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Case Reports

Diffuse intra-abdominal clear cell myomelanocytic tumor: report of an unusual presentation of "PEComatosis" simulating peritoneal mesothelioma

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Abstract We report a case of diffuse myomelanocytic tumor of the peritoneum that simulates, clinically and instrumentally, a malignant mesothelioma. The patient was a 70-year-old woman with a history of ancient hysterectomy for fibroids, who presented with abdominal discomfort. Exploratory laparotomy revealed diffuse encasing of the peritonealized organs by a thin, fleshy, gray-white tissue rind. Scattered tumor masses were present as well. A dominant lesion measuring 6 cm in larger size was resected from the pelvis. Histological examination revealed a tumor composed of epithelioid and spindle cells, exhibiting either a clear or slightly eosinophilic cytoplasm and a mild to moderate nuclear pleomorphism. Focal areas of necrosis could be documented. Immunohistochemically, tumor cells were positive for HMB45, melan-A, and smooth muscle actin, but negative for other antibodies, including epithelial markers, desmin, and S100 protein. We believe that this case represents an example of myomelanocytic tumor of uncertain biologic potential, a member of the recently devised perivascular epithelioid cell tumors (PEComa), with an unusual presentation simulating a diffuse mesothelial neoplasm. The origin of this particular lesion is briefly discussed in light of the recent literature published on the subject.

Keywords: Epithelioid neoplasms; Peritoneum; Immunohistochemistry; Female genital tract

1. Introduction

After their initial description in the early 1990s [1,2], tumors of perivascular epithelioid cells (PEComas) have now emerged as a group of typical microscopic entities sharing a spindled/epithelioid morphology and showing a consistent immunoreactivity to actins and the so-called melanoma antigens [3], including the pulmonary "sugar" tumor and lymphangiomyomatosis and, in the abdominal cavity, the renal angiomyolipoma and the clear cell myomelanocytic tumor (CCMMT) of the falciform ligament/ligamentum teres [4]. The latter usually arises within the female genital tract, occurs in young females, and pursues a benign course. Nevertheless, cases exhibiting a significant deviant clinical presentation as well as phenotypic features have been reported, including extra-pelvic, extra-abdominal location, and tumors occurring in aged or in male patients [3-8]. However, to the best of our knowledge, CCMMT has not presented with diffuse peritoneal involvement that simulates, clinically and macroscopically, a diffuse peritoneal neoplasm.

We report a case of CCMMT presenting with exclusive peritoneal involvement, raising clinically the suspicion of a diffuse mesothelial tumor.

2. Case report

A 70-year-old patient was admitted to the hospital because of pain and abdominal distension. About 27 years

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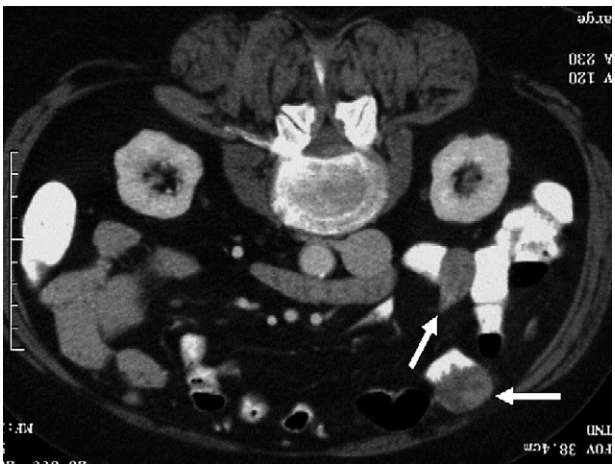

Fig. 1. Computed tomographic scan of the abdomen showing two distinct solid nodules adjacent to the small bowel (arrows). There is no radiological evidence of intestinal occlusion.

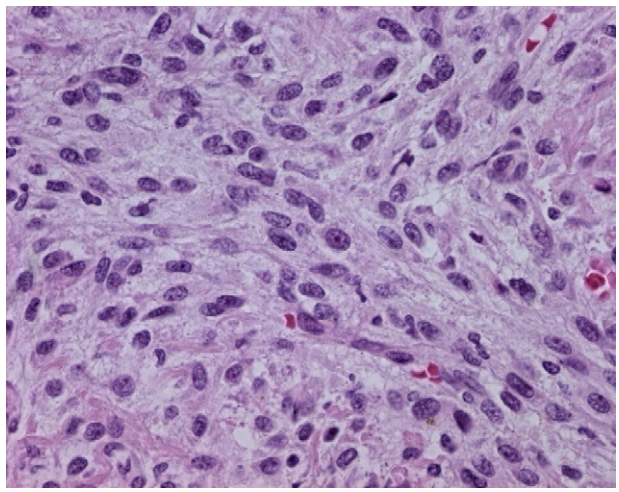

Fig. 3. A vague fascicular arrangement of tumor cells showing spindled morphology is focally present. Bland nuclear chromatin texture can be noted (H&E, intermediate power).

previously, she had undergone total hysterectomy with bilateral oophorectomy for "uterine leiomyomata." Glass slides pertaining to the uterine specimen were no longer accessible. Computed tomographic scans revealed confluent nodules in the pelvis, omentum, and visceral peritoneum (Fig. 1). A dominant mass measuring 6 cm was documented between the lower rectum and urinary bladder. The results of the remainder of the physical examination were negative. A chest x-ray film did not reveal abnormality of the lung and mediastinal profile. Exploratory laparotomy revealed diffuse encasing of the peritoneum and omentum by finely scattered nodules alternating with thin, gray-white tumor rinds showing hemorrhagic variegations on the cut surface. There was no evidence of peritoneal effusion. Histological evaluation of H&E-stained sections showed a diffuse proliferation of atypical spindle to polygonal cells, showing a round to oval nucleus. The cell cytoplasm was variably abundant, clear to granular (Figs. 2 and 3). Areas of moderate cell pleomorphism could be focally identified (Figs. 4 and 5). A panel of antibody was applied on paraffin tissue sections using the avidin-biotin method and commercially available antibodies. Optimal dilutions were assessed by means of a titration checkerboard as described elsewhere [9]. Tumor cells were positive for HMB45 (Fig. 6), melan-A (Fig. 7), smooth muscle actin (Fig. 8), and vimentin and negative for keratins, S100 protein, chromogranin, synaptophysin, α-inhibin, desmin, actins, H-caldesmon, c-kit, CD31, CD34, LCA, CD99, ER, and PR. A low proliferative fraction was detected by means of the MIB-1 antibody (Fig. 9). No further therapy was administered. The patient general condition remained stable for 1 year after abdominal surgery. Repeated computed tomographic scans of

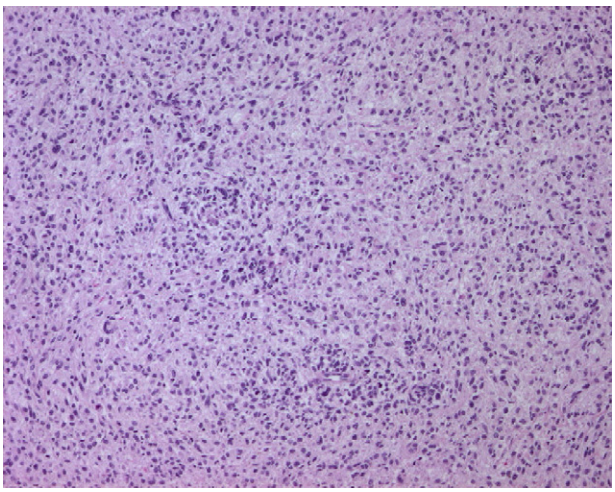

Fig. 2. A panoramic view of a PEComa tumor shows a diffuse proliferation of uniform cells (H&E, low power).

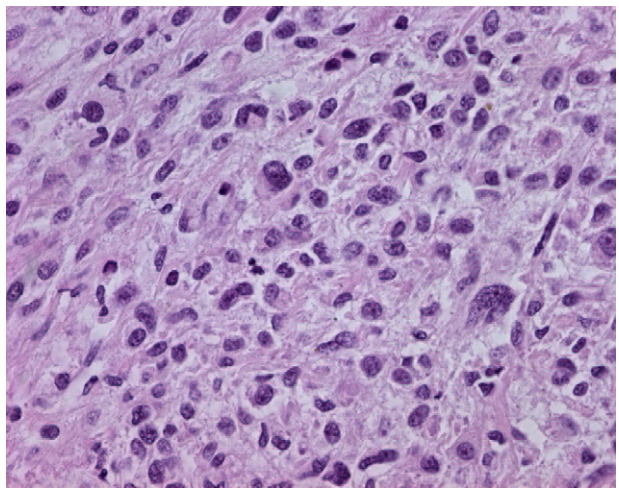

Fig. 4. Mild to moderate nuclear pleomorphism of tumor cells along to some cytoplasm granularity and slight eosinophilia (H&E, intermediate power).