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Case Report

Malignant Peripheral Nerve Sheath Tumor: Treat or Not Treat?

Abstract

Malignant peripheral nerve sheath tumors (MPNSTs) are uncommon, biologically aggressive soft tissue sarcomas of neural origin that poses tremendous challenges to effective therapy. MPNSTs are among the most challenging mesenchymal malignancies to treat with poor prognosis. They usually affect young and middle-aged adults, tend toward early metastasis, and often demonstrate resistance to chemotherapy. We present a case of a 23-year-old female who initially presented with the right temporal swelling for 1 month associated with constitutional symptom which progressively worsening. The right craniotomy and excision biopsy were done with histopathological examination results suggestive of MPNST. Thorax-abdominal-pelvic computed tomography and magnetic resonance imaging further revealed multiple metastatic lesions involving spine, retroperitoneal, pelvic, chest wall, and lungs. This case illustrates the typical presentation of MPNST with its known poorly outcome.

Keywords: *Malignant peripheral nerve sheath tumors, neurofibromatosis type 1, prognosis, survival*

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Introduction

Malignant peripheral nerve sheath tumors (MPNST) typically originate from nerves of the extremities and trunk or from preexisting neurofibromas. The occurrence of MPNST tumors within the neuroaxis is uncommon. Even rarer is the finding within brain parenchyma. Treating it is still a challenge. Here we presented a patient with MPNST and the suggested treatment, either to treat or not.

Case Report

A 23-year-old female was admitted to our hospital with a 1-month history of the right temporal swelling associated with intermittent headache, loss of appetite, and loss of weight, which is progressively worsening. She had a strong family history of neurofibromatosis type 1 (NF1) and one of her siblings passed away due to brain tumor. Clinically, there was a mass at her right scalp measuring 5 cm × 5 cm, which was firm and tender on palpation.

Computed tomography (CT) brain showed an enhancing extra-axial lesion at the right temporoparietal region with bony erosion [Figures 1 and 2]. We have proceeded with right craniotomy and excision of tumor. Histopathological examination results came back suggestive of malignant peripheral malignant nerve sheath tumor with immunohistochemical stain showing that the cells are only positive to vimentin and CD56, while Ki-67 is more than 50%. Thorax-abdominal-pelvic CT was done for surveillance and a synchronous retroperitoneal mass was noted with intraspinal extension and metastatic bony lesion to the right acetabulum [Figure 3]. Hence, magnetic resonance imaging spine was proceeded which revealed tumor deposits at the retroperitoneal and upper lumbar spine involving the L1 vertebral body and epidural extension was noted from T12 to L1 and bilateral L1/L2 foramina as well. However, no surgery was done in view of advance progression of the disease. Instead, she was referred to oncology team.

She had completed 4 cycles of palliative chemotherapy and 10 cycles of radiotherapy before coming again after 5 months completion of treatment with a new complaint of left-sided chest wall swelling with shortness of breath. On examination, there was a mass fixed on the left chest wall, which was firm and measuring 5 cm × 5 cm. Chest X-ray revealed mass at the left lower zone eroding the ribs with ipsilateral pleural effusion causing tracheal deviation. Therefore, she was then referred to palliative care unit.

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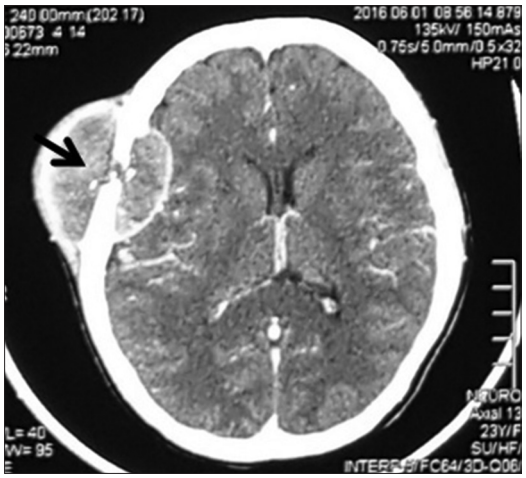

Figure 1: Computed tomography brain showing soft tissue view. An enhancing lesion measuring 7 cm × 7 cm

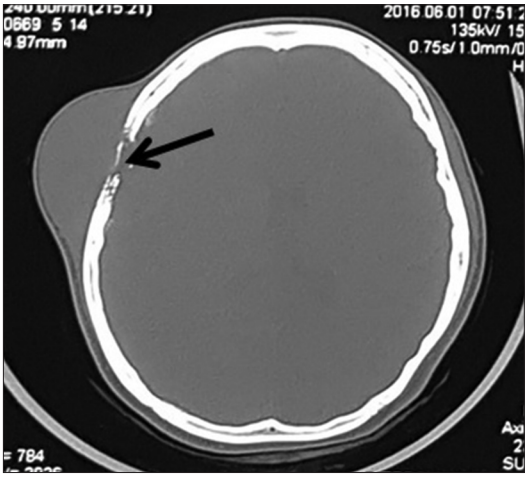

Figure 2: Computed tomography brain showing bone view bony erosion by the underlying lesion

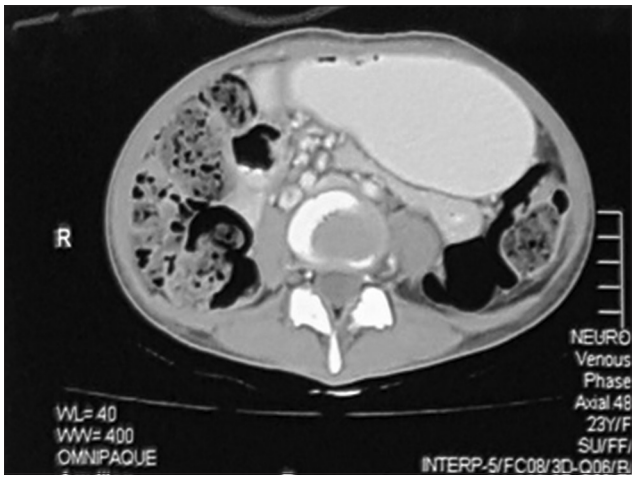

Figure 3: Thorax-abdominal-pelvic computed tomography showing retroperitoneal mass with intraspinal extension

Discussion

A malignant peripheral nerve sheath tumor (MPNST) is a tumor that develops in the protective lining that covers nerves. The first symptom of MPNST is often a lump or mass that increases in size, sometimes causing pain or a tingling sensation. Treatment of MPNST begins with surgery to remove as much of the tumor as possible and may or may not followed by radiation therapy to decrease the chance of a recurrence. Chemotherapy might be used if the whole tumor cannot be removed during surgery or to treat a metastasis.[1,2]

These tumors account for up to 10% of all soft tissue sarcomas[3] and are associated with poor prognosis unless wide excision of the tumor is undertaken before local invasion or distant metastasis can occur. The incidence of sporadic MPNST is low, with a lifetime risk of 0.001%,[4,5] but in association with the familial condition NF1, where these tumors often arise from malignant transformation of a plexiform neurofibroma, the incidence is much higher. Evans *et al*.[6] estimated the lifetime risk of developing MPNST in the population of patients with NF1 to be as high as 13%.

Due to the relative rarity of MPNST, there have been few large studies into survival. The chance of surviving a diagnosis of MPNST depends on the size and location of the tumor; people who have a small tumor tend to survive longer than those with a large tumor, and people with a tumor in the arms or legs tend to survive longer than those with a tumor in the head-and-neck regions.[5,7] Furthermore, MPNSTs that are treated when they first occur have a better prognosis than when the tumor has regrown after initial treatments or spread to distant parts of the body.[7]

One study of 140 patients found that 26% of individuals diagnosed with MPNST were living 10 years after the initial diagnosis.[8] Of those patients who developed a metastasis, 8% were living 10 years after the initial diagnosis.[8] Other than that, large tumor size at presentation (typically >5 cm) has been the most consistently determined adverse prognostic factor across all series.[8-10]

Meanwhile, several studies showed relation between prognosis and NF1 factor. MPNST patient with NF1 related showed worst prognosis compared to sporadic MPNST patient.[11] There has been some evidence that poor prognosis is also reflected by an increased proliferation index of Ki-67 as measured by immunohistochemical analysis, and a number of studies have identified Ki-67 as an independent prognostic factor.[12,13]

To summarize, the patient presented with tumor size >5 cm, tumor location at the head or neck, strong family history of NF1, recurrent or distant metastatic, and increase Ki-67 has poor prognosis.

Conclusion

Based on our illustrated case, our patient presented with brain lesion which its size was more than 5 cm, the patient had significant family history of NF1, presence of distant

metastases, and head-and-neck region as its location and immunohistochemical analysis showed Ki-67 of more than 50%. This is in tally with our aforementioned literature reviews which correlate these peculiar features with poor prognosis. This can indeed be appreciated when our patient came back with new symptoms in <6 months even after completing chemotherapy and radiotherapy. Thus, it can be clearly stated that any patient presented with all the criteria mentioned above would have no benefit for further treatment.

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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Conflicts of interest

There are no conflicts of interest.

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