nerve, brachial plexus, and sacral plexus, thus contributing to tumors in the proximal portions of the upper and lower extremities and trunk. Minimal data exist on MPNSTs primarily occurring in female genital organs. Up to now, only a few cases of MPNSTs arising in the uterine corpus, cervix, or vulva has been documented in the English literature. We report a rare case of an MPNST primarily occurring in the vaginal wall, to highlight the accurate diagnosis and optimal therapy of this unusual tumor.

Case Report. A 41-year-old gravida 2, para 1011 complained of noting a mass in the vagina prior to presentation. A round, well-circumscribed tumor protruding from the left vaginal wall was noted on examination (2.5 cm in diameter and 2 cm from the introitus). The tumor was rubbery in texture and beneath the vaginal mucosa. Both the uterus and bilateral adnexa were within normal limits. The rectovaginal examination suggested that the mass was located in the vaginal wall without apparent extension to the rectovaginal septum. There were no abnormal findings on the remainder of the physical examination and medical history. A vaginal mass biopsy was performed, and the original histopathologic diagnosis was leiomyosarcoma. To check for metastasis, she underwent systemic CT scan and it showed there was no evidence of lymphadenopathy or distant metastases. A radical local excision of the vaginal tumor was performed, removing the total tumor and at least 1 cm of surrounding tissue of the tumor. Grossly, the tumor was mainly located beneath the vaginal mucosal layer, and was not encapsulated. The rectum was not involved. The cut surface of the tumor was homogeneous without necrosis. Microscopically, the tumor was composed of spindled cells arranged in interlacing bundles. The neoplastic cells had abundant eosinophilic cytoplasm. The nuclei were hyperchromatic, partially with vesicular chromatin (**Figure 1**). The average mitotic count was between 7 and 8 mitotic figures per 10 high-power fields, and significant nuclear

pleomorphism was lacking. Immunohistochemistry showed that the tumor cells had strong diffuse positive staining for S-100 protein (**Figure 2**) and vimentin (**Figure 3**), but were not immunoreactive for cytokeratin, smooth muscle actin, desmin, or neuron-specific enolase (NSE). The final pathologic diagnosis was confirmed as an intermediate malignant peripheral nerve sheath tumor, and the tumor-free excision margin was 5 mm on pathologic analysis. A regular gynecologic examination and Pap smear of the vaginal region from which the tumor had been resected were performed at 3-month intervals for 2 years. Recently, a systematic CT scan revealed that there were no signs of local recurrence or remote metastasis.

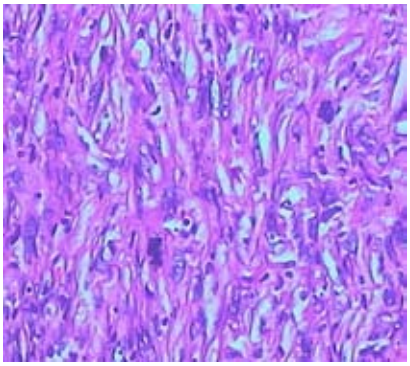

Figure 1 - Malignant peripheral nerve sheath tumor of the vagina. the tumor displays a fascicular growth pattern and is composed of crowded spindled cells with hyperchromatic nuclei (hematoxylin and eosin stain, original magnification x 400).

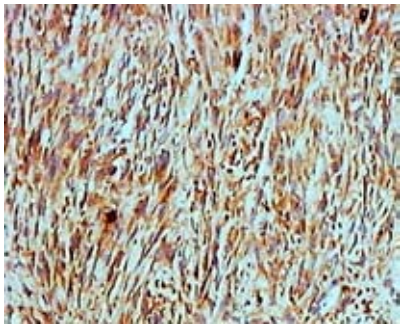

Figure 2 - Tumor cells show strong immunoreactivity to S-100 protein (original magnification x 200).

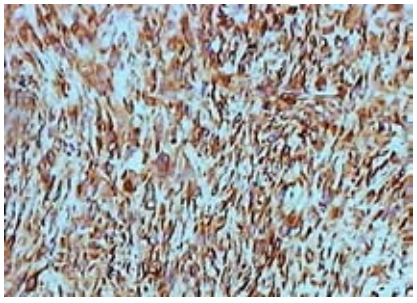

Figure 3 - Positive immunostaining for vimentin (original magnification x 200).

Discussion. Primary MPNSTs of the female genital tract are very rare. We report an MPNST of the vagina. The diagnosis of MPNST is primarily based on clinical features, pathologic appearance, and immunohistochemistry. It is not difficult to make a diagnosis of MPNST when a sarcoma arises from a peripheral nerve or from a pre-existing benign neurofibroma, especially in the setting of NF-1. Outside of the context of NF-1 or involvement of a nerve, it can be quite challenging to achieve such a diagnosis because of the relatively non-specific histologic and immunohistochemical findings. In our patient's circumstance, the vaginal soft-tissue tumor lacked an anatomic relationship to the nerves, and she had no history of neurofibromatosis. At this time, the histologic features and immunohistochemistry were crucial in confirming the diagnosis of this unusual tumor. The characteristic pathologic appearance of MPNST includes fascicles of spindle cells with intervening hypocellular myxoid, edematous, or fibrous areas. The spindle cells are typically large and uniform, which usually possess hyperchromatic nuclei and eosinophilic cytoplasm, and absent coagulative tumor necrosis, and severe nuclear atypia. Low-grade MPNSTs are less cellular and mitotically active than their high-grade counterparts.[3] Although there is no pathognomonic immunohistochemical marker for MPNST, the following markers can be helpful in the diagnosis of MPNSTs. The S-100 protein is the most commonly used antibody to identify nerve sheath differentiation, but <50% of MPNSTs are weakly or focally positive for S-100; additional markers for nerve sheath differentiation, such as glial fibrillary acidic protein(GFAP), CD56/neural cell adhesion molecule-1 and neurofilaments, can also be useful.[3] Most often, MPNSTs show strong staining for vimentin, whereas absence of staining to smooth muscle markers, such as desmin or muscle-specific actin, aids in ruling out a diagnosis of a malignant smooth muscle neoplasm. Furthermore, electron microscopy also can be used in demonstrating the histologic features of perineurial cell differentiation, and it is significantly helpful in the accurate diagnosis of this uncommon lesion. To date, the standard treatment modality has not been fully established because of the low incidence of MPNSTs. Surgery represents the mainstay of therapy. The paramount objective of surgery is complete removal of the tumor with histologically clear margins of resection, which can provide a chance for a cure. The MPNSTs have a high likelihood of producing local recurrence and distant metastasis. A multidisciplinary curative

modality has been adopted by some centers to improve local control and reduce distant metastasis, although benefits from adjuvant radiotherapy and chemotherapy in the treatment of MPNSTs have not been proven. Our patient did not undergo any adjuvant management after a tumor-free surgical margin was achieved by a radical local excision of the vaginal tumor. There was no evidence of recurrence or metastasis after 2 years of surgery. Our case provides evidence that complete surgical resection may be sufficient in the treatment of vaginal MPNSTs.

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Related topics

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