Case Report:

Malignant small round cell tumor masquerading as mesothelioma of peritoneum: diagnosed on immunohistochemistry

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Abstract:
Peripheral primitive neuroectodermal tumor (pPNET) arises in soft tissue and is thought to be of neural crest origin. PNET has been found in several sites: chest (44%), retroperitoneum and pelvis (26%), extremities (20%), head and neck (6%), and others (4%). Peripheral primitive neuroectodermal tumor is aggressive and rare, comprising 1% of soft tissue sarcomas. A 30-year-old woman had an omental cake. Histopathological examination revealed small round cell tumor confirmed as pPNET. Chemotherapy yielded complete response.

Key words: Peripheral primitive neuroectodermal tumor; Peritoneum; Chemotherapy

Introduction
Malignant small round cell tumors (MSRCT) is a term used for tumors composed of malignant round cells that are slightly larger or double the size of red blood cells in air-dried smears. This group of neoplasms is characterized by small, round, relatively undifferentiated cells. They generally include Ewing's sarcoma (EWS), peripheral neuroectodermal tumor, rhabdomyosarcoma, synovial sarcoma, non-Hodgkin's lymphoma, retinoblastoma, neuroblastoma, hepatoblastoma, and nephroblastoma. Other differential diagnoses of small round cell tumors include small cell osteogenic sarcoma, undifferentiated hepatoblastoma, granulocytic sarcoma, and intra-abdominal desmoplastic small round cell tumor. Differential diagnosis of small round cell tumors is particularly difficult due to their undifferentiated or primitive character. EWS/PNET is characterized by a balanced chromosomal translocation which generates a fusion transcript of the EWS gene and the Friend leukemia virus integration 1 (FLI-1). Cases of primary EWS/pPNET of the kidney, retroperitoneum, and omentum have been sporadically reported in the literature. To our knowledge, only two or three cases reported of EWS/pPNET observed in retroperitoneum/omentum. The tumor was unique with its location.

Diagnosis was confirmed on Immunohistochemistry (IHC), CD99, the product of the MIC2 gene, is a cell surface glycoprotein that is expressed in more than 95% of Ewing sarcoma/primitive neuroectodermal tumors with a diffuse membranous staining pattern.

Case report
A 30 years-old woman presented with distention of abdomen, nausea, vomiting and shortness of breath. Patient also complains of pain in epigastrium and bilateral hypochondriac region since two months. She was treated for gastritis but condition did not improve. Ultrasound abdomen showed thick fluid in
pelvis and CT abdomen revealed multinodular enhancing soft tissue mass involving omentum and peritoneum from subdiaphragmatic region to pelvis [Figure 1a,b]. Gut loops were centrally located, multiple mesenteric lymph nodes were seen along with free fluid. Radiologically, diagnosis of peritoneal mesothelioma was made. Upper gastrointestinal endoscopy and colonoscopy was normal. CEA = 0.6, CA125= 79.7, CA 19.9= 2.7, rest of the laboratory investigations were normal. Laprotomy with biopsy from mass and decompensation of ascetic fluid was done. Patient was diagnosed as a case of pseudomyxoma peritoni and debulking surgery was performed. Specimen was sent to department of Pathology. Macroscopic examination revealed a grey 12 cm, round, soft tissue mass with “fish flesh” consistency, partial necrosis and hemorrhagic foci [Figure 2]. H &E section showed the tumor was composed of small round cells with a finely distributed chromatin pattern. Rosette formations or ribbon-like cell arrangements were not apparent. Section from other areas demonstrated “storiform-like” pattern consisting of plump spindle cells arranged in short fascicles around slit-like vessels and contained areas showing more cellular pleomorphism [Fig 3,4]. Features were in favour of biphasic mesothelioma, but on immunohistochemical analysis spindled cells with storiform showed diffuse and intense membranous MIC-2 (CD99) positivity and negativity for calretinin and cytokeratin [Fig 5,6].

**Discussion**

Small round cell tumors comprise heterogeneous neoplasms composed of relatively small, round to oval, closey packed undifferentiated cells with high nuclear-cytolasmic ratio, scant cytoplasm, and round nuclei with evenly distributed, slightly coarse chromatin and small or inconspicuous nucleoli. In spite of a similar light microscopic morphology, SRCTs include pathologic entities from vastly different lineages, including

1. epithelial tumors, for example, small cell carcinoma (SmCC) (poorly differentiated neuroendocrine carcinoma);
2. mesenchymal tumors encompassing malignant solid neoplasms of childhood and other small round cell sarcomas; and
3. tumors with overlapping features, such as lymphoma and melanoma.

Because of similar routine light microscopic features of these tumors, immunohistochemistry is often mandated for a definitive diagnosis.  

[3] Ewing’s tumor arises from long bones and soft tissue. When Ewing’s tumor arises from soft tissues, it is called “extraskeletal Ewing’s tumor” (EES). PNET, similar to Ewing’s tumor is a round cell tumor originating from neuroectodermal crest. Histologically, EES and PNET are closely related tumors and have been grouped together as Ewing family of tumors. Patients with abdominal/retroperitoneal PNET present with nonspecific symptoms like pain and a palpable mass on examination. Diagnosis is routinely based on imaging studies; however, biopsy is essential for definitive diagnosis. The most commonly utilized biopsy techniques are either open biopsy or an imaging-guided core biopsy.  

[4] To distinguish between EWS/pPNET and other SRCTs is sometimes difficult by conventional morphologic methods, especially if the tumor arises in an unusual region. The typical PNET may resemble a primitive fibrosarcoma, malignant schwannoma, or malignant fibrous histiocytoma.  

[5] Because up to 80% of patients with apparently localized disease have occult metastatic disease, multidrug systemic chemotherapy
is indicated in patients with PNET. To our knowledge, this is a rare case to be reported of Ewing’s sarcoma/peripheral primitive neuroectodermal tumor arising in the omentum with unique pathological features and the occurrence of partial neural differentiation during the clinical course. This case pointed out to us, that only thorough examination confirms a definitive diagnosis of small round-cell tumor of the abdomen, it also shows that Ewing’s sarcoma/peripheral primitive neuroectodermal tumor should be included in the differential diagnosis of cystic lesions in the omentum.

References
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