

Malignant Peripheral Nerve Sheath Tumour of Scalp with Extradural Extension: Case Report

Saçlı Deriye Ekstradural Yayılım Gösteren Malign Periferik Sinir Kılıf Tümörü: Olgu Sunumu

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ABSTRACT

Malignant peripheral nerve sheath tumours are uncommon aggressive soft tissue sarcomas that originate from peripheral nerves or cells associated with the nerve sheath. Scalp malignant peripheral nerve sheath tumours are extremely rare and the role of multimodality management is unclear. We report a 43-year-old female with scalp swelling and rapid growth. Imaging demonstrated an aggressive lesion with extradural extension. A wide excision with negative margins was achieved and the defect reconstructed by rotational flap. The patient was given adjuvant radiation. A follow-up scan done one year post radiation did not show any recurrence. Malignant peripheral nerve sheath tumours of the scalp are extremely rare and such an extradural extension has not been described earlier.

KEYWORDS: Malignant peripheral nerve sheath tumour, Scalp swelling, Neurofibrosarcoma, Malignant schwannoma, Neurogenic sarcoma

ÖZ

Kötü huylu periferik sinir kılıf tümörleri nadir görülen, saldırgan seyir gösteren yumuşak doku sarkomlarıdır. Bu tümörler ya periferik sinirlerden ya da sinir kılıfı ile ilişkili hücrelerden köken alır. Saçlı derinin kötü huylu periferik sinir kılıf tümörleri oldukça nadir görülür ve çoklu tedavi yöntemlerine verdiği yanıt net değildir. Saçlı derisinde hızlı şişme görülen 43 yaşındaki kadın hasta olgu olarak sunulmaktadır. Görüntüleme çalışmalarında ekstradural uzanım gösteren lezyon gösterilmiştir. Saçlı derideki lezyon tümör sınırı negatif olana kadar çıkarılmış, oluşan defekt rotasyonel flep ile kapatılmıştır. Hastaya adjuvan radyoterapi verilmiştir. Radyoterapiden 1 yıl sonra yapılan kontrolde rekürrens görülmemiştir. Saçlı derinin kötü huylu periferik sinir kılıf tümörleri oldukça nadir görülür ve bu tümörlerin ekstradural olarak yayıldığı daha önce gösterilmemiştir.

ANAHTAR SÖZCÜKLER: Malign periferik sinir kılıfı tümörü, Saçlı deride şişme, Nörofibrosarkom, Malign şivannom, Nörojenik sarkom

INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) is a malignant tumour arising from a peripheral nerve or showing a nerve sheath differentiation, with the exception of tumours originating from the epineurium or the peripheral nerve vasculature (5). The term MPNST replaces a number of previously used terms like malignant schwannoma, malignant neurilemmoma, neurofibrosarcoma, and neurogenic sarcoma. MPNSTs arising from large peripheral nerves such as the sciatic nerve, the brachial plexus and the sacral plexus are not uncommon. However, primary MPNST of the scalp is extremely rare and poses additional surgical challenges. Here we describe a presentation and management of a rare case of scalp MPNST with calvarial erosion and extradural extension.

CASE REPORT

A 43-year old woman had noticed a small painless swelling over the left high parietal region of the scalp approximately five years back while combing her hair. The size of the swelling

remained static till six months prior to presentation when it started growing to reach the present size. There was dull aching pain over the swelling. On local examination, there was a firm, globular, nontender swelling of 6x8x8 cm size over the left parietal region. The swelling had indistinct margins from surrounding bone and it was non-compressible and non-transilluminating. The skin over the swelling was tense and shiny and was attached to the underlying swelling. There were no discharging sinuses or dilated veins over the swelling. On systemic examination, there was no lymphadenopathy, neck rigidity or any neurocutaneous stigmata. Magnetic resonance (MR) imaging showed a brilliantly contrast-enhancing extradural tumor in the left parietal region with erosion of the skull (Figures 1, 2). The patient underwent surgery with gross total excision of tumour. The tumour was highly vascular with necrotic areas and was involving overlying skin. The dura was stretched and displaced inwards but was not involved by the tumour. The bone margins and overlying involved skin were also excised with the swelling. The defect thus created was covered by a local rotational flap. The postoperative

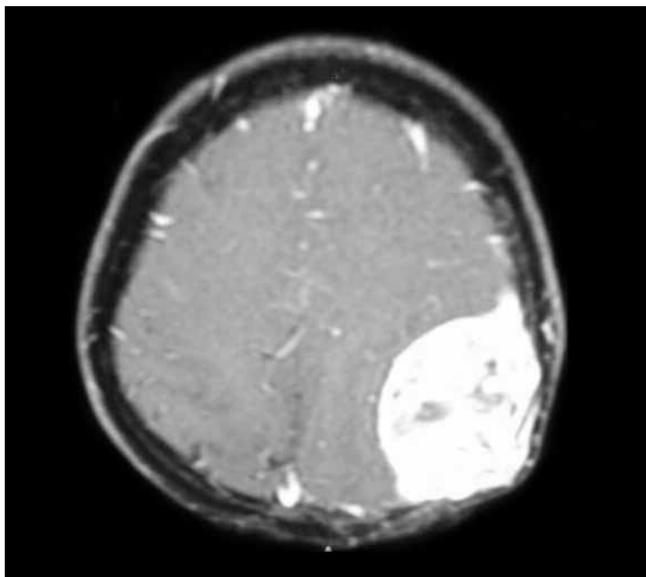


Figure 1: Gadolinium-enhanced axial magnetic resonance image showing large well define irregular mass lesion arising from the scalp eroding the calvarium and extending into the extradural space.

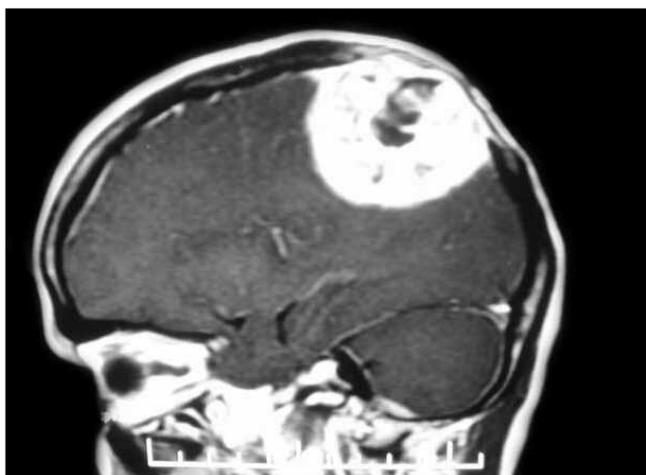


Figure 2: Gadolinium-enhanced sagittal magnetic resonance image showing the lesion to be heterogeneous with necrotic areas and invading the overlying skin.

period was uneventful. Grossly, the operated specimen revealed a partially skin-covered grayish brown globular tissue. Microscopic examination revealed a highly cellular tumour composed of small spindle cells and oval to spindle-shaped cells with bland nuclear chromatin embedded in a collagenous stroma forming intersecting fascicles suggestive of MPNST (Figure 3). The patient was given adjuvant radiation. There was no recurrence at one year of follow-up.

DISCUSSION

MPNSTs are uncommon malignancies accounting for 5 to 10% of all soft-tissue sarcomas with an incidence of approximately 0.001% in the general population (6). MPNST are commonly located in the buttocks, thighs, brachial

plexus, and paraspinal region. To the best of our knowledge, very few cases of MPNSTs of the scalp have been reported in the English language literature (5). This clinical diagnosis should nevertheless be considered for any patient with a rapidly enlarging and painful soft-tissue mass, particularly in the context of neurofibromatosis. Tumor location has been found to be a strong prognostic factor with those in the scalp, thoracic and retroperitoneum having worse outcomes (5, 6).

The etiology is unknown. Approximately one third of MPNST cases arise de-novo but the majority arise from malignant transformation of a preexisting neurofibroma with or without neurofibromatosis1 (NF1). 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). The development of plexiform neurofibroma has been linked to the loss of NF1 gene expression in a mouse model, while the development of MPNST has been related to other genetic insults, such as those involving p53 and p16 (2). While NF1 gene activity does not independently cause MPNSTs, it may in fact predispose these patients to such an event. Any rapid enlargement in the setting of NF1 should raise the suspicion of malignant degeneration of neurofibroma. A higher incidence of MPNST has also been noticed in patients with a history of radiation exposure. Our case appears to have malignant transformation of preexisting neurofibroma as there was no evidence of NF1 or radiation exposure.

Magnetic resonance imaging is the imaging modality of choice. MPNSTs share imaging characters with their benign counterpart but large size, heterogeneity, ill-defined margins, invasion of body planes, destruction of normal tissues like bones and the edema surrounding the lesion are more suggestive of a malignant nature.

Microscopically, MPNST is a densely cellular tumour that shows fascicular areas with alternate myxoid regions. The swirling arrangement of intermixed dense and myxoid areas has been

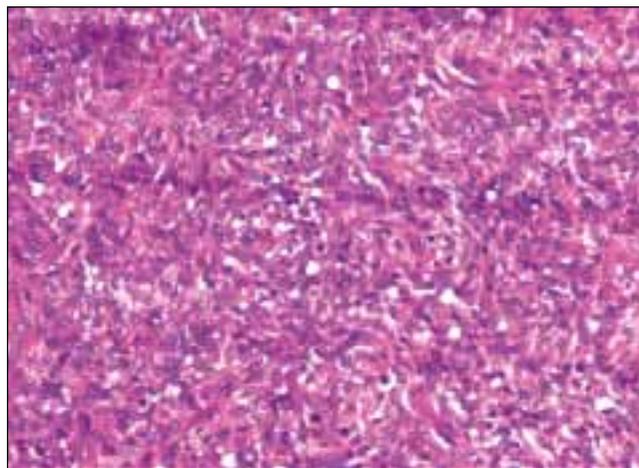


Figure 3: Photomicrograph of the tumour showing highly cellular tumour with variable mitotic activity and nuclear pleomorphism. Areas of hemorrhage and necrosis are also seen.

described as a marbled pattern. 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 has been identified in approximately 50 - 90% of MPNST cases. Leu -7 and myelin basic protein are noted in 50% and 40% of cases respectively (1). In general, a combination of antigens is used to help exclude other spindle cell lesions and to confirm the diagnosis of MPNST.

The International Consensus Group has recommended that the current management of MPNST should be identical to that of any other soft tissue tumours. The mainstay of treatment is surgical excision. The goal is to achieve complete surgical excision of the tumour with negative (wide) margins (5). Positive tumour margins were determined as the most important prognostic marker. Adjuvant radiotherapy should be considered for all intermediate- and high-grade lesions as well as low-grade tumours with positive margins (4). The supportive literature is generally in the context of sarcomas and not specific to MPNSTs. The role of chemotherapy is usually limited to the treatment of metastatic disease. 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% (4, 5). Disease-free survival and overall survival were reported to be approximately 64 and 30% at 5 years, respectively. This is in contrast to the 5-year survival rates of 72 to 78% reported in the soft tissue sarcomas (STS), thereby suggesting the MPNST subgroup has a selectively worse prognosis than STS (5).

In conclusion, MPNSTs should be considered in the differential diagnosis for any patient with a rapidly enlarging and painful soft tissue mass of scalp, particularly with a background of preexisting neurofibroma. Scalp MPNSTs are aggressive lesions, and multimodality approaches are necessary to optimize outcomes.

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