Would a Massive Intra-abdominal Malignant Peripheral Nerve Sheath Tumor with Growth into the Inguinal Canal and Scrotum Preclude Surgical Option? A Case Report and Review of Literature

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Received: 26 April 2009 / Accepted: 2 May 2009 / Published online: 31 July 2013
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Abstract Malignant peripheral nerve sheath tumors (MPNST) are rare spindle-cell sarcomas derived from Schwann cells or pluripotent cells of the neural crest accounting for less than 10 % of all soft tissue sarcomas. They arise from major or minor peripheral nerve fibers or their sheaths. The World Health Organization coined the term MPNST for tumors of neurogenic origin with similar biological behavior replacing all the previous heterogeneous and, often, confusing nomenclature including malignant schwannoma, malignant neurilemmoma, and neurofibrosarcoma. The retroperitoneum and the lower extremities are the most common sites, but MPNST may arise anywhere in the body. Its location in the retroperitoneum in a patient without neurofibromatosis is an exceedingly rare occurrence. Imaging is routinely performed to assess the extent of the disease and to plan surgical resection. Surgical resection is the first line of therapy, ideally with total removal of the tumor. Owing to a high risk of recurrence with incomplete resection, postoperative irradiation and chemotherapy are necessary; however, they are often used as adjuvant therapy even if the tumor is completely resected.

Keywords Malignant peripheral nerve sheath tumor · MPNST · Retroperitoneal sarcomas

Introduction

Malignant peripheral nerve sheath tumors (MPNST) are rare spindle-cell sarcomas derived from Schwann cells or pluripotent cells of the neural crest [1]. They account for less than 10 % of all soft tissue sarcomas, and out of this, up to 60 % occurs in the setting of neurofibromatosis I [1]. The retroperitoneum and the lower extremities are the most common sites, but MPNST may arise anywhere in the body [2]. Its location in the retroperitoneum in a patient without neurofibromatosis is an exceedingly rare occurrence [2]. Moreover, the herniation of such a tumor into the inguinal canal and the scrotum is even rarer. We present such a rare case in which the tumor had grown to a massive size of 10 kg and had herniated into the right side of the scrotum, and review the literature.

Case Report

A 37-year-old male presented with a 3-month history of progressively increasing abdominal distention associated with diffuse dull aching abdominal pain. He also complained of a painless swelling in the right groin and the scrotum of 1-month duration, with steady increase in size. He also had anorexia and an unquantified weight loss, without any history of altered bowel habits, jaundice, or fever.

On examination, the patient was found to be of average build and nourishment. His abdomen was grossly distended and tense with an everted umbilicus (Fig. 1). There was a lobulated firm large mass occupying almost the entire abdomen pushing the bowel loops to the left side (as detected on percussion). The mass

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Fig. 1 Preoperative photograph showing the grossly protuberant abdomen

had continued into the right scrotum as an indirect inguinal hernia. The liver and the spleen were not palpable, and there was no shifting dullness. Digital rectal examination revealed no abnormality, and the systemic examination was essentially normal.

The hemogram and biochemical parameters were within normal limits. Ultrasound examination of the abdomen revealed a large mass occupying most of the abdominal cavity with minimal ascites. Contrast-enhanced CT scan of the abdomen and pelvis revealed a lobulated soft tissue density mass lesion in the abdomen and pelvis composed of multiple well-defined lesions displacing the viscera to the left and indenting the urinary bladder (Fig. 2). The liver was normal, and there was no ascites. The impression was that the mass was of neoplastic pathology, but the site of origin was not discernable. FNAC was inconclusive, and a core needle biopsy suggested low-grade spindle-cell sarcoma.

Fig. 2 The large retroperitoneal multiloculated tumor on CECT abdomen

The patient was offered exploratory laparotomy. At surgery, a well-encapsulated mass occupying the whole of the abdomen and pushing the bowel loops to the left was found. It was adherent to the right iliopsoas fascia, right genitofemoral nerve, and partly to the right femoral nerve. The tumor was found herniating into the right inguinal canal and stuck to the epididymis (Fig. 3). The entire lump measuring 24 cm×22-cm×20 cm and weighing 10 kg was removed en masse (Fig. 4) along with the right testis and the cord structures, but the femoral nerve was spared.

The cut surface showed the tumor to be solid comprising multiple lobulated masses separated by necrotic, mucinous, and fibrofatty tissue. Microscopically, spindle cells were seen arranged in broad sheets with 10–12 mitoses per 10 HPF. The tumor cells stained positive for S-100, confirming MPNST of intermediate grade arising from the retroperitoneum.

Fig. 3 The large retroperitoneal tumor being dissected out at surgery

After an uneventful postoperative recovery, the patient was advised adjuvant radiotherapy, which he declined. However, at 2-year follow-up, there was no recurrence of the tumor.

Discussion

The World Health Organization coined the term MPNST for tumors of neurogenic origin with similar biological behavior replacing all the previous heterogeneous and, often, confusing nomenclature including malignant schwannoma, malignant neurilemmoma, and neurofibrosarcoma [3]. They arise from major or minor peripheral nerve fibers or their sheaths [3].

These tumors may arise de novo in adult patients, although 10 to 60 % of MPNST tend to have an association with multiple neurofibromatosis type I [1].

Our patient did not manifest any neurofibromatosis, and the tumor seems to have originated sui generis. The presentation was atypical as, despite the huge intra-abdominal mass, there was no element of overt intestinal obstruction of any kind nor were there compression effects. In addition, the tumor had herniated into the right side of the scrotum which is extremely rare, and moreover, there was no evidence of local invasion that is expected in such a massive tumor. Nerve sheath tumors can grow to giant sizes. Vincent et al. reported a 4-kg tumor [4]. Our case was even larger, tipping the scales at 10 kg.

Fig. 4 Complete tumor excision along with the right testis

Imaging is routinely performed to assess the extent of the disease and to plan surgical resection. However, it does not reliably determine the malignant transformation from neurofibroma to MPNST [5]. MRI is the investigation of choice as it can reveal the nerve of origin and its relationship to adjacent structures [5].

Surgical resection is the first line of therapy, ideally with total removal of the tumor [6]. Owing to a high risk of recurrence with incomplete resection, postoperative irradiation and chemotherapy are necessary [1]; however, they are often used as adjuvant therapy even if the tumor is completely resected [1]. Even with aggressive therapy, local recurrence of tumor is seen in 50 % of patients [6]. Hematogenous spread occurs most commonly to the lungs. The reported 5-year survival rate for patients with MPNST without neurofibromatosis type I is as high as 50 % [7], but drops to as low as 10 % for those with neurofibromatosis type I [1].

A combination of gross and microscopic findings along with immunohistochemical studies are commonly used to diagnose a case of MPNST [8]. One of the macroscopic criteria laid down for diagnosis is demonstration of nerve of origin at surgery [4]. However, this is often not possible in view of the fact that they arise from small and insignificant nerves [9]. Immunohistochemistry and electron microscopic studies are essential as confirmatory diagnostic methods. S-100 protein positivity and cytokeratin negativity is a supplementary tool for confirmation [4].

Conclusion

MPNST are rare tumors which are known to grow in large masses. However, a retroperitoneal MPNST of 10-kg mass

with no features of overt obstruction or any pressure symptoms along with tumor herniation into the right side of the scrotum is unique and to our knowledge has not been reported. That the patient has no recurrence at 2 years despite defaulting on the adjuvant therapy makes our case even more interesting and rare.

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