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Epithelioid malignant peripheral nerve sheath tumor of the uterine corpus

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Abstract Epithelioid variant of a malignant peripheral nerve sheath tumor (MPNST) is a rare sarcoma. Rarer still is its occurrence at uncommon sites like the uterine corpus where an index of suspicion for this diagnosis is extremely low. Herein, we report a rare case of a uterine epithelioid MPNST in a young girl who underwent a total abdominal hysterectomy for a uterine tumor that was initially diagnosed as an undifferentiated sarcoma and whose paraffin blocks were submitted to us for review. Biopsy sections showed a malignant tumor, predominantly composed of polygonal cells, including "rhabdoid" forms with conspicuous mitoses. On immunohistochemistry, tumor cells were diffusely positive for vimentin and S-100 and negative for smooth muscle actin, desmin, myogenin cytokeratin, epithelial membrane antigen, melan A, HMB-45, CD10, glial fibrillary acid protein inhibin, synaptophysin, chromogranin, MIC2, FLI-1, and neuron-specific enolase. Diagnosis of an epithelioid MPNST was offered. The case is presented in view of its rarity and also to highlight the value of immunohistochemistry in objectively identifying unusual sarcomas at uncommon sites.

Keywords: Epithelial malignant peripheral nerve sheath tumor; Uterine mesenchymal tumor; Uterine sarcoma; Immunohistochemisty; S-100 protein

1. Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas that originate from peripheral nerves or from nerve sheath cells like Schwann cells, perineural cells, or fibroblasts [1]. Histologically, these are invariably composed of spindle cells. An epithelioid differentiation is relatively uncommon in an MPNST, designated as an *epithelioid MPNST* [2]. Malignant peripheral nerve sheath tumor has been rarely documented at unusual sites like uterus and cervix where an index of suspicion for its diagnosis is exceedingly low, over the relatively more common leiomyosarcoma and endometrial stromal sarcoma (ESS) [3-5]. Immunohistochemical (IHC) analysis is essential to substantiate this diagnosis [1-3]. Herein, we report a rare case of an epithelioid MPNST of the uterine corpus with the role of IHC in sorting out this entity from its other differentials.

2. Case presentation

A 22-year-old unmarried, nulliparous lady presented to her gynecologist with a 2-month history of irregular vaginal bleeding, associated with a pelvic mass and mild abdominal pain. On examination, she had mild pallor. Per abdominal examination showed a protuberant uterus (22 weeks' size). Per speculum examination revealed mild vaginal discharge. Per vaginal examination showed uterus of 22 weeks' size, firm with restricted mobility. Bilateral adnexae and fornices were free. The pouch of Douglas was unremarkable.

Her hemoglobin was low, that is, 8.1 g/dL. Results of all the other laboratory investigations were within normal limits. She underwent radiological investigations elsewhere and, subsequently, a total abdominal hysterectomy with bilateral salpingopherectomy. Paraffin blocks from the uterine tumor were submitted to us for review.

2.1. Radiological findings

Magnetic resonance imaging revealed a large fusiform tumor, invading the fat planes and displaying heterogeneity,

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ill-defined margins, and edema surrounding the tumor. Ultrasonography showed a large irregular hypoechoic lesion of 10 × 8-cm size, arising from uterus with areas of degeneration. Bilateral adnexae were normal.

2.2. Pathologic findings

Grossly, as per the referring pathologists' notes, a uterine tumor was identified with a fleshy appearance and pale yellow areas on cut section. Microscopically, it was diagnosed as an undifferentiated sarcoma.

On review, conventional hematoxylin and eosin (H & E)–stained sections showed an ill-defined nodular tumor composed of oval to short spindle cells arranged in sheets and fascicles with prominent areas showing epithelioid morphology in the form of well-defined, abundant eosinophilic cytoplasm with focally eccentric nuclei, leading to "rhabdoid" morphology. Nuclei showed moderate pleomorphism with brisk mitotic activity. There was no necrosis or concentric periarteriolar arrangements of tumor cells. (Fig. 1A-D)

Immunohistochemistry was performed on the paraffin blocks by polymer technique (Dako REAL Envision detection system, Glostrup, Denmark), including peroxidase/39-39-diaminobenzidine trichloracetic acid, including various antibody markers (Table 1). Immunohistochemistry revealed diffuse positivity for vimentin and S100 protein and negativity for desmin, smooth muscle actin (SMA), H-caldesmon, HMB45, synaptophysin, chromogranin, melan A, myogenin, inhibin, glial fibrillary acid protein (GFAP), neuron-specific enolase, CD99/MIC2, and FLI-1. CD10 highlighted the endometrial stroma and was negative in tumor cells. Desmin and SMA highlighted the myometrial tissue. Cytokeratin (CK) and epithelial membrane antigen (EMA) highlighted the entrapped glands and were negative in the tumor cells. (Fig. 2A-D).

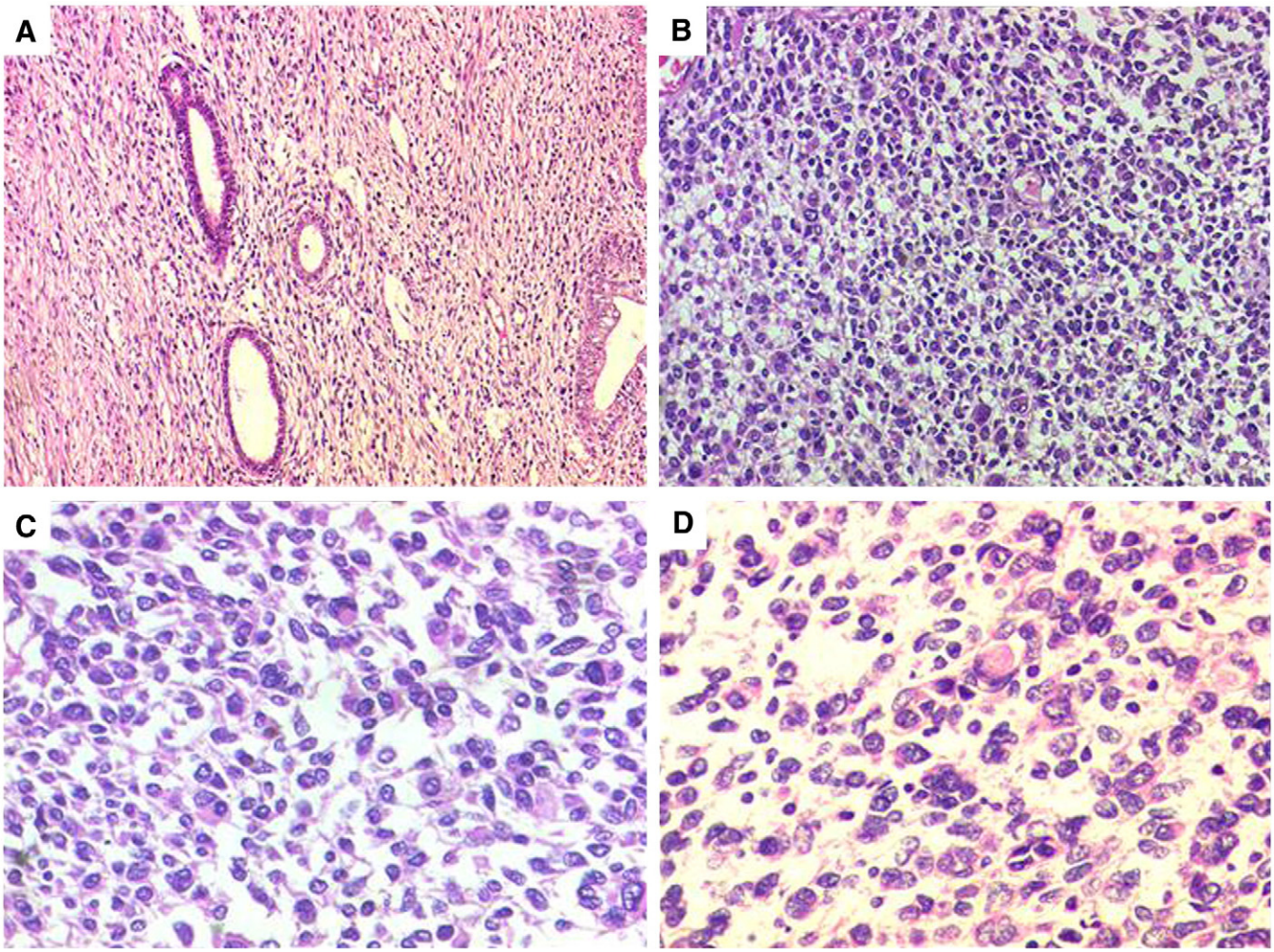

Fig. 1. Histopathologic features of a uterine epithelioid MPNST. (A) Cellular tumor composed of polygonal to short spindly cells with interspersed benign endometrial glands. H & E ×40. (B) Tumor showing predominantly epithelioid/polygonal cells. H & E ×100. (C) Epithelioid tumor cells with hyperchromatic nuclear and focal "rhabdoid" cells with abundant eosinophilic cytoplasm and eccentric nuclei. H & E ×200. (D) Higher magnification showing "rhabdoid" cells. H & E ×400.