CLINCAL CASE

RECURRENCE OF MALIGNANT PERIPHERAL NERVE SHEATH TUMOR

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Abstract

Peripheral nerve sheath tumor (PNST) is known as a benign tumor called Schwannomas, developed from the Schwann cells that produce myelin and collagen, and can occur in a wide variety of locations. In some cases, it can take a malignant turn and grow as a pelvic mass known as malignant peripheral nerve sheath tumor (MPNST), malignant schwannoma, malignant neurofibroma, malignant mesenchynoma and neurogenic sarcoma. Our case report presents a female patient presenting dysuria and pelvic pain. The clinical examination revealed a palpable mass in the lower abdomen. The computer tomography (CT) scan of the abdomen and pelvic region showed a tumor mass that was in contact with the nearby organs, but not infiltrating them. After the surgery, histopathology confirmed the MPNST proliferation. A month after, the patient returned for a follow-up and presenting pelvic pain. The CT scan of the abdomen and pelvic region showed a tumoral mass compressing the nearby organs, but not infiltrating them. The patient was referred to oncology board for palliative treatment. The particularity of this case report is the short period of time recurrence of the MPNST, rather than metastases. The most important treatment for MPNSTs remains surgery, trying to achieve negative margins. However, there have been reported two cases of unresectable MPSNT with partial remission to chemo- and radiotherapy.

Keywords*: malignant peripheral nerve sheath tumor, recurrence, sarcoma*

Introduction

Malignant peripheral nerve sheath tumor (MPSNT) is known as an uncommon sarcoma, achieving 5%-10% of all soft tissue sarcomas, with an incidence of 1:100 000/year [1]. MPNSTs are also known as malignant schwannoma, malignant neurofibroma, malignant mesenchynoma and neurogenic sarcoma. MPSNT results through dysplasia of peripheral nerves cells, such as Schwann cells or perineural cells. The histogenesis of MPNST remains unclear, but there is a higher incidence in patients with NF1, 50% of MPSNTs arise from plexiform neurofibromas, 47% sporadically and RT-associated with a 10% lifetime risk [2-4]. The typically localization of MPSNT are in the extremities (40%), trunk/retroperitoneal (38%) and head and neck region (21%) [4]. MPSNT frequently metastasizes hematogenous, most often to the lung and bone [5]. MPSNT is known as being resistant to chemo- and radiotherapy.

Case presentation

A 54-year-old woman presented with dysuria, pelvic pain and abdominal discomfort. The

patient had no history of renal or bowel disease and did not accuse considerable weight loss. Physical examination showed a palpable mass in the right lower quadrant (RLQ) and left lower quadrant (LLQ).

On investigation, urea and creatinine levels were elevated. Cancer antigen (CA) 125 was 391.90 U/L (normal range, 0-35 U/L). A CT scan of the abdomen and pelvis showed two polylobate masses in the recto-vesical pouch and at the fundus of the uterus emerging from both ovaries. Right side axial length measurement 8.6/12.7 cm, left side axial length measurement 5.5/9.2 cm, caudal forming a unique mass measuring 11/15 cm and cranio-caudal length 11 cm. Although the masses were in contact with the gall bladder, lumbosacral spine, small bowel and the anterior wall of rectum (Figure 1). Both ureters and the gall bladder were compressed by the tumoral mass. Left kidney had a grade 2 and the right kidney grade 1 hydronephrosis (Figure 2). Minimal ascites was observed.

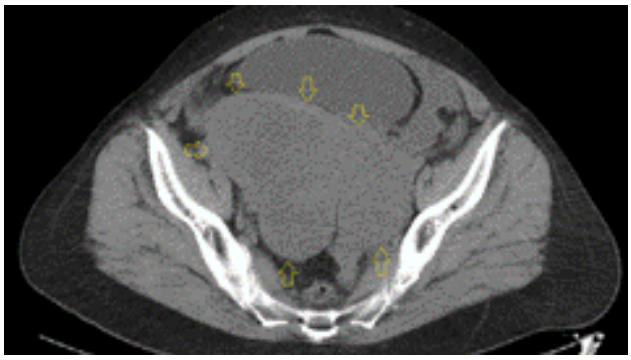

Figure 1 – Axial CT view of malignant peripheral sheath nerve tumor (lesion is indicated by yellow arrows)

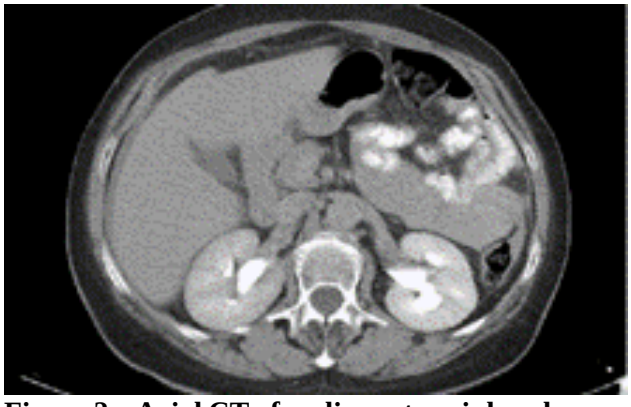

Figure 2 – Axial CT of malignant peripheral nerve sheath tumor (hydronephrosis type 2 in the left kidney)

A surgical resection of the tumor was decided, based on the clinical exam, laboratory and imagistic results. After standard preoperative preparation, the patient had undergone surgery. During surgery a pelvic tumor was seen to be adherent to both ovaries, ovaries measuring 3/2/1 cm, compressing both ureters and the gall bladder. Without direct extension and penetration into them. The surgical intervention included a resection of the tumor, a total hysterectomy with bilateral salpingo-oophorectomy and a left double J stent insertion. Postoperative recovery was uneventful.

Histopathology on macroscopic exam revealed a brown-yellow, encapsulated, irregular tumor deforming the uterus. On section, multiple compartments were filled with a cloudy brown liquid and a soft hemorrhage material. Microscopically, cells had a fusiform shape, nuclear atypia, mitotic activity and growth pattern. The surgical resection was graded as R0. Immunohistochemistry was negative for CK AE1/AE3, enolase, epithelial membrane antigen (EMA) but positive for S-100 protein. Thus, confirming to be a MPSNT proliferation.

After the MPNST confirmation further investigations were made. No stigmata of von Recklinghausen's disease nor radiation exposure were noted. Radiotherapy or chemotherapy were not recommended because they are known not to improve survival rate.

After a month the patient returned for a follow up accusing pain in the LLQ. Physical examination showed a palpable mass in the LLQ. A CT scan of the abdomen and pelvis was recommended and revealed a polylobate mass with multiple sized inner walls, axial length 9.2/11.5 cm and cranio-caudal length 10.9 cm. The mass was in contact with the left common iliac arteries, sacral spine, recto-sigmoid junction, right wall of rectal ampulla and the proximal region of sigmoid colon (Figure 3). The left ureter was compressed in the presacral and distal region leading to proximal dilatation. Minimal ascites was seen. The patient was referred to the oncology board for palliative treatment.

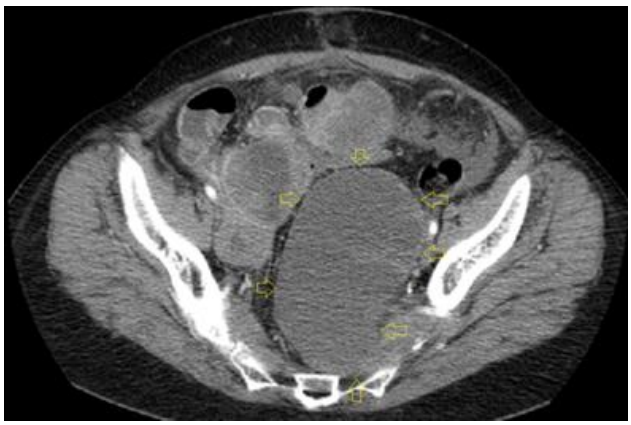

Figure 3 – Axial CT of malignant peripheral nerve sheath tumor (recurrence lesion indicated)

Discussions

MPNSTs are known to be rare and aggressive soft tissue sarcomas, occurring in adults between 20 and 60 years of age [6]. These tumors arise in nerve roots and bundles in lower or upper extremities and pelvis from the sciatic nerve, brachial and sacral plexus, known as large peripheral nerves. In our case report the origin of MPNST was from the sacral plexus and had no history from NF1, known to cause half of MPNSTs in patients with this disease. The other half is known to appear sporadically, as in our case report, and a smaller percent are RT-associated. It is known that the most commonly affected nerve for MPNST located on the trunk is within the lumbosacral nerve roots [7]. In this case report the tumoral mass was located intra-abdominal, obstructing both ureters and leading to dysuria and local discomfort, leading the patient to seek medical attention. Although the tumor resection was successful, the recurrence developed in an unexpected short period of time, growing to the same size within a month from the first surgery. The resection margins were R0. The local recurrence after a MPNST surgery is known to be at 31,4% for sporadic, 35,7% NF1-associated and 53,8% RT-associated [7].

In some cases of MPNST it was found that patients which underwent resection with negative margins and tumors were under 10 cm, had better prognosis [7-10]. Also, radiation therapy in combination with an R0/R1 resection is associated with better local control (5-year local recurrence at 82% versus 67% for those that were not radiated) [7],[11]. Chemotherapy has shown not to provide a significant survival benefit in most patients. Only three patients had complete response to chemotherapy, which illustrates an important aid in select patients. Therefor the therapeutic benefits of chemotherapy remain unclear. A promising treatment remains by targeting activated signaling pathways like: RTKs, RAS/RAF/MEK pathway, PI3K/AKT/mTOR pathway, VEGF pathway and others.

Our case report presents a female patient presenting dysuria and pelvic pain. The clinical examination revealed a palpable mass in the lower abdomen. The computer tomography (CT) scan of the abdomen and pelvic region showed a tumor mass that was in contact with the nearby organs, but not infiltrating them. After the surgery, histopathology confirmed the MPNST proliferation. A month after, the patient returned for a follow-up and presenting pelvic pain. The CT scan of the abdomen and pelvic region showed a tumoral mass compressing the nearby organs, but not infiltrating them. The patient was referred to oncology board for palliative treatment.

The particularity of this case report is the short period of time recurrence of the MPNST, rather than metastases. The most important treatment for MPNSTs remains surgery, trying to achieve negative margins. However, there have been reported two cases of unresectable MPSNT with partial remission to chemo- and radiotherapy.

Conclusion

We address this case to alert clinicians to the aggressive nature of MPNST, which can recur in a month to the same size after a surgical resection R0 grade. A frequent18F-FDG PET/CT or an MRI can be useful in early detection of recurrence.

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