Pleomorphic Rhabdomyosarcoma with Neuroendocrine Differentiation in Abdominal Wall

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Abstract

The present study presents a case of primary pleomorphic rhabdomyosarcoma that occurs in the abdominal wall. A 52-year-old patient arrived in our department. In his clinical history they have been: 2016 advanced serous ovary papillary carcinoma; a diagnosis of a primary gynecological tumor with secondary extension, and the patient was prepared for bilateral total abdominal hysterectomy salpingo-oophorectomy, omentectomy. Macroscopic and histopathological evaluation of the specimen removed surgically showed a medially differentiated serous papillary carcinoma. The patient received Carbo-Taxol and avastin as postoperative chemotherapy. Postoperative follow-up and CT twelve months after surgery, revealed signs of tumor recurrence: two multilobate neoplasms of 15 cm and 5 cm in size respectively with intestinal obstruction. The final histological diagnosis was pleomorphic rhabdomyosarcoma with neuroendocrine differentiation.

Keywords: Ovary cancer; Pleomorphic rhabdomyosarcoma; Immunohistochemistry

Introduction

Rhabdomyosarcomas (RMS) more commonly afflict children and adolescents. It is rare in adults, accounting for 1% of all soft tissue sarcomas. In adults, rhabdomyosarcomas are embryonal (34%), alveolar (23%) or pleomorphic (43%), rarely spindle cell or sclerosing. Adult-type excludes embryonal and alveolar types. Most so-called abdominosarcomas in adults within the internal trunk are in fact dedifferentiated liposarcomas with heterologous rhabdomyoblastic differentiation. Clinical features: true adult rhabdomyosarcomas occur predominantly in the lower limb, trunk wall or upper limb. RMS has been divided into 3 main subtypes: Embryonal, alveolar and pleomorphic RMS (PRMS). The most common subtypes are the embryonal and alveolar subtypes. Primary PRMS is relatively rare and primarily affects adults, with a peak incidence in the fifth decade of life. It most commonly arises in the deep soft tissues of the extremities. Sarcomas showing neuroendocrine/neuronal differentiation are uncommon: apart from tumors such as peripheral primitive neuroectodermal tumor (pPNET) [1-5] and malignant gastrointestinal stromal/autonomic nerve tumor (plexosarcoma) [6], examples include extra skeletal myxoid chondrosarcoma [7-9] malignant peripheral nerve sheath tumor and ectomesenchymoma [10-12]. This report provides the detailed clinicopathologic findings of 1 case of pleomorphic rhabdomyosarcoma (PRMS) showing neuroendocrine/neuronal differentiation based on classical techniques of histology and immunostaining.

Case Report

In September 2017 a 52-year-old female patient presented to our department. In her clinical history were: 2016 advanced ovary papillary serous carcinoma; a diagnosis of a primary gynecological tumor with secondary extension, and the patient was prepared for total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy. The resected tumor tissue was fixed in 10% formalin, embedded in paraffin and cut into 5 μm sections using a microtome. The sections were subsequently stained with haematoxylin and eosin and visualized under a microscope. The tumor displayed cellular admixtures of pleomorphic spindle cells and polygonal, rhabdomyoblastic cells arranged in poorly defined clusters. The spindle cells were configured in vague fascicles and formed the background proliferation in which variable numbers of polygonal cells were distributed. The ratio of spindle to polygonal cells was at least 10:1, but the latter cells were easily identifiable in almost every high-power microscopic field. The polygonal cells displayed abundant, brightly eosinophilic cytoplasm and eccentrically located, pleomorphic nuclei. Both cellular populations displayed marked cytologic atypia, with marked anisonucleosis, abnormalities of chromatin, and nuclear membrane irregularities. Osteoclast-like giant cells (Figure 1) and numerous mitotic figures were present, including highly atypical forms (average mitotic index: 30–40 mitotic figures per 10 high power fields). The cells revealed scanty to moderate amounts of pale eosinophilic cytoplasm with indistinct borders. The nuclei were vesicular, moderately large, round to by Oliveira et al. [7] oval with clumped or coarse chromatin and contained one or more prominent nucleoli. Microscopic analysis showed full-thickness infiltration of the colon and small intestine wall and diffuse infiltration of the peritoneum.

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Results

Immunohistochemical reactions for cytokeratins (AE1/3, CK7, CK20, CAM5.2), WT-1, CA125, MART-1, S100 all found negative. In contrast, vimentin, desmin, actin, myogenin (Figure 2), reacted with tumor cells that show a diffuse neuroendocrine differentiation: synaptophysin, NSE and chromogranin (Figure 3) positive. The tumor was negative for estrogen and progesterone. The proliferation index was very high, revealing 30-40 mitoses for 10 high power fields, that was demonstrated by immunostaining with anti-Ki-67 antibodies. The final diagnosis was pleomorphic rhabdomyosarcoma with neuroendocrine differentiation. The patient died about 90 days after the first hospitalization due to organ failure. For postoperative chemotherapy he received only avastin, non-reactive and with rapidity progression to death approximately 2 months after surgery.

Discussion

RMS is divided into 3 main subtypes: Embryonal and alveolar RMS and PRMS, according to the 2002 World Health Organization Classification of Soft Tissue and Bone Neoplasms [14-21]. PRMS was first described by Stout in 1946. Primary PRMS is relatively rare and primarily affects adults in the fifth decade of life and are mostly located in the lower extremity. Less frequent sites of presentation are the abdomen, retroperitoneum, chest wall, spermatic cord/charges. These larger rhabdomyoblasts are often arranged in clusters, sheets, or scattered individual cells. Atypical, vesicular nuclei with prominent nucleoli predominate. The rhabdomyoblasts in the background that surround the large, pleomorphic rhabdomyoblasts vary from round to spindled. Neuroendocrine/neuronal differentiation was not [7] demonstrated completely in rhabdomyosarcoma. This study wants to highlight [7] the neuroendocrine/neuronal differentiation in rhabdomyosarcoma, and specifically the pleomorphic variant [24-26].

Conclusion

The positive immunoreactivity for myogenic markers supported rhabdomyoblastic differentiation. According to all classical criteria, therefore, the tumor conform to PRMS. In the definition of neuroendocrine differentiation in tumors, in addition to the histologic features, chromogranin and synaptophysin immunostaining forms the most practical and widely used criterion. Chromogranin and synaptophysin are widely regarded as reliable and specific. In rhabdomyosarcoma, neurone specific enolase (NSE), and CD56 have been demonstrated; these markers were originally thought to be specific for neuroendocrine differentiation, but their specificity has been brought into question by their demonstration in a wide variety of non neuroendocrine cells. These papers show carcinomas with neuroendocrine and rhabdomyosarcomatous differentiation. To our knowledge this is a rare case report of abdomen wall pleomorphic rhabdomyosarcoma. The presentation of a rare adult sarcoma with neuroendocrine differentiation mimicking a gynecologic malignancy was an unusual feature that complicated the diagnosis in this case.

References


