

Malignant Peripheral Nerve Sheath Tumors of the Scalp: Case Report and Review of Literature

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Abstract

Malignant peripheral nerve sheath tumors of the scalp are rare lesions of the nervous system. Only 16 cases have been reported to date. In this report, we present a case of a malignant peripheral nerve sheath tumor (MPNST) of the scalp and retrospectively analyze the clinical features, imaging findings, pathological features, and prognoses of these tumors.

Keywords

Malignant Peripheral Nerve Sheath Tumors, The Scalp, Treatment

1. Background

Malignant peripheral nerve sheath tumors of the scalp are rare neoplasms of the nervous system. A malignant peripheral nerve sheath tumor (also known as “Malignant schwannoma”, “Neurofibrosarcoma” and “Neurosarcoma”) is a form of cancer of the connective tissue surrounding nerves. As defined by the World Health Organization, however, the universal or standard terminology should be a malignant peripheral nerve sheath tumor [1]-[5]. Given its origin and behavior it is classified as a sarcoma [1]-[4]. Although the incidence of a malignant peripheral nerve sheath tumor of the scalp is very low, it is highly malignant and is associated with poor prognosis [1]-[3].

Approximately 25% to 50% of observed MPNSTs occur in patients with NF-1. Like other soft tissue sarcomas, MPNSTs have a tendency to recur locally and spread hematogenously. Despite aggressive surgery and adjuvant therapy, the prognosis for patients with MPNST remains poor; however, to date, prognostic factors have not been identified consistently in the literature [6]-[10].

In this article we describe a new case of MPNSTs of the scalp.

2. Case Presentation

A 15-year-old male patient presented with a swelling in the occipital region that had been gradually increasing for past two years.

It had shown rapid growth during the year prior to being our hospital.

Initially, the swelling was the size of a peanut. The patient had no history of trauma, fever, or infection at this site.

On examination, we found a globular (goose egg size), non-tender, non-compressible, firm to fluctuant swelling measuring approximately 6×6 cm in the occipital region.

A cranial computed topography (CT) revealed a lesion that exhibited higher density to the brain parenchyma, with an adjacent bony defect measuring 46×37 mm in the occipital area.

Magnetic resonance imaging (MRI) revealed a swelling with bony and dural involvement measuring $47 \times 38 \times 52$ mm in the occipital region, which showed nearly equal T1 and T2 signals. Mixed signals were also observed in various portions of the lesions, with partial enhancement in the lesion overall.

The tumor was misdiagnosed as a hemangioma in the occipital area before surgery. After excising the large mass, the wound healed well, and no surgical complications arose.

The diagnosis of malignant peripheral nerve sheath tumor was made through postoperative pathological examination. Light microscopy revealed that the tumor cells were spindle-shaped, with variable mitotic activity and nuclear pleomorphism (Figure 1). Focal hemorrhage and necrosis were also observed.

Immunohistochemistry revealed positive immunoreactivity with Sangtec 100 (S-100) (Figure 2).

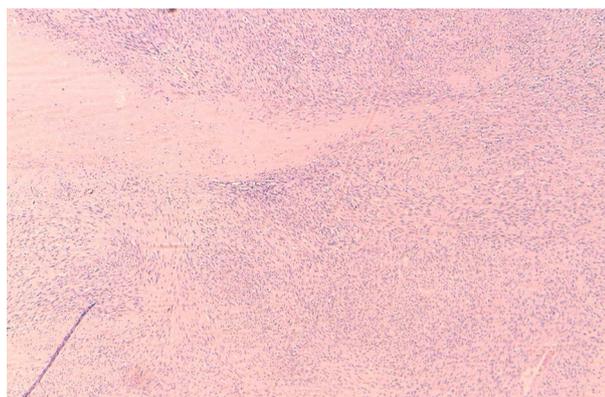


Figure 1. HES $\times 4$: tumor cells were spindle-shaped, with high cell density, variable mitotic activity, nuclear pleomorphism and foci of necrosis.

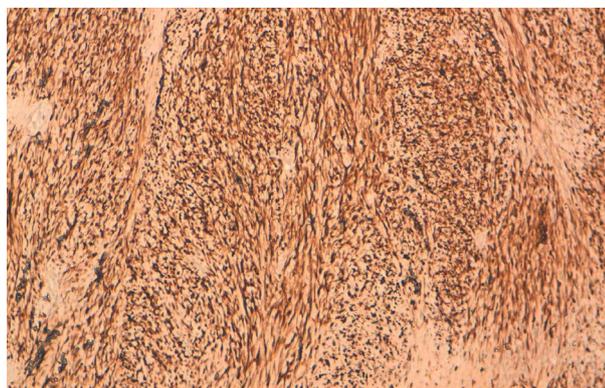


Figure 2. $\times 10$: marquage positif des cellules tumorales à l'anti-corps anti PS100.

3. Discussion

Malignant peripheral nerve sheath tumors (MPNSTs) are rare, with an expected incidence of 0.1/100,000 per year [11], they account for approximately 5% to 10% of all soft tissue tumors with slight male predominance [1]-[4]. Most studies show that the peak incidence of MPNSTs is in the seventh decade of life in the general population, but in the third or fourth decade in people with NF1 [12]; however, these tumors can occur at a much younger age in either population [12] [13].

MPNSTs are defined as any malignant tumor arising from or differentiating toward cells of the peripheral nerve sheath.

The etiology is unknown but they have a known association with neurofibromatosis type 1 (NF-1), an autosomal-dominant disorder that involves the NF1 tumor suppressor gene, which is located on chromosome 17. Up to 50% of MPNSTs occur in patients with NF1 [14], and there is a higher incidence in patients with a history of radiation exposure [1] [14].

MPNST is located mainly in the trunk and extremities, such as the buttocks, thighs, brachial plexus, sciatic nerve, and paraspinal region. Primary scalp MPNST is extremely rare, with only 16 cases reported to date in English literature [1].

Theoretically, MPNST of the scalp can originate from any site on the scalp. Of the 16 reported cases in the literature, the occipital region seemed to be the most common site, accounting for more than 50%.

The initial clinical features of MPNST of the scalp are generally atypical. Most patients present with a gradually increasing swelling in the scalp (cutaneous or subcutaneous) [1] [15]-[21]. Symptoms of headache, vomiting, seizures, vertigo, visual impairment, or focal neurological deficits may affect patients with giant MPNST of the scalp based on the intracranial extension of the tumor [1].

On local examination, the swelling is commonly globular, firm, non-tender, non-compressible and non-pulsatile [1].

Although MPNSTs of the scalp generally lack specificity on imaging features at the early stage, CT or MRI scans are useful for demonstrating the relationship with surrounding structures.

On MRI scans, MPNSTs of the scalp are often hypointense or isointense in T1-weighted series and hyperintense in T2-weighted and Flair sections [1] [22]. MRI scans often reveal features of MPNSTs that differ from those of other scalp tumors, such as lipoma and malignant melanoma [1]. Tumors with cystic degeneration, hemorrhage, or calcification, however, may show mixed densities on CT or MRI scans, which contributes to the high chance of misdiagnosis. Of our case MRI scans demonstrated mixed densities in T1- and T2-weighted series.

The pathological diagnosis is the gold standard for diagnosing MPNST of the scalp. Although MPNST is a heterogeneous group of neoplasms that shows diverse differentiation potential, routine hematoxylin-eosin (H&E) and immunohistochemical staining techniques can help diagnose the majority of MPNSTs of the scalp [1].

Malignant peripheral nerve sheath tumors are unencapsulated infiltrating tumors composed of spindle cells arranged in a whorling pattern with irregular nuclei, cyst formation, and nuclear palisading [23].

Mitotic figures are readily visible, with more than one per hpf, and in 50% to 90% of cases the cells are immunoreactive to S-100 protein staining [24]. Necrosis, pseudocystic change, or hemorrhage may also be found. The pathological criteria for malignancy include invasion of surrounding tissues by tumor cells, vascular invasion, marked nuclear pleomorphism, necrosis, and the presence of mitoses [24].

Immunohistochemical staining of the tumor tissue with a variety of antibodies can diagnose the disease and differentiate it from other diseases. The S-100, EMA, Vimentin (VIM), and CD34 antibodies are highly specific to MPNST of the scalp [4] [6] [25].

Of the 16 cases described in the literature, 12 were immunoreactive for the S-100 protein (12 of 14 cases (86%); not available in two cases), suggesting a high specificity [1].

Other markers, such as glial fibrillary acidic protein (GFAP), cell keratin (CK) and HMB, are usually not expressed in MPNST of the scalp and are therefore used only as references [1]. In general, a combination of antigens is used to help exclude other scalp tumors and to confirm the diagnosis of MPNST especially if S-100-negative.

The International Consensus Group has recommended that the current management of MPNST should be identical to that of any other soft tissue tumors [26]. Surgery, therefore, is the mainstay of treatment for MPNST of the scalp. The goal of surgery is to achieve complete excision of the tumor with wide (negative) margins (≥ 2

cm) [1] [21] [26] [27]. Therefore, patients who had positive surgical margins had a greater risk of dying, mainly from locoregional disease [10].

As reported, surgical excision followed by adjuvant radiotherapy can improve local control [27]. Thus, some authors have recommended that adjuvant radiotherapy should be considered for all intermediate- and high-grade MPNST of the scalp, as well as low-grade tumors with positive margins [27].

Of the 16 cases, surgical excision followed by adjuvant radiotherapy had been done in eight cases. It seems that the current treatment of these highly malignant tumors, however, is not that pessimistic. As some authors have pointed out, the survival rates of patients with MPNSTs were significantly better for superficial tumors, such as MPNST of the scalp [5]. Therefore, further studies with greater numbers of patients are required to confirm the prognosis for MPNST of the scalp [1].

4. Conclusion

The diagnosis of MPNST of the scalp is based on integrating clinical, imaging, histopathological and immunohistochemical findings. Surgical excision with wide margins (≥ 2 cm) and adjuvant radiation should be considered standard treatment.

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