ABDOMINOPELVIC PERIVASCULAR EPITHELIOID CELL TUMOR WITH OVERT MALIGNANCY: A CASE REPORT

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Perivascular epithelioid cell tumor (PEComa) is a group of rare tumors composed of epithelioid cells with characteristic perivascular distribution and co-expression of the melanogenic marker HMB-45 and muscular markers. There are no documented parameters referring to the biologic behavior of PEComa. We report an abdominopelvic PEComa with overt malignancy in a 16-year-old girl. Histologically, the tumor showed the typical morphophenotypic characteristics of PEComa. Though the cytologic appearance of the tumor cells was relatively bland, the extensive necrosis, presence of lymph node metastases, and surrounding tissue invasion were all indicative of malignancy. Relapse of the tumor with multiple lymphadenopathy shortly after debulking surgery for the primary lesion, and postoperative adjuvant chemotherapy, further denoted its aggressive behavior.

Key Words: perivascular epithelioid cell, PEComa, angiomyolipoma, clear cell sarcoma, clear cell renal cell carcinoma
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Perivascular epithelioid cells, first discussed by Bonetti et al [1], are found in a group of disorders including angiomyolipoma (AML), lymphangioleiomyoma (LAM), and clear-cell "sugar" tumor (CCST) of the lung. The term "PEComa" (perivascular epithelioid cell tumor) refers to tumors composed primarily of perivascular epithelioid cells [2]. More tumors are being reported and categorized as members of the PEComa family, including monotypic epithelioid AML [3–9], pulmonary or extrapulmonary CCST [10–13], and clear-cell myomelanocytic tumor (CCMMT) of the falciform ligament/ligamentum teres [14,15]. They all share common morphophenotypic features: round to polygonal (epithelioid) tumor cells, clear to variable eosinophilic granular cytoplasm, round vesicular usually centrally located nuclei, as well as immunoreactivity to both the melanogenic-related marker HMB-45 and muscular markers.

The biologic behavior of PEComa has not been well documented, perhaps due to the rarity of these disease entities and because the duration of follow-up is too short. In general, most AMLs [4,6,16], CCSTs of the lung and pancreas [10,12,13], and CCMMTs of the falciform ligament/ligamentum teres [14,15] are thought to follow a benign course. However, malignant AML [16], malignant pulmonary CCST [10], and malignant PEComas of the prostate and uterus [17,18] have also been reported, as well as four cases of abdominopelvic sarcoma with perivascular epithelioid cells reported by Bonetti et al in 2001 [18]. We report a case of abdominopelvic PEComa in a 16-year-old girl with overt malignancy shown by extensive necrosis, surrounding tissue invasion, and lymph node metastases at diagnosis.

CASE PRESENTATION

A 16-year-old girl was referred to the gyneco-obstetric outpatient department of our hospital on January 7, 2003, due to left lower quadrant abdominal pain and a huge intra-abdominal mass with bilateral hydronephrosis found by

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abdominal ultrasonography at a regional hospital. Physical examination revealed a distended abdomen with a palpable mass. Hematologic examination revealed normocytic anemia (red blood cells, 3.35×10^9/L; hemoglobin, 93 g/L; hematocrit, 0.31; mean corpuscular volume, 92.2 fL), while biochemistry parameters were all within normal limits. Magnetic resonance imaging (MRI) of the abdomen revealed a lobulated, relatively solitary tumor mass occupying the lower abdomen mainly in the pelvic area, with bilateral compressive obstructive uropathy over the distal lower ureter and a small amount of ascites. No remarkable fat component was discernable. Serum levels of human chorionic gonadotropin β subunit, α-fetoprotein, carcinoembryonic antigen, CA19-9, tissue polypeptide-specific antigen, and squamous cell carcinoma antigen were all within normal ranges, but the serum lactate dehydrogenase level was 1,645 U/L (normal range, 91–180 U/L) and the CA125 level was 141,300 U/L (normal range, < 35,000 U/L). Laparotomy showed a huge, encapsulated, hypervasculated tumor in the pelvis with invasion to the rectosigmoid colon and mesocolon and adhesion to the uterus. The tumor was partially covered by the mesocolon. Bilateral ovaries were grossly normal. Several enlarged lymph nodes over the mesocolon, iliac region, and pelvic floor were also found. Intraoperative frozen-section analysis of the tumor revealed a nested arrangement of bland-looking cells with clear-to-pale eosinophilic granular cytoplasm segregated by a delicate vascular network, mimicking the architecture of clear-cell renal cell carcinoma. In addition to excision of the tumor, resection of the sigmoid colon with Hartmann's procedure, wedge resection of bilateral ovaries, and pelvic floor lymph node dissection were performed.

On pathologic examination, the tumor mass measured 27 × 22 × 9 cm and weighed 1,500 g. Grossly, it was partially encapsulated by thin membranous tissue. The cut surfaces were grayish-white with areas of fresh and old hemorrhage and necrosis. The tumor had adhered to the serosa of the large bowel, but the mucosa and muscle wall of the colon were not involved.

Histologically, the tumor was composed of cells with abundant clear-to-fine eosinophilic granular cytoplasm and round, rather uniform, nuclei arranged in nests or wide fascicles with delicate vascular septae (Figures 1 and 2A). Mitoses were inconspicuous. Proliferation of thin-walled capillary-like vessels with occasional glomerulus-like vascular tuft formation was also noted in foci. There was extensive coagulative necrosis and hemorrhage. No mature adipose tissue, spindle-shaped smooth-muscle bundles, or abnormal thick-walled blood vessels characteristic of classic AML were found. The tumor extended to the colonic serosa without penetrating to the muscular bowel wall. Surprisingly, despite the bland cytologic appearance of the tumor cells, lymph node metastases were found. The bilateral ovaries were unremarkable. Immunohistochemically, the tumor cells were negative for cytokeratin (CK), epithelial membrane antigen (EMA), vimentin, and chromogranin-A, but strongly expressed the melanogenic marker HMB-45 (Figure 2B) and focally positively stained with smooth muscle actin (SMA) (Figure 2C). Intracytoplasmic or

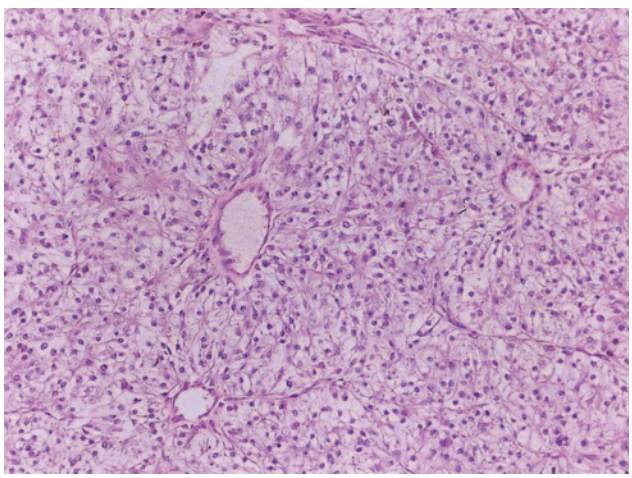

Figure 1. *Epithelioid tumor cells are arranged around delicate vasculature (perivascular arrangement; hematoxylin & eosin, original magnification × 100).*

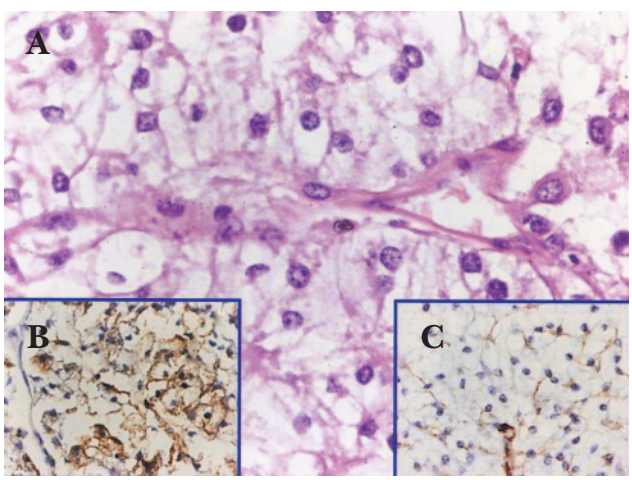

Figure 2. *(A) Perivascular epithelioid cells are round to polygonal with clear to finely eosinophilic granular cytoplasm and usually centrally located small nuclei. Note the intimately perivascular arrangement of the cells (hematoxylin & eosin, original magnification × 400). (B) Tumor cells show diffusely strong positive staining with HMB-45 (immunoperoxidase, original magnification × 400). (C) Some cells also stain positively with smooth muscle actin (immunoperoxidase, original magnification × 400).*