Case Report

Retroperitoneal Malignant Peripheral Nerve Sheath Tumor

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Abstract

Primary retroperitoneal neoplasms are a rare entity and account for 0.1%–0.2% of all malignancies. In addition, only 1% of malignant peripheral nerve sheath tumors (MPNSTs) occur in retroperitoneal region. Herein, we report a case of retroperitoneal peripheral nerve sheath tumor in a 50-year-old man who presented with right lower limb pain. On physical examination, an abdominal mass was palpable. Computed tomography of abdomen with contrast, showed a large, lobulated mass, in the right paravertebral region extending to the middle area of abdomen and pelvis. The patient underwent exploratory laparotomy and mass excision. Histopathological examination was consistent with "MPNSTs." This case report emphasizes that, although rare, MPNSTs may arise from retroperitoneal region. They have varied clinical presentations, and combination of microscopic, radiological, and immunohistochemical analysis is needed for diagnosis of MPNSTs.

Keywords: *Immunohistochemistry, laparotomy, malignant peripheral nerve sheath tumor, retroperitoneal, sarcoma*

Introduction

Primary retroperitoneal neoplasms are rare lesion accounting for 0.1%–0.2% of all malignancies.[1] Malignant peripheral nerve sheath tumors (MPNSTs) are any malignant tumors arising from peripheral neurons or their sheaths.[2] The incidence rate of MPNSTs is estimated to be 1/1,000,000.[3] In addition, only 1% of MPNSTs occurs in retroperitoneal region.[4] Herein, we report a case of MPNST arising in the retroperitoneum region in a 50-year-old man. To the best of our knowledge, the present case is a unique condition regarding the weight of tumor (5.0 kg).

Case Report

A 50-year-old male patient was referred to our outpatient clinic for the right lower limb pain. There was no history of nausea, vomiting, jaundice, hematemesis, or melena. Urinary disturbance, weight loss, or anorexia was not noted. The patient did not have any family history of neurofibromatosis (NF). Other clinical manifestations including muscular atrophy, weakness, and functional limitation were not existed. On examination, a mass was palpated in the right lower quadrant of abdomen. Physical examination did not show any significant findings including cafe au lait macules in skin or lisch nodules in eyes. Routine laboratory tests all were all normal. Ultrasonography of abdomen showed two hypoechoic masses, 5 and 10 cm in diameter in the right lower quadrant. This finding was suggestive of (1) carcinoma of cecum, (2) lymphadenopathy, or (3) retroperitoneal sarcoma. Colonoscopy was normal. Computed tomography (CT) of abdomen with contrast, showed a large and lobulated mass, 120 mm × 120 mm in size in the right paravertebral region extending to the middle area of abdomen and pelvis [Figure 1]. Informed consent for surgery was taken from the patient. He underwent exploratory laparotomy. Under general anesthesia, the mass was resected.

Macroscopically, it consisted of a fragmented soft-tissue mass weighing 5.0 kg and measuring 30 cm in greatest dimension [Figure 2a]. Cut surfaces were lobulated tan and solid. Microscopic examination showed a neoformated tissue consisting of proliferation of spindle cells. Some cells had irregular and pleomorphic nuclei. Tumor cells had a patternless growth pattern, collagen-rich network [Figure 2b].

The primary diagnosis was spindle cell tumor that was suggestive of solitary

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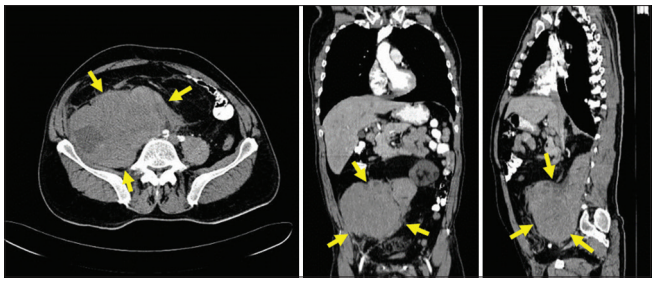

Figure 1: Abdominal computed tomography (axial, coronal and sagittal views) shows a large, lobulated mass in the right paravertebral region extending to the middle area of abdomen and pelvis

fibrous tumor (regarding to nuclear pleomorphism and the presence of necrosis in CT scan, most probably malignant). Immunohistochemical (IHC) markers including cytokeratin, EMA, desmin, S100, GFAP, CD-117, and CD-68 were all negative except for Vimentin and CD-34 which were strongly positive.

Finally, histopathological findings were consistent with "MPNSTs, low grade." The patient has been followed up regularly and has been recurrence free for 6 months.

Discussion

Retroperitoneal neoplasms often arise from mesodermal system which form over 80% of primary retroperitoneal sarcomas. The remaining primary retroperitoneal masses usually arise from nervous system.[3] MPNSTs are defined as any tumor arising from peripheral nerve sheath cells. The incidence of MPNST is 1/000000 and from 5% to 10% of all soft-tissue tumors.[5]

MPNST can occur either sporadically or in association with (NF type 1 or von Recklinghausen disease). Approximately half of the cases with MPNST are observed in patients with hereditary syndrome NF1. The factors affecting the development of MPNSTs include NF1 and radiation exposure. The median age among patients with NF1-associated MPNST is less than of patients with sporadic MPNST.[6] To the best of our knowledge, only a few cases of retroperitoneal MPNSTs have been reported in the literature.[7] Only 1% of MPNSTs occur in retroperitoneal region. In the retroperitoneum region, MPNST may arise from the vertebral nerve roots or the sacral or lumbar plexus.[8]

These tumors are often clinically silent. They may present with pelvic or abdominal mass. In our case, a mass was palpable in the right lower quadrant of the abdomen. Paraspinal region is the most common site of MPNSTs arising from retroperitoneum.[1] In the present case, the mass was in the right paravertebral region extending to the middle area of abdomen and pelvis.

CT, magnetic resonance imaging and positron emission tomography scan have been used for diagnosis of MPNSTs as well as differentiation from benign lesions.[7]

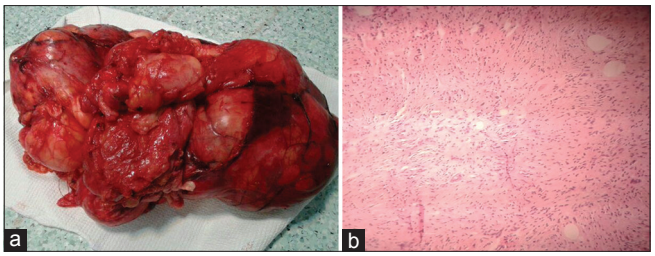

Figure 2: (a) Resected specimen weighing 5.0 kg and measuring 30 cm in greatest dimension, (b) microscopic examination shows a neoformated tissue consisting of proliferation of spindle cells

There is no specific criteria for the diagnosis of MPNST. Correlation of clinical manifestations with the exclusion of mimicking neoplasms by IHC is required for achieving the diagnosis of MPNST. Microscopically, MPNSTs usually show a varied range of cell morphologies including spindle, epithelioid, pleomorphic, or small round cell. There is no diagnostic immunoprofile for MPNST. However, S-100 staining is positive in 50%–60% of MPNST nuclei. CD34 is also expressed in about a quarter of tumors.[9] In our case, Vimentin and CD34 were positive.

A combination of macroscopic, microscopic examination beside IHC study is generally used to diagnose a case of MPNST.[10]

Unfortunately, MPNST has a poor prognosis, with survival rates between 35% and 50%.[11] The factors affecting the survival of cases with MPNST include tumor location, size, resection margin, and underlying disease such as the presence of the NF-1.[12]

Kuznetsov *et al*. reported a schwannoma of 2.89 kg.[13] Our patient had a retroperitoneal mass weighing approximately 5 kg.

There are several dilemmas in the management of retroperitoneal MPNSTs including (1) The difficulty of distinguish from benign lesions such as neurofibromas; (2) differentiation of low-grade MPNST from atypical neurofibroma; and (3) difficult surgical resection. Hence, the main treatment of MPNSTs is surgical resection.[8] Our patient underwent laparotomy and mass excision.

We introduced a case of MPNST in a 50-year-old man who presented with lower limb pain. To the best of our knowledge, this case is one of the giant cases of retroperitoneal MPNSTs weighing about 5.0 kg. A combination of clinical presentations, histopathological examination, and IHC analysis is required for the diagnosis of MPNSTs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The

patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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