

Case Report

Recurrent giant malignant peripheral nerve sheath tumor of the scalp with intracranial extension

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Malignant peripheral nerve sheath tumors (MPNSTs) are rare neoplasms, usually arising from somatic soft tissues or peripheral nerves. The incidence of MPNSTs is approximately 0.001% in the general population. MPNSTs are mainly located in the buttocks, thighs, brachial plexus, and paraspinal region. Scalp is an unusual and surgically challenging site of occurrence. We report a case of giant recurrent MPNST in the left fronto-parietal region, with intracranial extension, in a 58 year old female which was excised completely along with the involved overlying skin, and reconstruction was done for the bony and skin defect. She was given adjuvant radiotherapy. She is doing well after treatment and is in regular follow up. MPNST should be considered by neurosurgeons in the differential of an enlarging scalp soft-tissue lesion with bony and intracranial involvement. Scalp MPNSTs are mostly aggressive lesions, and multimodality approaches are necessary to optimize outcomes

Key words: Malignant peripheral nerve sheath tumors (MPNST), scalp, intracranial, surgery.

INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) are rare neoplasms, usually arising from somatic soft tissues or peripheral nerves. The incidence of MPNSTs is approximately 0.001% in the general population (Al-Gahtamy et al., 2005). MPNSTs are mainly located in the buttocks, thighs, brachial plexus, and paraspinal region (Wanebo et al., 1993). To the best of our knowledge, only 3 cases of MPNSTs of the scalp with intracranial extension have been reported in the English literature (Garg et al., 2004; Ge et al., 2010; Kumar et al., 2007).

We report a case of giant recurrent MPNST in the left fronto-parietal region, with intracranial extension, in a 58 year old female which was excised completely along with the involved overlying skin and bony reconstruction was done for the bony defect.

CASE REPORT

A 58 year old female presented to us with progressive

right sided weakness for the past 3 months along with a gradually increasing nodular swelling in the left parietal scalp. The patient was operated 22 years ago for the swelling at the same site with surgical excision, and bony removal and was advised adjuvant radiotherapy which the patient did not undergo. The details of the previous treatment including the biopsy report were not available to us. She remained asymptomatic for all these years till 3 months back when she started having the present problem. On examination, she was conscious, and oriented with normal higher mental functions. There was a right upper motor neuron type facial involvement. She had right hemiparesis with a power of 0/5 in the right upper limb and 2/5 in the right lower limb. Local examination revealed a 3 × 3 cm hard nodular swelling in the left parietal scalp with erythematous and indurated overlying skin, fixed to the underlying mass. There was a 5 × 5 cm sized bony defect in the left fronto-parietal region reaching midline, and previous surgical scar was healthy.

The brain magnetic resonance imaging (MRI) revealed a 7 × 6 × 6 cm sized extraxial lobulated mass lesion in the left fronto-parietal region, hypointense on T1 weighted images and hyperintense on T2 weighted images with

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Figure 1. (a) Preoperative MRI brain contrast enhanced T1 weighted axial image showing a large extra axial lobulated left fronto-parietal mass lesion compression the brain parenchyma with resultant mass effect. (b) Preoperative MRI brain contrast enhanced T1 weighted sagittal image showing a large extra axial lobulated left fronto-parietal mass lesion compression the brain parenchyma with resultant mass effect along with involved overlying skin.

homogenous enhancement on contrast administration, compressing the brain parenchyma and growing out of the bony defect involving the overlying skin (Figure 1; a and b). The patient was investigated for any lesion elsewhere in the body but none was found. The patient underwent exploration via previous scalp incision and complete surgical excision of the lesion, along with removal of overlying involved skin with cranioplasty for bony defect with methylmethacrylate and closure of the scalp defect with transpositioned scalp flap from right fronto-parietal region and split skin grafting.

Post-operative computed tomography (CT) head showed complete excision of the mass (Figure 2). The patient improved neurologically after surgery. The biopsy report showed proliferation of spindle cells with mild pleomorphism in interlacing fascicles, with alternating hypercellular and hypocellular areas (marbled effect), with cartilaginous metaplasia and buckling of nuclei corresponding to schwannian differentiation. No significant mitoses or necrosis was seen. On immunohistochemistry, the tumor cells were positive for vimentin and S-100, and negative for EMA, CK, CD34 and SMA, and a final diagnosis of MPNST was established (Figure 3). She was given adjuvant radiotherapy and has been in regular follow up for 2 months and is fine.

DISCUSSION

As defined by the World Health Organization, MPNSTs

are malignant tumors arising from a peripheral nerve or showing a nerve sheath differentiation, with the exception of tumors originating from the epineurium or the peripheral nerve vasculature (Kumar et al., 2007). These tumors are treated as a subcategory of soft tissue sarcomas, in which they comprise 3 to 10% of all such tumors (Al-Gahtamy et al., 2005). MPNSTs are associated with genetic alterations such as NF1 loss of heterozygosity.

Approximately one-third of MPNSTs arise *de novo*, whereas the remainder represent a sarcomatous degeneration of a pre-existing plexiform neurofibroma in a neurofibromatosis I or non-neurofibromatosis I patient (World Health Organization, 2000). In present case, as the previous biopsy report was not available, we presume that it was a primary scalp MPNST involving the underlying bone which was excised 22 years ago and which recurred. There were no features of neurofibromatosis I in the present case. MPNSTs usually occur in the third to sixth decades of life and usually affect the medium and larger nerves. Tumor location has been found to be a strong prognostic factor with those in the thoracic and retroperitoneum having worse outcomes (Wanebo et al., 1993). Histopathologically, these are highly cellular tumors that characteristically show a fascicular pattern, spindle-shaped nuclei, and scant cytoplasm. The majority of tumors show geographic necrosis and mitotic activity. Most features were seen in this case, except for mitosis and necrosis.

Magnetic resonance imaging is the investigation of



Figure 2. Postoperative contrast enhanced axial CT head image showing complete excision of the tumor and bony reconstruction.

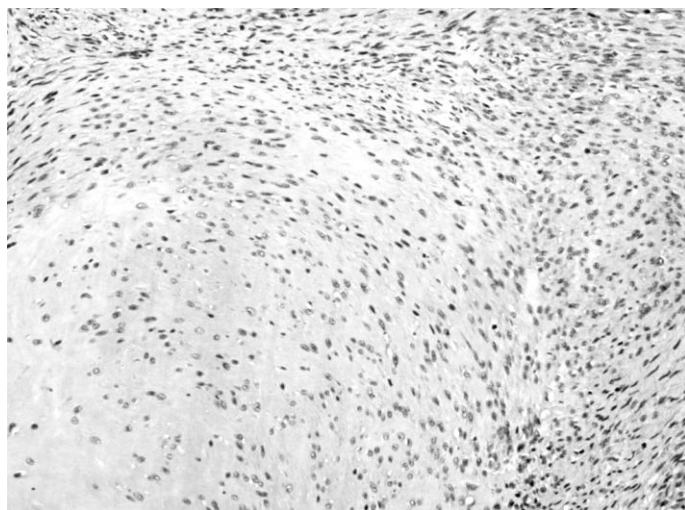


Figure 3. Microphotograph showing interlacing fascicles of spindle cells exhibiting minimal pleomorphism along with buckling of nuclei and cartilaginous metaplasia. (Hematoxylin and eosin, $\times 100$).

choice because it reveals the nerve origin and the relationship to adjacent structures in most cases (Al-Gahtamy et al., 2005). The International Consensus Group has recommended that the current management

of MPNST should be identical to that of any other soft tissue tumors. Adjuvant radiation therapy (RT) should be considered for all intermediate- and high-grade lesions, as well as low-grade tumors with positive margins and was given in this case as well (Angelov et al., 1998; Vege et al., 1994). Local recurrences have been reported to vary from 52 to 88.9% for different sites, whereas metastasis (mainly in the lungs and liver) ranged from 11.1 to 18%. The 5-year survival rate among patients with MPNSTs ranges from 30 to 50% (Angelov et al., 1998; Ferner and Gutmann., 2002; Vege et al., 1994). In this case, it recurred locally and there was no metastatic lesion elsewhere in the body.

In view of the high incidence of local recurrence in MPNSTs after surgery, adjuvant postoperative RT has been shown to improve local control. This could be explored further, especially in sites in which complete surgical resection may not always be feasible (for example, the scalp) with intracranial extension (Kumar et al., 2007; Wilson et al., 1994). With MPNSTs being relatively radioresistant, similar to soft tissue sarcomas, an attempt should always be made for near total surgical debulking of the tumors, and adjuvant postoperative RT could help to reduce the local recurrence.

Conclusion

MPNST should be considered by neurosurgeons in the differential of an enlarging scalp soft-tissue lesion with bony and intracranial involvement. Scalp MPNSTs are mostly aggressive lesions, and multimodality approaches are necessary to optimize outcomes

REFERENCES

- Al-Gahtamy M, Midha R, Guha A, Jacobs WB (2005). Malignant peripheral nerve tumors, in Beyer MS, Prados MD: Textbook of Neuro-oncology. Elsevier Saunders, Philadelphia. pp. 564–571.
- Angelov L, Davis A, O'Sullivan B, Bell R, Guha A (1998). Neurogenic sarcomas: Experience at the University of Toronto. *Neurosurgery* 43:56–65.
- Ferner RE, Gutmann DH (2002). International consensus statement on malignant peripheral nerve sheath tumors in neurofibromatosis 1. *Cancer Res.* 62:1573–1577.
- Garg A, Gupta V, Gaikwad SB, Mishra NK, Ojha BK, Chugh M, Sharma MC (2004). Scalp malignant peripheral nerve sheath tumor (MPNST) with bony involvement and new bone formation: Case report. *Clin Neurol. Neurosurg.* 106:340–344.
- Ge P, Fu S, Lu L, Zhong Y, Qi B, Luo Y (2010). Diffuse scalp malignant peripheral nerve sheath tumor with intracranial extension in a patient with neurofibromatosis type 1. *J. Clin. Neurosci.* 17(11):1443-4.
- Kumar P, Jaiswal S, Agrawal T, Verma A, Datta NR (2007). Malignant peripheral nerve sheath tumor of occipital region. *Neurosurgery* 61(6):1334-5; Discussion E1335.
- Wanebo JE, Malik JM, VandenBerg SR, Wanebo HJ, Driesen N, Persing JA (1993). Malignant peripheral nerve sheath tumors. A clinicopathological study of 28 cases. *Cancer* 71:1247–1253.
- Wilson AN, Davis A, Bell RS, O'Sullivan B, Catton C, Madadi F, Kandel R, Fornasier VL (1994). Local control of soft tissue sarcoma of the extremity: The experience of a multidisciplinary sarcoma group

with definite surgery and radiotherapy. *Eur. J. Cancer* 30:746–751.

World Health Organization (2000). *Pathology and Genetics of Tumors of the Nervous System*. IARC Press, Lyon. pp. 169–171.

Vege DS, Chinoy RF, Ganesh B, Parikh (1994). Malignant peripheral nerve sheath tumors of the head and neck: A clinicopathological study. *J. Surg. Oncol.* 55:100–103.