CASE REPORT

Ileus secondary to a retroperitoneal malignant melanoma

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Abstract  Retroperitoneal malignant melanomas, either primary or metastatic, are rare. We present our clinical experience concerning one case with ileus secondary to a huge retroperitoneal malignant melanoma. A 77-year-old woman was admitted to the hospital due to progressive abdominal fullness and decreased appetite for 4 months. Plain films showed soft-tissue density in the left lower quadrant of the abdomen, as well as rightward displacement of the intestine. Laboratory data excluded any metabolic or septic causes of ileus. Abdominal computed tomography scan identified a huge retroperitoneal tumor with invasion of the left lower peritoneal space. The mass measured approximately 18.2 × 21.5 cm in the largest section. Immunohistochemical analysis of the tumor biopsies at minilaparotomy showed positive staining of tumor cells for S-100 protein, human melanoma black-45, and vimentin. Thus, a diagnosis of malignant melanoma with peritoneal metastases was established. This case highlights the possibility of a retroperitoneal malignant melanoma exerting a mass effect on the surrounding organs. The authors suggest that malignant melanoma should be taken into consideration as a possible differential diagnosis of retroperitoneal neoplasms.

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Introduction

Melanomas are derived from melanocytes, which originate from the neural ectoderm and migrate to the skin, eyes, and leptomeninges in early embryonal life. Compared with most malignant melanoma that occurs primarily in the skin, fewer than 10% of malignant melanomas arise from noncutaneous regions, including the choroid layer of the eyes, the leptomeninges, oral mucosa, and the mucosa of the gastrointestinal, respiratory, and genitourinary tracts. Malignant melanoma arising in the retroperitoneum is extremely rare. Here, we report a case with ileus secondary to a huge retroperitoneal malignant melanoma and briefly review the literature.

Case report

A 77-year-old woman visited our department of emergency due to progressive abdominal fullness and decreased appetite over the previous 4 months. In addition, she complained of nausea and vomiting in the previous 3 days. She had underlying diabetes mellitus as well as ischemic heart disease, and lived in a nursing home. General physical examination revealed abdominal distension and hypoactive bowel sound, but no abdominal tenderness. Plain films showed soft-tissue density in the left lower quadrant of the abdomen, as well as rightward displacement of the intestine. Laboratory data excluded any metabolic or septic causes of ileus. Abdominal computed tomography (CT) scan identified a huge retroperitoneal tumor with irregular margin, heterogenous density, and invasion of the left lower peritoneal space anteroinferiorly. The mass measured approximately 18.2 × 21.5 cm in the largest section (Fig. 1A) and was clearly separable from the pancreas, kidney, and bilateral adrenal glands. The tumor extended cross-midline of the abdomen, partially enclosing the left common iliac artery (Fig. 1B), and was inseparable from the left psoas muscle. The patient underwent minilaparotomy thereafter. We noted that the gross appearance of the mass was soft and gray-white in color (Fig. 2A). The tumor had a parenchymal green-black necrotic area as well as a bleeding tendency. Multiple metastatic lesions were noted over the visceral and parietal peritoneum (Fig. 2B). We checked the small intestine carefully during the operation and no obvious intestinal mass lesions were identified. In consideration of the patient’s intraoperative critical hemodynamics and late stage of the malignancy, we performed incisional biopsy instead of en bloc resection of the tumor as well as the peritoneal lesions. The specimen was evaluated by histologic analysis and immunohistochemical staining. Microscopic study revealed abundant tumor necrosis, high nuclear/cytoplasmic ratio, and hypercellularity; with spindle or irregular nucleoli in tumor cells (Fig. 3A). The cells showed positive staining for S-100 protein (Fig. 3B), human melanoma black-45 (HMB-45; Fig. 3C) and vimentin (Fig. 3D), whereas cytokeratin AE1/AE3, cytokeratin 7, epithelial membrane antigen, and CD34 were negative. Therefore, the diagnosis of malignant melanoma with peritoneal metastases was established. The detailed history revealed that the patient had no previous melanoma. Subsequently, she was referred for further dermatologic and ophthalmologic examination, and no suspicious primary lesion was identified. Further investigations were also scheduled, but the patient declined to receive subsequent examinations. Due to poor nutritional status on admission, she received postoperative nutritional support therapy prior to further treatment of the Stage IV malignant melanoma. However, systemic chemotherapy was postponed because the patient developed pneumonia in the postoperative course. Consequently, she died 2 months after surgery.

Discussion

Primary retroperitoneal neoplasm includes a diverse group of benign and malignant tumors that arise in the retroperitoneum. Due to the inaccessibility of the region and because these tumors are often asymptomatic until they have reached a substantial size, they are usually large at presentation. Therefore, diagnosis and management of
these tumors is often challenging for clinicians because of their clinical features.

The most common symptoms in patients with retroperitoneal tumors remain nonspecific, including early satiety, anemia, abdominal swelling, abdominal pain, and back pain. Additionally, patients may also present with a palpable abdominal mass. The most common histology of primary retroperitoneal malignant tumors is liposarcoma, followed by leiomyosarcoma. By contrast, malignant melanoma arising in the retroperitoneum is extremely rare.

In this case report, we present a case with ileus secondary to a huge retroperitoneal malignant melanoma. A review of the literature was performed using PubMed with the keywords “malignant melanoma” and “retroperitoneum” to search for all case reports and reviews published between 1963 and 2013 on malignant melanoma originating from the retroperitoneal space. Similar searches were performed using Ovid MEDLINE. A solitary retroperitoneal malignant melanoma with a maximal diameter >20 cm at diagnosis has not been previously reported in the literature. Most cases reported so far with primary retroperitoneal malignant melanoma are of the adrenal medulla origin. There are only seven cases reported to date as a primary retroperitoneal malignant melanoma that originated from retroperitoneal tissue other than the adrenal gland. Because of their rarity, the epidemiology and clinical behavior of these tumors have not been well characterized.

Regarding the possible etiology of primary retroperitoneal malignant melanoma, several authors have pointed out that melanoma lesions may arise from the adrenal glands, lumbar sympathetic chains, and autonomic nerve plexuses. The findings may explain why melanoma can arise within the retroperitoneal space. Previous studies have reported that primary retroperitoneal malignant melanoma can cause a mass effect on the surrounding organ. It is interesting to note that the findings in the current case, like a solitary bulky tumor with a mass effect, are coincident with previous case reports of primary retroperitoneal melanoma. Most retroperitoneal metastatic melanomas (i.e., pancreas, kidney, lymph nodes) are usually multiple and small lesions.

Diagnosis of malignant melanoma in the abdomen is generally made by radiographic studies, histopathologic analysis, and immunohistochemical stains. Abdominal CT scan is currently the most widely used imaging method for tumor staging, preoperative surveillance, and a follow-up to therapeutic response. However, a final diagnosis of malignant melanoma can only be made after histopathologic and immunohistochemical analysis. Cytologic features of malignant melanoma include enlarged nuclei, prominent nucleoli, thick and irregular nuclear membranes, abnormal cytoplasmic melanization, and atypical mitotic figures. Histologically, tumor cells may be classified into four cell types: epithelioid, spindle cell, lymphoma-like, and pleomorphic. Immunohistochemical stains are now widely used for the diagnosis of malignant melanoma. Extracutaneous melanomas exhibit the same immunohistochemical and ultrastructural features as their cutaneous counterparts. S-100 protein, HMB-45, melanin A, vimentin, and occasionally antityrosinase antibodies are used as immunohistochemical tools in the diagnosis of melanoma. S-100 protein is the most common screening immunohistochemical stain used in the diagnosis of melanoma and it remains the most sensitive marker for melanocytic lesions (sensitivity nearly 100%). Other markers such as HMB-45, melanin A, vimentin, and antityrosinase antibody demonstrate relatively good specificity but not the same sensitivity as S-100.

The sensitivity of HMB-45 for melanoma is reported to reach 93–100%, with a distribution similar to that of melanin A, and the reported sensitivity of vimentin is 93%. Along with the wide application of immunohistochemical stains, Tousignant et al. reported that simultaneous positivity for S-100 protein and vimentin, confronted with the patient’s history and with histologic features, firmly establishes the diagnosis of malignant melanoma in virtually all cases.

In conclusion, we report a case with ileus secondary to a huge primary retroperitoneal malignant melanoma. This case highlights the possibility of a retroperitoneal malignant melanoma exerting a mass effect on the surrounding organs. Finally, the authors suggest that malignant melanoma should be taken into consideration as a primary retroperitoneal malignant melanoma that originated from retroperitoneal tissue other than the adrenal gland.
possible differential diagnosis of retroperitoneal neoplasm.

References


Figure 3  Histologic and immunohistochemical analysis. (A) Hematoxylin and eosin stain (×400) showed high nuclear/cytoplasmic ratio and hypercellularity, with spindle or irregular nucleoli in tumor cells. (B–D) Immunohistochemical analysis demonstrated positive staining with S-100 protein (B, ×400, arrow), HMB-45 (C, ×400, arrow), and vimentin (D, ×400, arrow).