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Case Report

Huge Malignant Peripheral Nerve Sheath Tumor in the Back: A Case Report and Literature Review

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Abstract

Malignant peripheral nerve sheath tumors (MPNSTs) are rare soft tissue sarcomas that arise from peripheral nerve fibers and are derived from Schwann cells, perineural cells, or fibroblasts. Up to half of these tumors arise primarily in the setting of neurofibromatosis. Large tumors could pose difficulty for surgery. We report 53-year-old female with neurofibromatosis type 1 who presented 18.4*16.2* 27.8 cm3 progressively enlarging, painless soft tissue mass in the back present for 30 years. Her daily activity was severely affected by the huge tumor. She received surgical excision and adjuvant radiotherapy. After operation, this patient returned to normal life within 6 months.

Keywords: MPNST; Malignant peripheral nerve sheath tumors; Soft tissue sarcoma; Back tumor

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are defined as malignant tumors arising from a peripheral nerve or showing nerve sheath differentiation, with the exception of tumors originating from the epineurium or peripheral nerve vasculature [1,2]. MPNSTs account for 5-10% of all soft tissue sarcomas. The incidence of MPNST is 0.001% in the general clinic (hospital) population and 4.6% in patients with neurofibromatosis type 1 (NF-1) [3]. These tumors are highly malignant; often affect the head, trunk, and extremities; and lead to poor overall survival [4]. We report a case 53-year-old female with NF-1 who underwent excision for huge MPNST in the back.

Case Report

The 53-year-old female was evaluated for a progressively enlarging painless soft tissue mass in the back present for around 30 years (Figure 1).

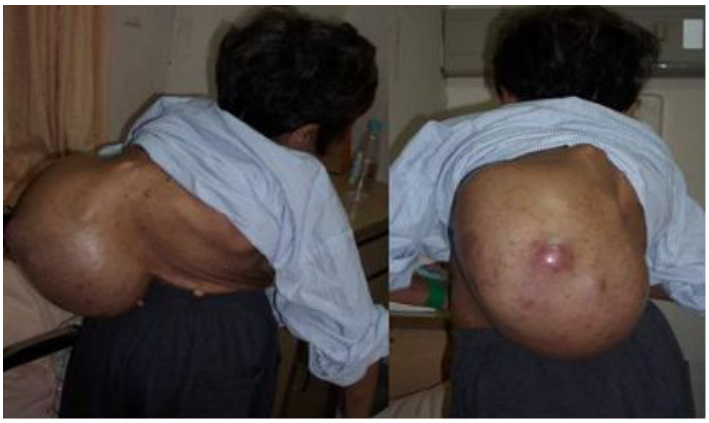

Figure 1: A firm, non-tender soft tissue mass around 18.4*16.2*27.8 cm3 lobulated from her upper back.

She had previous history of NF-1. More than six cafe-au-lait spots were found on her trunk in addition to several skin-fold freckles on her axillary and neck base regions. She had positive family history and her daughter also had NF-1.In out-patient-department, physical examination revealed a firm, non-tender soft

tissue mass lobulated from her upper back. Size of the mass was about 18.4*16.2*27.8cm3. Her daily activity was severely affected by the huge tumor. Although physical examination showed angular deformity of kyphoscoliosis (convexity to left side) in thoracolumbar junction, the motor and sensory function of bilateral lower limbs were free without neurological deficit.

Computed tomography (CT) showed a huge mass (size about 18.4*16.2* 27.8 cm3) at left back and right paraspinal region, at the level of T7 to S4. Intraspinal extension to left side T8-T10 spinal canal with dura ectasia and destruction of left 9th and 10th ribs were also noticed. There are several nodules in left lung, the largest one was around 4.5*3.9 cm over left upper lobe; hence, lung metastasis was suspected (Figure 2).

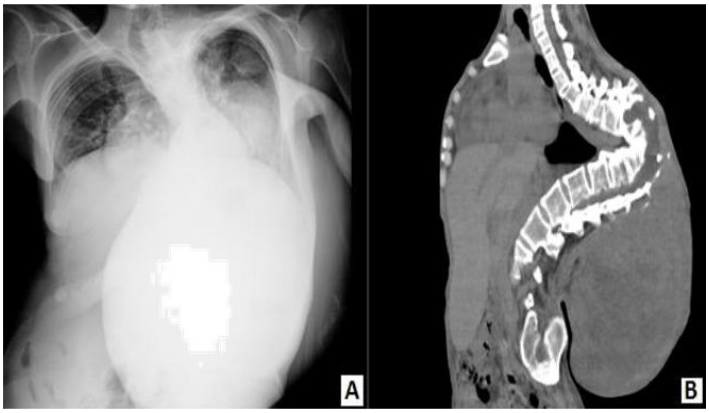

Figure 2: A huge mass at left back and right paraspinal region, at the level of T7 to S4 level. It mainly located in subcutaneous layer, but with intraspinal extension to left side T8-10 spinal canal, suspicious bilateral parapinal muscles invasion at the T7-L1 level. (A)X-ray, (B)Computed tomography.

Magnetic resonance imaging (MRI) revealed left lower back soft tissue mass lesion with prominent tumor vessels and multiple cystic components that might be central necrosis. The patient was put under endotracheal general anesthesia in the right decubitus position, and debulking surgery was performed with longitudinal skin incision along the plane between soft tissue and bony structure. Report of intra-operative frozen section showed malignancy. The excised tumor weighed about 3 kg.

Pre-operation CT showed defect of left chest wall around 8*6cm and a solid nodule around 4.5*3.9 cm over left upper lobe of lung; therefore, thoracoplasty with mesh repair over defect area and wedge resection of left upper lobe for solid tumor was performed by the thoracic surgeon at the same time. Pathology revealed a malignant peripheral nerve sheath tumor arising in neurofibroma characterized by spindle cells displaying a fascicular or whorling growth pattern, with branching hemangiopericytoma-like vascular pattern, as well as alternating hypercellular and hypocellular areas. Proliferation of tumor cells concentrating around blood vessels is also seen with geographic necrosis. The tumor cells are mitotically active (11 mitosis/10hpf), serpentine-shaped with pleomophism, hyperchromatic nuclei and pale cytoplasm. Immunohistochemically, the tumor cells express S-100 but not CD34, desmin, HMB-45, melan A or smooth muscle actin. The pathological report proved the diagnosis of MPNST (Figure 3).

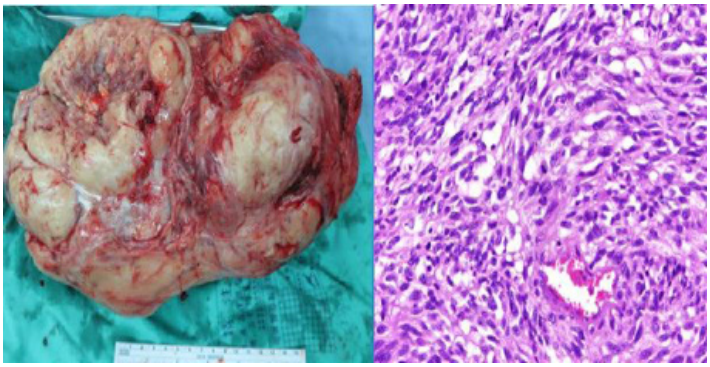

Figure 3: (Left) Tumor gross picture, (Right) Pathology show a malignant peripheral nerve sheath tumor arising in neurofibroma.

The tumor staging was pT2bNxM1, stage 4.After operation, this patient was sent to intensive care unit (ICU) for compromised respiratory function and transferred to ordinary ward 1 week later. The total course of hospitalization was about 3 weeks. Post operation image showed still some residual soft tissue tumor along T8-T9 paravertebral space (Figure 4).

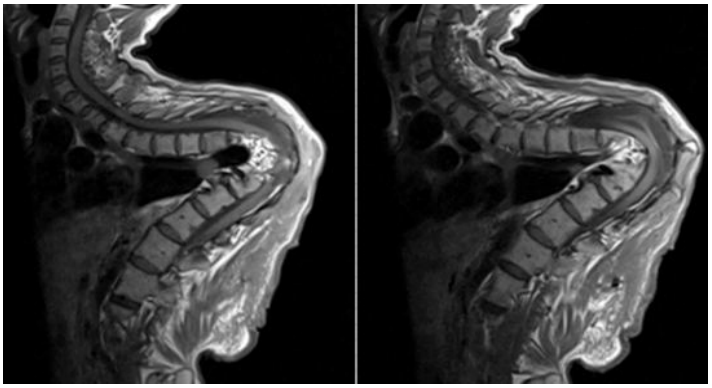

Figure 4: Post-operation MRI showed excision of the tumor with some residual soft tissue tumor.

In post-op third month, the patient began to receive adjuvant radiotherapy (7000cGy in 35 fractions) and chemotherapy (Doxorubicin, Ifosphamide and Mesna). By 6 months follow up, her pain improved and she can lie down and ambulation.

Discussion

MPNST are typically seen in patients between 20 and 50 years of age. Some cases may be seen in the pediatric population. It may occur in the general population sporadically or in patients who have NF-1[5]. People with NF-1 have higher risk to develop MPNST. More than 50% patients with MPNSTs suffer from NF-1, and the life-time risk is 7-13%. Moreover, MPNST tend to pres-

ent at an earlier age in patients with NF1 (3rd or 4th decade of life) compared with patients without NF1 (7th decade)[6]. If the patient had plexiform neurofibroma, the risk of malignant change to MPNST will increase 20-fold[5]. Hint of malignant transformation included plexiform neurofibroma with persistent or nocturnal pain, rapid growing size, change in texture, or new or unexplained neurological deficit. Malignant transformation of neurofibroma is the main cause of mortality in patients having NF-1 with overall 5-year survival rate about 30-50% [7]. In view of the above, differentiating benign from malignant tumors has important prognostic and therapeutic implications.MRI and CT can be employed to determine the site and extent of a lesion, but positron emission tomography (PET) scan may be a sensitive and specific test to differentiate benign plexiform neurofibromas from MPNSTs [8].

Standard care for MPNSTs typically consists of wide surgical excision. Radiation therapy is an important adjunct to surgery in improving local control and may be administered in the neoadjuvant, adjuvant setting or intraoperative therapy [9].The use of chemotherapy as a second adjuvant option in the treatment of MPNSTs remains controversial. Adjuvant chemotherapy (usually consisting of anthracycline-ifosfamide-based combinations) has also been incorporated into the treatment regimen to improve systemic control and reduce rates of distant recurrence [10].

In plexiform neurofibroma in NF-1, tumor size ≥ 5 cm and new onset of pain or neurological symptoms may predict malignant change that required advanced survey [11,12]. Complete surgical excision is needed, but tumor-free margin is difficult to achieve, especially in large tumors. Complete resectability rates are determined primarily by neuroanatomic location, and are reported to be around 95% for extremity lesions and 20% for paraspinal lesions [13]. According to the reviewed literature, combination of multidisciplinary treatment regimen is suggested to obtain a relatively better outcome.In our patient, the treatment plan focused on relief of symptoms and improvement of life quality. Although PET-scan was not performed before operation because of the patient's economic difficulties, malignant change of the tumor should be taken into consideration according to the gross morphology and pre-op image study. Furthermore, pre-op tumor embolization to decrease intra-operation blood loss had been considered but was not recommended by radiologist because of the distorted anatomic structure. After wide excision, adjuvant radiotherapy is needed because it is very difficult to achieve a clear surgical margin in our patient. Moreover, careful pre-op planning is also important for any combined lesion. The thoracic defect and lung tumor of our patient were both resolved by the chest surgeon. After operation, this patient returned to normal life within 6 months. By proper treatment, better life quality for patients with huge MPNSTs can be provided.

Conclusion

Among tumors occurring in the spinal region, malignant peripheral nerve sheath tumors are very rare and may arise either in a sporadic form or from a pre-existing neurofibroma associated with underlying NF1.Most important of all, in NF-1 patients with large tumors, malignant change should always be kept in mind. Complete surgical excision can be achieved in relatively small tumors, but it is very difficult to make a tumor-free surgery for large tumors. Though existing treatment has its limitation for the outcome, well-planned surgery with combined adjuvant radiotherapy is still the treatment of choice for this kind of situation to provide a better life quality.

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