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Case Report

Malignant peripheral nerve sheath tumor of the scalp: Two rare case reports

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Abstract

Background: Malignant peripheral nerve sheath tumors (MPNSTs) constitute a group of soft tissue neoplasm with neuroectodermal origin. Most cases are small at presentation and only some have been described reaching giant dimensions.

Case Description: We report two cases that were diagnosed and treated as giant MPNST of the scalp. Both patients had extensive lesion on the head with intracranial infiltration. Microsurgical resection was indicated and a vascularized free flap was used to cover the defect. During follow-up the tumors recurred and further surgical excision treatment by adjuvant radiation therapy was performed.

Conclusion: MPNSTs of the scalp are rare neoplasm of the peripheral nervous system. They are aggressive lesion that can recur and their management requires a multimodality approach.

Key Words: Malignant peripheral nerve sheath tumors, scalp, soft tissue tumor

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INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) of the scalp are rare neoplasms of the nervous system. These tumors are considered to be a subcategory of soft tissue sarcomas based on World Health Organization (WHO) classification on central nervous system (CNS) tumors,[14] because they arise from peripheral nerve sheaths and their cells of origin show divergent differentiation potentials.[1,9,16] The lesion in CNS was first described by Kudo *et al.*, and introduced into the English literature in 1983.[11] It is currently classified by the WHO as MPNST in 2016, which further designates two subtypes of MPNST: epithelioid MPNST and MPNST with perineurial differentiation.[14] A variety of terminologies including neurofibrosarcoma, neurogenic sarcoma, malignant schwannoma, and malignant neurilemmoma have been used to describe the tumor.[17]

MPNSTs are very rare tumors with incidents of approximately 0.001% in the population;[4] these tumors usually affect the proximal extremities and trunk and are very rare in the head/scalp and neck[15] and these tumors behave in a highly malignant fashion and they are associated with a poor prognosis with only about 30% of patients surviving

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beyond 5 years.[1,17] The association of MPNST with neurofibromatosis type 1 (NF-1) has been reported; 20–50% of patients with MPNST also have NF-1,[5] and besides *de novo* cases MPNST are usually seen as a result of malignant transformation of a preexisting plexiform neurofibroma.[6]

In this report, we present two cases adding the evidence of scalp MPNST in world literature that were diagnosed and treated as giant MPNST of the scalp. Patients underwent microsurgery and adjuvant radiotherapy.

CASE REPORTS

Case 1

A 45-year-old male presented with painless and progressive swelling on the right frontal region over 2 years prior to examination. According to patient history, the "swelling" started over his eyebrow and progressively increased in size. He underwent a biopsy at an outside facility and the pathology report diagnosed neurofibroma but patient failed to follow-up. He was later referred to our hospital after the lesion enlarged in size and he had developed difficulties opening his eyes. He had no history of trauma, bone pain, systemic disease, or neurological symptoms. Physical examination found an extensive scalp lesion that measured 30 × 20 cm in size, extending from the right orbital rim toward the contralateral side and the parietal region. Upon palpation the lesion was firm but without any tenderness [Figure 1]. The skin on the tumorous area was adherent to the underlying soft tissue and the lesion was immobile. There was no visible venous dilatation or any audible bruit over the lesion. A small post biopsy scar was present at the middle of the lump. Neurological examination was unremarkable and the patient did not bear any signs of NF-1 as well as in their family tree.

Computed tomography (CT) scan of the head revealed a calvarial soft tissue mass predominantly located in right frontoparietal with infiltrating mass on the frontal region and associated right frontal bone defect [Figure 2a and b]. Magnetic resonance imaging (MRI) of the brain revealed a soft tissue mass that was iso-hypointense on T1 showing lobulated mass at frontal region [Figure 3a], which enhanced homogenously after contrast administration [Figure 3b]. The mass extends to the frontal base viewed by coronal plane [Figure 3c] and various intensity on T2 imaging [Figure 3d]. The tumor showed extension from extracranial compartment through the bony defect into the right frontal lobe and frontal base. The decision was made to pursue resection.

The patient was positioned supine with back slightly elevated 20° and without any head fixation. A wide marginal excision with 4 cm distance from neoplasm margin was performed. At surgery, tumor tissue was found to be soft, fleshy, moderately vascular, and mostly encapsulated with some areas displaying ill-defined margins. The mass was eroding through the internal table of the bone and infiltrated the dura mater as well as the intradural compartment. The mass was highly vascular and bled easily when touched. The bone at the right frontal region appeared moth-eaten and was removed with rongeurs until a normal hard and thick border was identified. A wide intracranial portion of the lesion was removed without any involvement of brain parenchyma, and a fascia lata graft was used for duroplasty. The postoperative bone defect measuring 10 × 10 cm was closed using a titanium mesh. A vascularized free flap was raised from anterolateral thigh and sewn in by the plastic surgeon to close the skin defect. The patient's neurological status remained intact postoperatively. He was discharged from the hospital uneventfully.

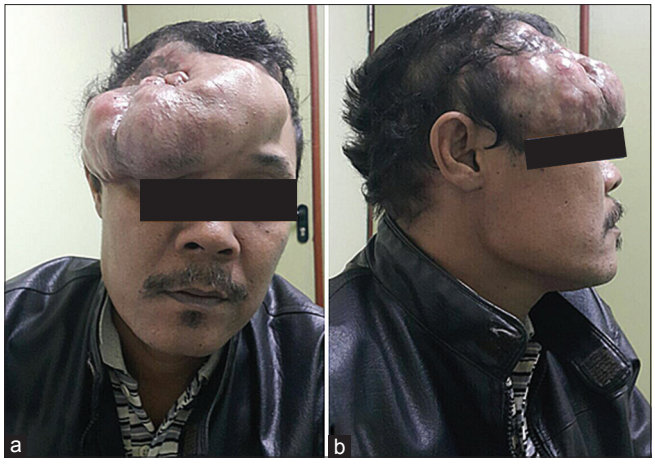

Figure 1: Clinical picture of the patient that had a large swelling on his scalp covering his right eye lid. (a) Front side and (b) right side

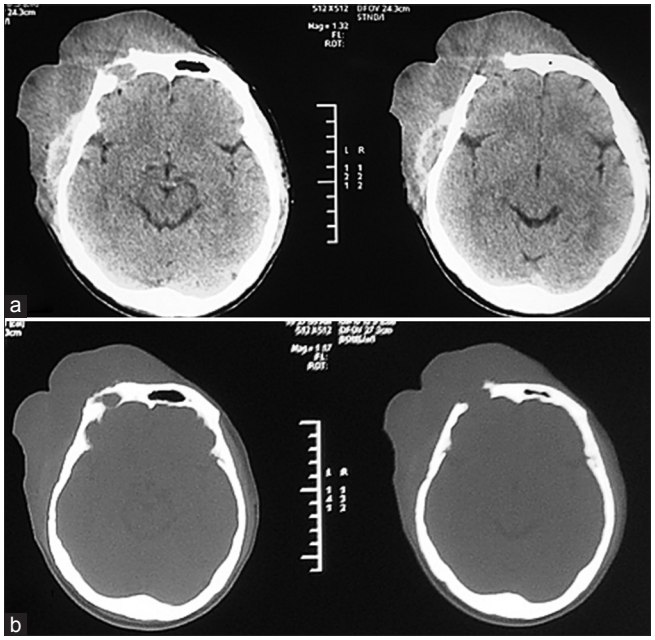

Figure 2: (a) CT scan imaging showing infiltrating mass at the frontal region. (b) CT scan reveal bone discontinuity due to bony destruction

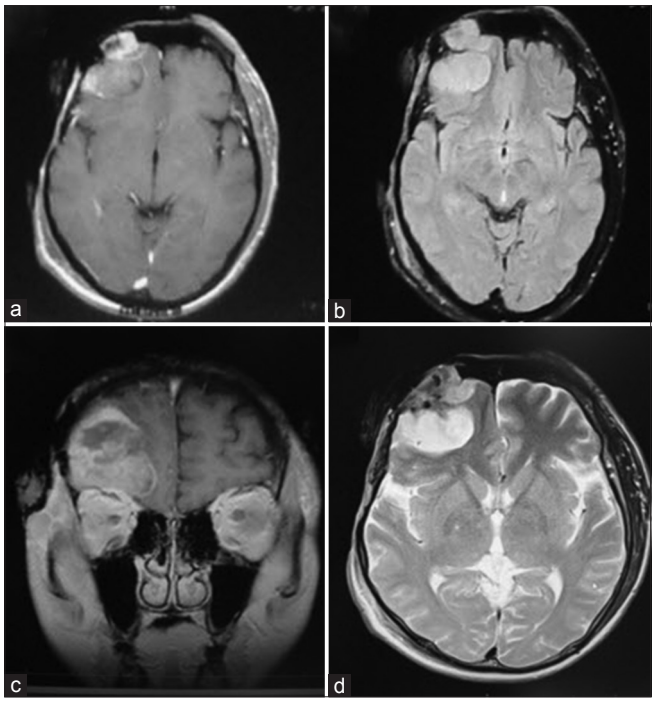

Figure 3:The MRI scan showing (a) lobulated mass at frontal region which (b) enhanced homogenously after contrast administration. (c) The mass extends to the frontal base viewed by coronal plane and (d) shows various intensity on T2 imaging

Over a period of 6 months, the patient was seen in regular follow-up when a recurrence was seen on routine imaging and also new complaint of ptosis on left side. The images show local recurrence on right frontal lobe; interestingly, a new lesion was prominent on contralateral cavernous sinus [Figure 4]. Patient underwent surgery for the right frontal lobe mass and 20 gray external beam radiosurgery for cavernous sinus lesion. There was partial response as the ptosis complaint got better and mass shrink nearly 50% of its size. After 2 years the tumor recurred again on both right frontal and also cavernous sinus, but the patient refused any other medical intervention. Patient died two-and-a-half years after initial diagnosis of this disease.

Case 2

A 49-year-old male patient came to our hospital with a tumorous growth located in the frontal region of his head, which developed over approximately 3 years prior to presentation. He had undergone an operation in another hospital about 2 years earlier but the lump had been growing rapidly over the preceding 6 months. Pathologic analysis revealed malignant schwannoma. On physical examination, we encountered two firm, noncompressible, nontender, nonpulsatile masses that measured approximately 6 × 5 cm and 8 × 8 cm in the frontal region with head CT scan reveal bone discontinuity due to bony destruction [Figure 5].

There were no clinical signs suggesting neurofibromatosis and the family history was negative. The patient underwent microsurgical resection via a frontal craniotomy

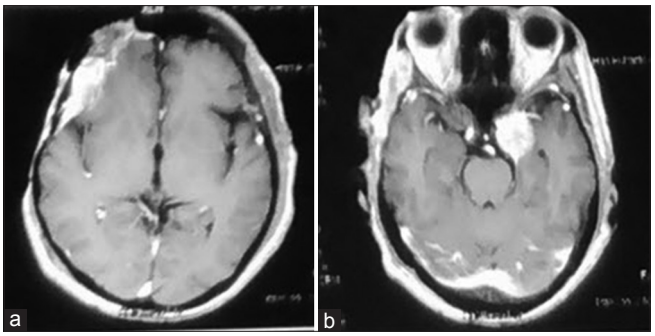

Figure 4: (a) Recurrence of mass on the previous surgery site with marked enhancement on T1 imaging. (b) Contralateral cavernous sinus showing new mass that was not seen on previous images

followed by plastic and reconstructive in a single surgery. The involved bone and a 2-cm margin of healthy tissue were excised together with the tumor mass. The involved bone was brittle and soft, so it rongeured until thick and healthy bone was encountered. There was no infiltration of the underlying dura mater and the lesion could be completely excised en bloc. Cranioplasty was required to close the resulting 4 × 4 cm calvarial defect. After excising the mass, the scalp defect measured approximately 15 × 12 cm and the reconstruction was completed using a myocutaneous flap with a muscle cuff along with the vascular pedicle [Figure 6a and b]. The graft was taken from anterolateral thigh and femoral circumflexa artery – superficial temporal artery anastomosis was performed. The wound healed well and no surgical complications arose. Conventional radiotherapy of 50 gray was performed 2 months after the surgery. The final pathological diagnosis of MPNST was made based on examination of the surgical specimen. Light microscopy revealed that the tumor cells were monomorphic spindle-shaped cells with frequent mitotic figures [Figure 6c]. Immunohistochemistry revealed positive immunoreactivity with S-100 and ki-67 protein [Figure 6d and e]. During 1 year follow-up and 3 months interval MRI, no recurrence was seen.

DISCUSSION

The scalp consists of five layers: skin, connective tissue, aponeurosis (epicranial tissue), loose connective tissue, and periosteum. Any of these tissue layers may be associated with tumors, either benign or malignant.[13]

MPNSTs are rare tumors with an incidence of approximately 0.001% in the general population with slight male predominance and accounting for 5–10% of all soft tissue sarcomas.[4,16] MPNST is located mainly in the trunk and extremities, such as the buttocks, thighs, brachial plexus, sciatic nerve, and paraspinal region. Primary MPNST occurring in the scalp appears extremely rare,[15] and in 2013, Wang *et al.* reported only 16 such cases in the English literature.[17]

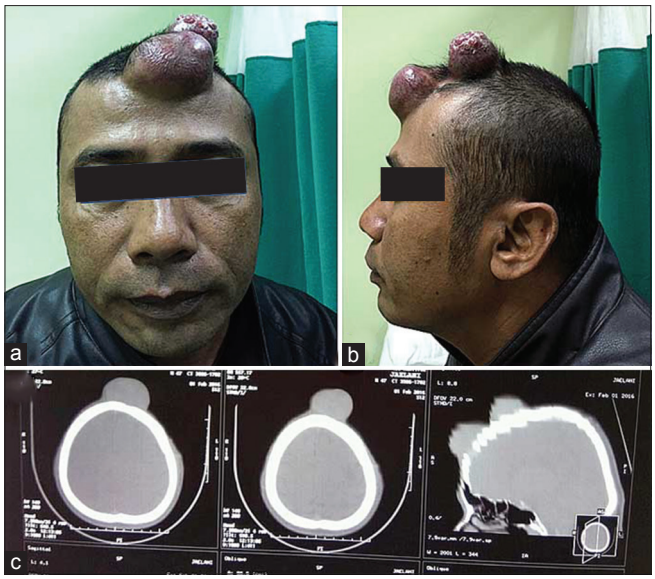

Figure 5: Clinical picture of the patient with two large lumps on his head. (a) Front side and (b) left side. (c) Head CT scan reveal bone discontinuity due to bony destruction

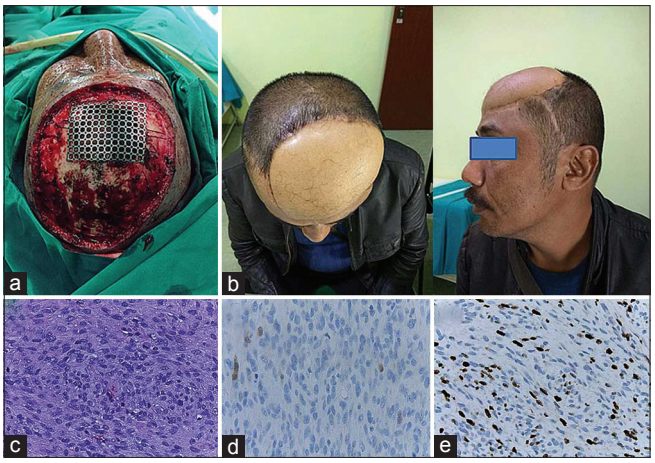

Figure 6: Intraoperative findings. (a) A cranioplasty measuring 4 × 4 cm was performed with a large scalp defect. (b) Postoperative image after 6 months. (c) High-cellular density of spindle-shaped tumor cells with frequent mitotic figures (high power fields with hematoxylin and eosin stain). (d) S-100 immuno-histochemistry, IHC. (e) Ki-67 IHC

The etiology is unknown but they have a known association with NF-1,[5] an autosomal-dominant disorder that involves the NF-1 tumor suppressor gene, which is located on chromosome 17. Up to 50% of MPNSTs occur in patients with NF-1, and there is a higher incidence in patients with a history of radiation exposure.[17,18]

The initial clinical features of MPNST of the scalp are somewhat atypical when compared to other peripheral MPNSTs. Most patients present with a gradually increasing mass in the scalp (cutaneous or subcutaneous). Symptoms of headache, vomiting, seizures, vertigo, visual impairment, or focal neurological deficits may occur in patients with giant MPNST of the scalp if there is any intracranial extension of the tumor.[2,7]

Diagnostic imaging plays a significant a role in determining tumor resectability, assessing surgical risks, and in planning reconstruction. Sequential MRI is necessary for assessing the patient for possible tumor recurrence. CT scan imaging shows the extent of bony involvement, as in both the cases there was destruction of cortical bone. MRI is the investigative tool of choice because it defines the anatomic relationships between the lesion and the adjacent soft tissues, including muscular, vascular, and neural structures.[7,18]

Microscopically, MPNST is a densely cellular tumor that shows fascicular areas with alternate myxoid regions. The swirling arrangement of intermixed dense and myxoid areas has been described as a marbleized pattern.[10] The cells may be spindle-shaped with irregular contours. Malignancy is suggested by features such as invasion of surrounding tissues, invasion of vascular structures, nuclear pleomorphism, necrosis, and mitotic activity. S-100 immuno-positivity has been reported in approximately 50–90% of MPNST cases.[8] In general, a soft tissue panel of antigen stains is used to help exclude other spindle cell lesions and to confirm the diagnosis of MPNST.[8,10,18]

The current management of MPNST is comparable to that of other soft tissue tumors.[4,10,17] Surgery is pivotal in the treatment of MPNST of the scalp. The goal of surgery has to be to achieve complete excision of the tumor with wide (negative) margins (≥2 cm) using frozen section pathology analysis during surgery.[17] After extensive resections, scalp defects are common and reconstruction needs to be undertaken by employing cutaneous or myocutaneous flaps. To this end, a plastic and reconstructive surgery team should be involved, as was done in both of our cases.[4,17]

MPNSTs are aggressive malignancies treated in the same way as other high-grade sarcomas. There are no specific guidelines for MPNST of the head or scalp. These guidelines usually follow soft tissue sarcomas. For those patients with resectable disease, a wide excision through normal uninvolved tissues is the surgical procedure of choice. Defining a rts and a literature reversial, but with the addition of effective adjuvant therapy (e.g. radiotherapy) a tumor-free margin (R0) may be adequate. Where a wide excision is not possible due to anatomical constraints, a planned marginal or microscopically positive margin against a critical structure, plus radiotherapy, for intermediate and high-grade tumors, may be an appropriate means of achieving tumor control while maintaining physical function.[3]

Although complete excisions can seemingly be achieved during surgery, local recurrence occurs frequently and will need further radiotherapy and/or chemotherapy. Local recurrences have been reported in as many as 52–88.9%

of MPNSTs for different sites.[17] Overall survival for these aggressive tumors is often limited and was reported to be around only 30% at 5 years.[3,10,12,17]

CONCLUSION

We describe two rare cases of giant primary MPNST of the scalp that were treated surgically followed by radiotherapy. MPNST should be considered as a differential diagnosis of an enlarging scalp soft tissue lesion and these tumors can show bony and intracranial involvement. MPNSTs are aggressive lesion and a multimodal approach is recommended for management.

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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