

Giant Malignant Peripheral Nerve Sheath Tumor of Scalp in Non-neurofibromatosis Person: A Rare Case Report

Amit Gupta¹ Rakesh Kumar¹ Bal Krishna Ojha¹ Anil Chandra¹

¹Department of Neurosurgery, King Georges Medical Collage, Lucknow, Uttar Pradesh, India

Address for correspondence Dr. Rakesh Kumar, MCh, 4kh-2 Housing Board, Shastri Nagar, Jaipur, Rajasthan 302016, India (e-mail: rksingh2226@gmail.com).

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Abstract

Keywords

- ▶ malignant peripheral nerve sheath tumor
- ▶ occipital
- ▶ scalp
- ▶ S100
- ▶ surgical resection
- ▶ radiotherapy

Malignant peripheral nerve sheath tumors (MPNSTs) are rare neoplasms, usually arising from components of nerve sheath such as perineural fibroblasts or Schwann cells. Primary scalp MPNST is an exceptionally rare entity and only few cases have been reported till date. We encountered an unusual case of giant scalp MPNST in a 70-year-old man without any history suggestive of neurofibromatosis associated with intracranial extension with underlying bone destruction. The tumor was treated with complete surgical excision followed by adjuvant radiotherapy. Histopathological examination showed hyper- and hypocellular areas of spindle cells admixed with myxoid areas and immunohistochemistry revealed positivity to vimentin and S100 on the basis of which the diagnosis of MPNST was made.

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are rare neoplasms with an incidence of about 0.001% in general population. They usually arise from components of nerve sheath such as perineural fibroblasts or Schwann cells, and are often considered as a form of soft tissue sarcoma.¹ Most of these MPNSTs are located in the buttocks, thighs, brachial plexus, and paraspinal region, and are rarely seen in the head and neck. Only few cases of primary scalp MPNST have been reported till date.^{2,3} This article describes a rare case report of MPNST of the occipital scalp region in non-neurofibromatosis person with diagnosis and management protocol.

Case Report

A 70-year-old man presented to the department with a gradually increasing swelling in the occipital region of scalp since childhood. Initially the asymptomatic swelling was of peanut size, which progressively increased over time to the size of a watermelon. Despite such a large swelling, the patient had no neurological symptoms. There was no history suggestive of neurofibromatosis.

On systemic examination the patient's cardiovascular, respiratory, and gastrointestinal systems were normal without lymphadenopathy, subcutaneous nodules, or neurofibromatosis markers. Physical examination of the swelling revealed a giant hemispherical, firm to hard, well-marginated, nonpulsatile, noncompressible/nonreducible, and painless mass that grossly measured 12 × 10 cm in the occipital region more toward the left side of midline (▶ **Fig. 1A**). Computed tomographic (CT) scan and magnetic resonance imaging (MRI) were suggestive of a large well-defined soft tissue-attenuated lesion in the left parieto-occipital region with large underlying bony defect in occipital bone of approximately 8.1 × 14.1 × 14.3 cm (▶ **Fig. 2A–C**).

Surgical excision of the mass was planned. A bone deep circumferential incision was given 2 cm away all around the base of swelling after applying a series of bone deep sutures to reduce the intraoperative blood loss (▶ **Fig. 3A**). The swelling was well-encapsulated even at places with bone defect, and complete excision along with overlying skin and underlying pericranium was achieved. Grossly the swelling was grayish pink in color, soft to firm in consistency, and measured approximately 12 × 10 cm in size (▶ **Fig. 3B**).

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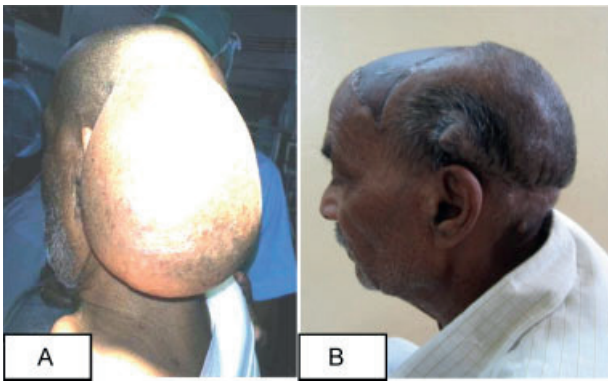


Fig. 1 (A) Preoperative photograph of patient during initial presentation showing a large scalp swelling in the occipital region and (B) postoperative photograph after rotation flap and grafting.

Excision was followed by repair with transposition flap and donor site was repaired with split skin graft from thigh.

On histopathological examination, the sections showed hyper- and hypocellular areas of spindle cells admixed with myxoid areas. Hypercellular areas were composed of dense fascicles of malignant spindle cells having pleomorphic, wavy, or buckled hyperchromatic nuclei and fibrillary eosinophilic cytoplasm. Few areas were composed of rounded, short fusiform cells arranged in nodules, circles, and whorls (→ Fig. 4). Fair numbers of mitotic figures were also seen, but cells were CD34 negative. Depending on these findings, a diagnosis of malignant peripheral nerve sheath tumor was made.

The patient was advised to undergo radiotherapy, but he failed to follow the advice and came back to the department after 6 months with recurrence of swelling at the previous site. The clinical and radiographic presentation was same with the size being slightly larger and the swelling bearing a focus of skin necrosis over its most prominent part. The patient was reoperated and tumor was excised along with wide excision of underlying pericranium and involved bone. Postoperative

radiotherapy was given and the patient was under observation and doing well in last 6 months follow-up.

Discussion

MPNSTs are uncommon malignancies accounting for 5 to 10% of all soft tissue sarcomas.^{1,4} They usually arise from somatic soft tissues or peripheral nerves, or show a nerve sheath differentiation with rare occurrence in the head and neck region. Approximately one-third of MPNST cases arise de novo, but the majority arise from malignant transformation of a preexisting neurofibroma with or without NF1.⁵ The estimated lifetime risk of developing an MPNST in NF1 syndrome is reportedly 10% and could be as high as 30% in patients with symptomatic plexiform neurofibroma.³ Any rapid enlargement in the setting of NF1 should raise the suspicion of malignant degeneration of neurofibroma.

Our patient had a gradually increasing mass in occipital region with rapid enlargement in last 6 months and appeared to have malignant transformation of preexisting neurofibroma as there was no evidence of NF1 or radiation exposure.⁶

MRI is the imaging modality of choice with features of large size, heterogeneity, ill-defined margins, and invasion of bony planes; destruction of normal tissues such as the bone and the edema surrounding the lesion suggest malignant nature.

Grossly the tumor was well circumscribed. The cut surface was multilobular, grayish to white in color with some scattered myxoid, cystic, or necrotic areas.⁷ Microscopically; MPNST is a dense cellular tumor that shows fascicular areas with alternate myxoid regions. The swirling arrangement of intermixed dense and myxoid areas with spindle-shaped, irregular contoured cells has been described as a marbled pattern. Malignancy is suggested by features such as invasion of vascular structures, nuclear pleomorphism, necrosis, and mitotic activity. S100 has been identified in approximately 50 to 90% cases. Leu-7 and myelin basic protein are noted in 50 and 40% of cases, respectively.²

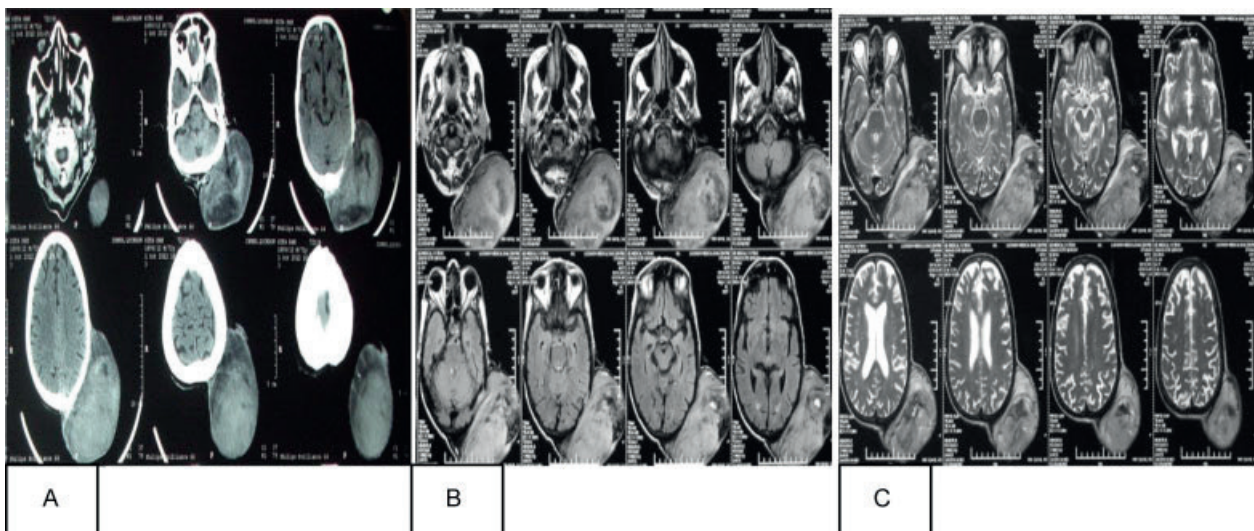


Fig. 2 (A) CT scan (B) MRI T1WI, and (C) MRI T2WI during initial presentation suggestive of a large well-defined soft tissue-attenuated lesion in the left parieto-occipital region with large underlying bony defect in occipital bone.

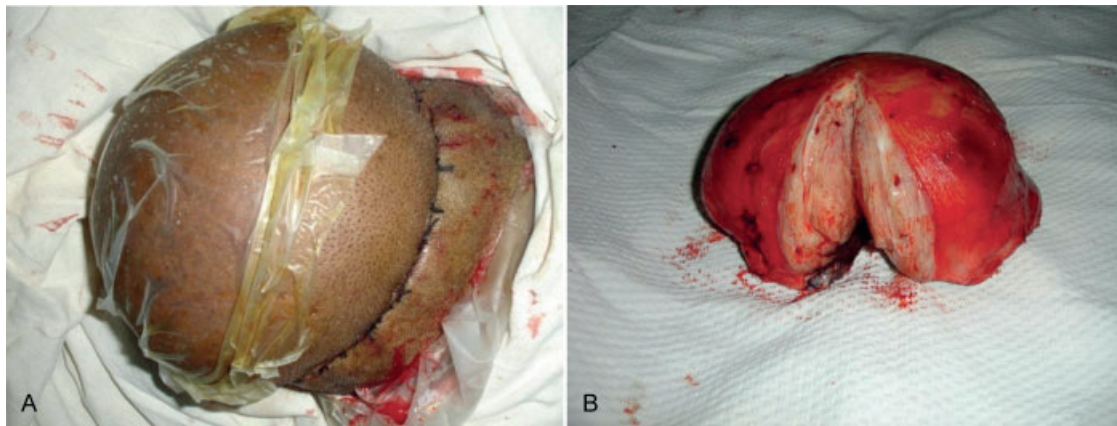


Fig. 3 Intraoperative photograph during surgery (A) and gross specimen after resection (B).

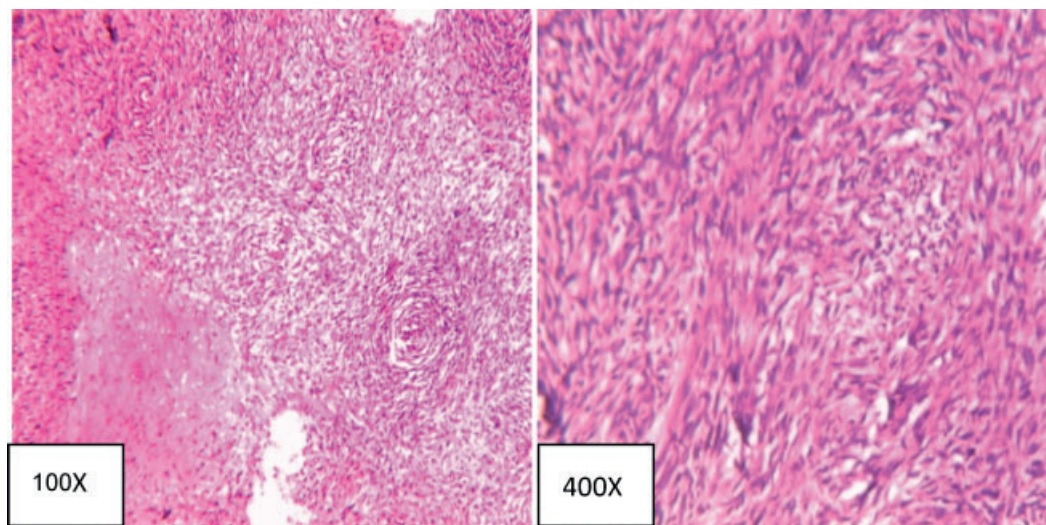


Fig. 4 Tumor cells arranged in fascicles and storiform pattern admixed with few myxoid areas H&E ($\times 100$). Pleomorphic spindle cells with tapered and wavy hyperchromatic nuclei and eosinophilic cytoplasm H&E ($\times 400$).

According to The International Consensus group, the current management of MPNST should be identical to that of any other soft tissue tumors.⁸ The mainstay of treatment is to achieve complete surgical excision of tumor with negative (wide) margins. Positive tumor margins were determined as the most important prognostic factor. Adjuvant radiotherapy should be considered for all intermediate- and high-grade lesions as well as low-grade tumors with positive margins.^{2,3} The role of chemotherapy is usually limited to the treatment of metastatic disease. Local recurrences have been reported to vary from 53 to 88.9% for different sites, whereas metastasis (mainly in the lungs and liver) ranged from 11.1 to 18%.⁹ Angelov et al¹ reported that disease-free and overall survival rates of MPNSTs were reported to be approximately 64 and 30% at 5 years, respectively. Compared with 72 to 78% reported in the soft tissue sarcomas, the MPNST subgroup has a selectively worse prognosis than soft tissue sarcomas.

Conclusion

MPNST of occipital scalp are rare tumors with a poor prognosis. Complete surgical resection with clear margins, if

possible, is recommended. Adjuvant radiotherapy should be considered for all intermediate- and high-grade lesions as well as low-grade tumors with positive margins.

Source of Support

None.

Conflict of Interest

None.

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