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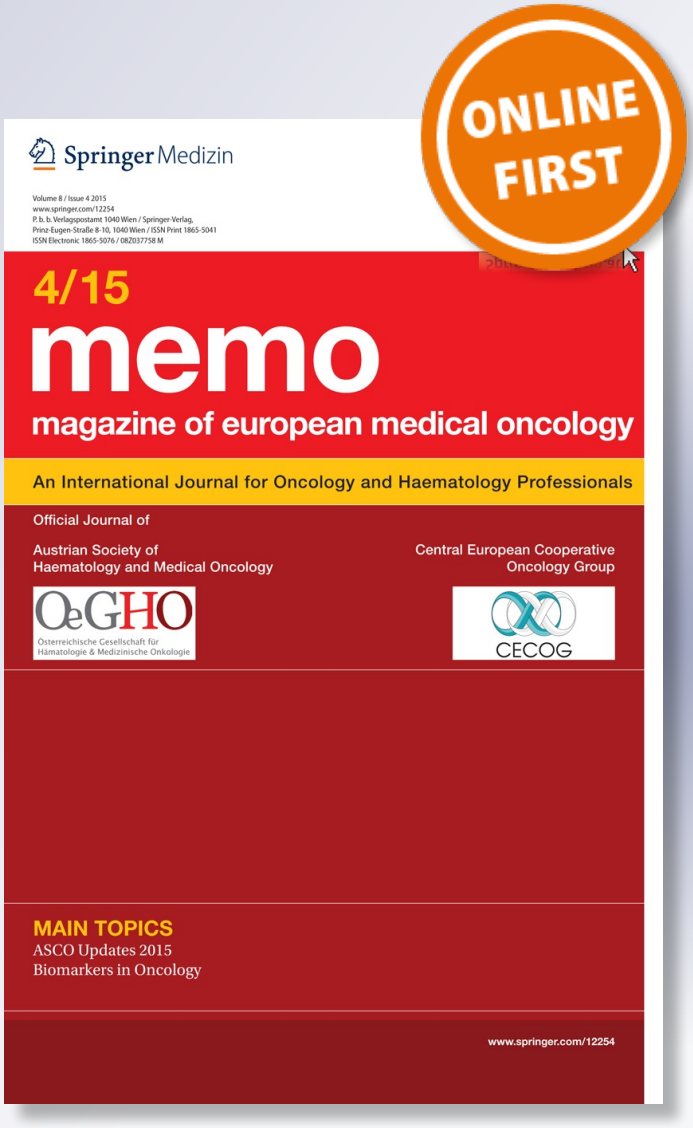

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Parapharyngeal malignant peripheral nerve sheath tumor: a case report and review of the literature

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Abstract We report on a patient who presented to the ear, nose, and throat (ENT) clinic with swelling on the right side of the neck. Imaging studies revealed a large expansive heterogeneous process encroaching on the parapharyngeal region. Postintravenous contrast administration revealed an ovoid mass with moderate enhancement without affecting the parotid gland. The patient underwent full tumor resection, and a biopsy confirmed the presence of a malignant peripheral nerve sheath tumor (MPNST). In addition, as part of the treatment the patient received radiation therapy and is currently disease-free with no visible complications or signs of recurrence.

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Keywords Parapharyngeal · Tumor · Magnetic resonance imaging (MRI) · Nerve · Sheath · Radiotherapy · Computed tomography (CT)

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are uncommon soft tissue neoplasms that usually arise from peripheral nerves. They mainly affect males and females adults in a wide age distribution. Imaging studies are essential for preoperative staging and surgical planning. MPNSTs are staged and treated as malignant soft tissue sarcomas, and the treatment of choice is complete surgical resection.

Case report

A 60-year-old female without a significant medical history presented to the Otolaryngology and Head and Neck Surgery Service complaining of swelling on the right side of the neck. The swelling had been present for 7 years without pain or any neurological deficits. A fine-needle aspiration biopsy was performed yielding inconclusive results. Subsequently, a magnetic resonance imaging (MRI) study was performed revealing a large solid expansive parapharyngeal process with no infringement on the parotid gland. The mass was approximately $2 \times 5 \times 5$ cm in size. An isointense area was seen in T1 sequence and a mildly hyperintense in short T1 inversion recovery (STIR) with moderate enhancement postintravenous injection of paramagnetic contrast. The mass involved the pharyngeal posterior wall decreasing the airway space. The internal carotid artery was displaced anteriorly. Lymphadenopathies were not observed (Fig. 1).

The case was reviewed by the Head and Neck Tumor Board (HNTB), where it was staged as a T3N0M0 para-

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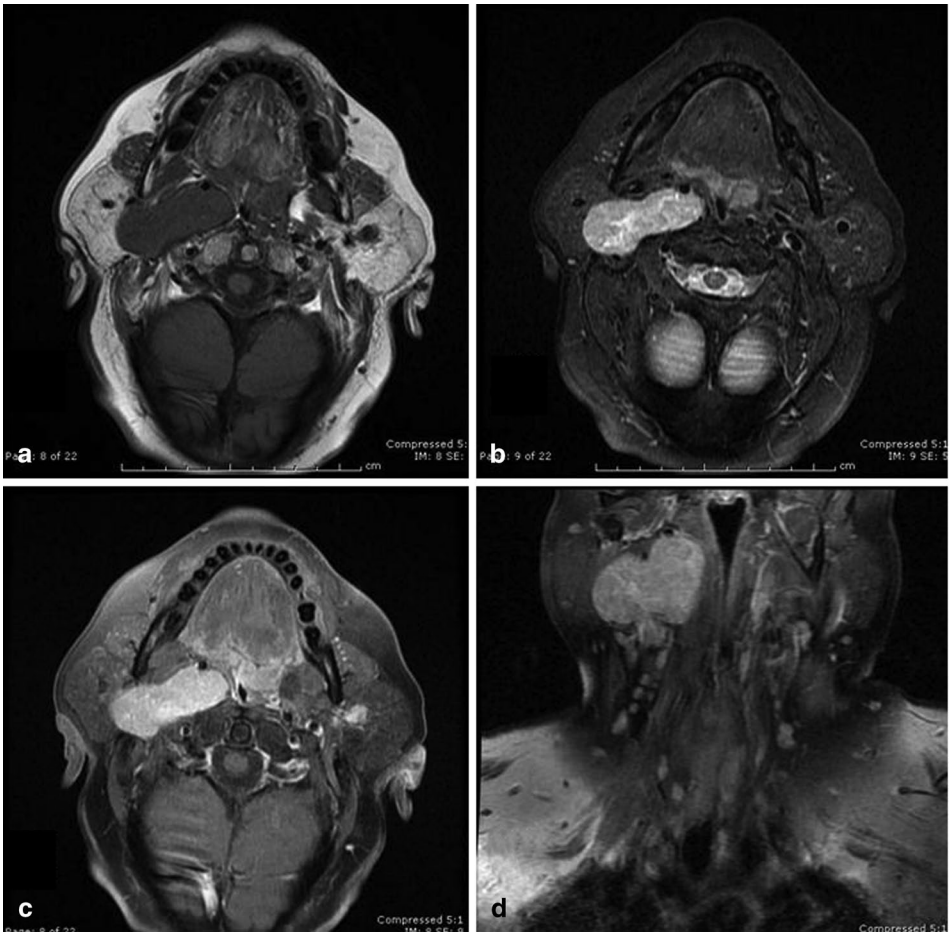

Fig. 1 Magnetic resonance imaging (MRI). Large solid expansive parapharyngeal process with moderate enhancement postintravenous injection of paramagnetic contrast. **a** T1 axial view. **b** Short T1 inversion recovery (STIR) axial view. **c** T1 fat sat Gadolinium axial view. **d** T1 fat sat and gadolinium coronal view

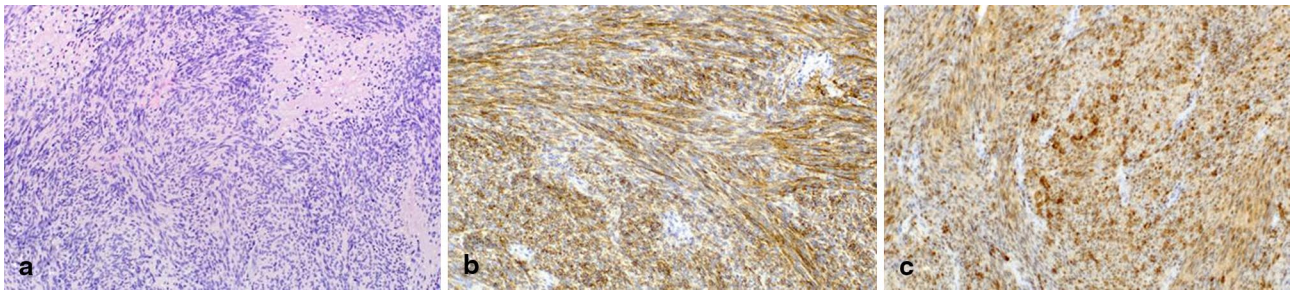

Fig. 2 Immunohistochemistry study (20x). **a** Hematoxylin and eosin stain. **b** Positive CD 56 stain. **c** Positive S100 stain

pharyngeal tumor. The HNTB recommended surgical excision as the best treatment option due to the size of the mass. The postsurgical biopsy confirmed the presence of a low-grade MPNST with no infiltration of adjacent structures (Fig. 2).

Follow-up by the HNTB recommended postoperative adjuvant radiotherapy. Conventional radiotherapy treatment was performed with a Varian 21 iX linear accelerator, with a total dose of 66 Gy, administered in 2-Gy doses during a 7-week period (Fig. 3).

Six months after treatment was completed, a follow-up computed tomography (CT) exam showed no sign of residual tumor or local or systemic recurrence. Currently the patient is disease-free without complications or signs of recurrence (Fig. 4).

Discussion

MPNSTs are uncommon but devastating tumors of peripheral nerve tissues because of their tendency to reoccur and metastasize. MPNSTs are soft tissue neoplasms that usually arise from peripheral nerves and show variable differentiation towards one of the cellular components of the nerve sheath. The incidence of MPNSTs in the general population is 0.001 % [1].

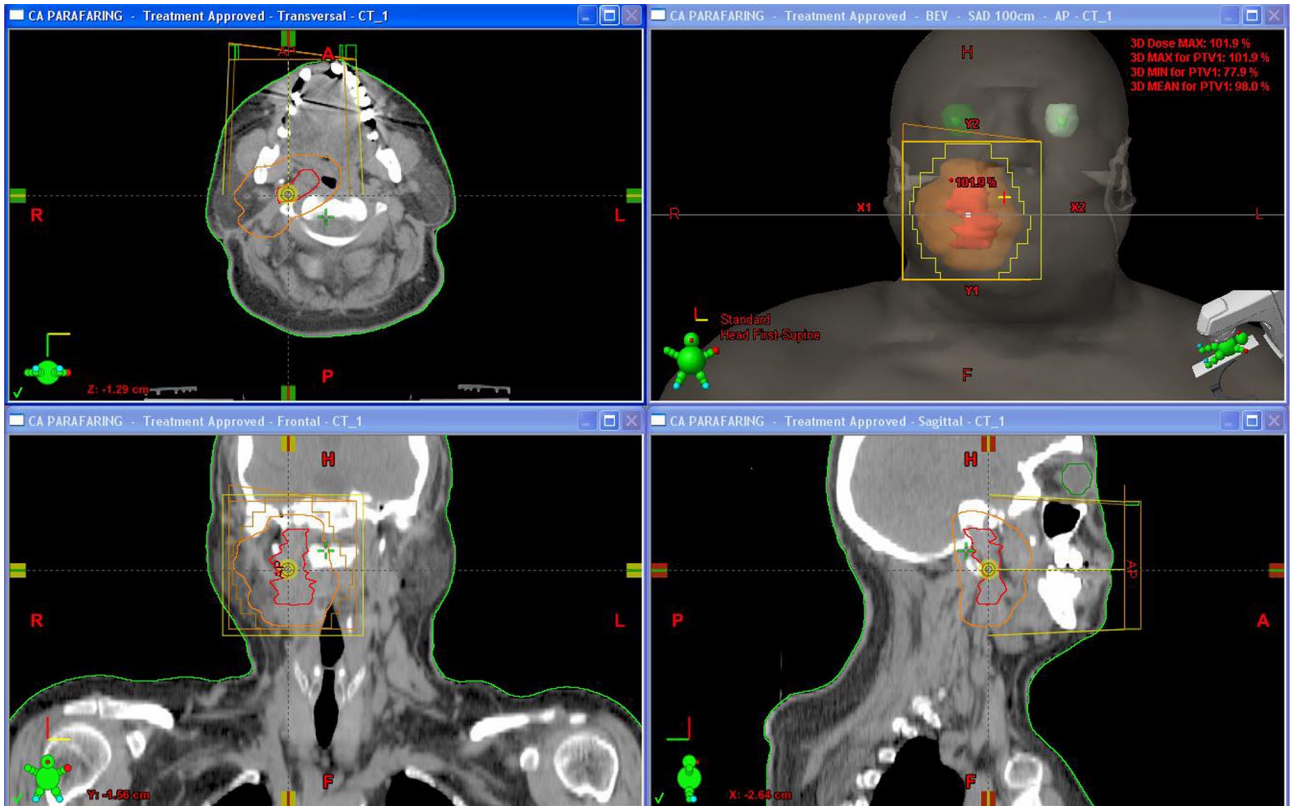

Fig. 3 3-dimentional conformal radiotherapy plan

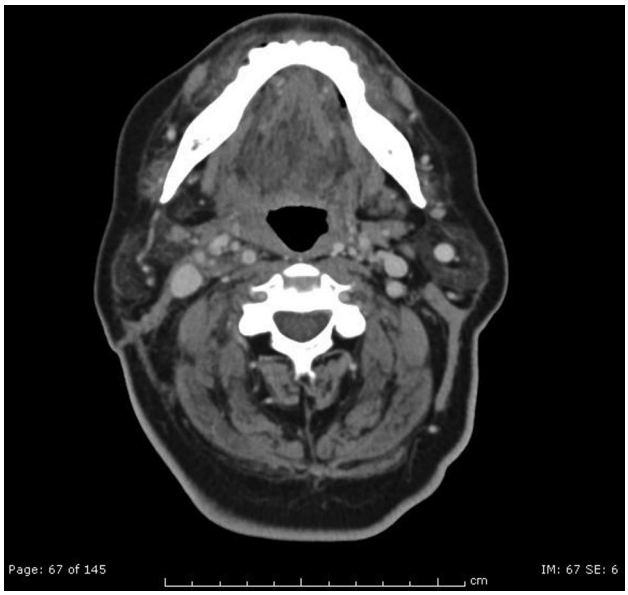

Fig. 4 Follow-up CT, soft tissues window after intravenous contrast injection, after 6 months: patient appears to be disease-free

They can occur sporadically or in patients with neurofibromatosis type 1 (NF1, 20–50 % of MPNSTs) or as a secondary neoplasm 10–20 years following radiation therapy (10 %). Patients with NF1 have a higher risk of developing MPNST (between 8–13 %) [2].

MPNSTs can arise from preexisting plexiform neurofibromas, perineuriomas, or normal nerves. MPNSTs are most commonly found on the extremities and trunk and less often on the head and neck (less than 10 % of total MPNSTs) [3]. They mainly affect adults with a roughly equal sex distribution, and although the age range is wide (20–50 years), they tend to occur at a younger mean age in patients with NF1 [4].

The clinical picture is characterized by a painful or rapidly enlarging mass with associated neurologic deficits [5]. MPNSTs share common imaging findings with other neurogenic tumors such as typical fusiform shape oriented longitudinally along the nerve distribution. Certain findings on CT or MRI should raise the suspicion of a malignant tumor, such as a large tumor greater than 5 cm, with ill-defined margins, heterogeneous, and invading the fat planes with perilesional edema [6].

The diagnosis of a malignant versus benign tumor requires a biopsy, which should be open and involve multiple sections of the tumor. An alternative is CT-guided core-needle biopsy with a good histologic correlation with the final specimen obtained during surgery. CT-guided core-needle biopsies generally have a low rate of complications, mainly exacerbated by pain through the distribution of the nerve during the procedure [7].

MPNSTs tend to be white, large, and solid tumors, containing areas of necrosis and hemorrhage. Microscopically, most MPNSTs are highly cellular, comprised of spindle cells reminiscent of Schwann cells. The cells are mitotically active. The cells are weakly S100 protein

positive, consistent with dedifferentiation from Schwann cells [4, 8].

MPNSTs are staged and treated as malignant soft tissue sarcomas. Because of their rarity and the frequent need for multimodal treatment, the evaluation and management of MPNSTs ideally should be carried out in a facility with expertise in the treatment of sarcomas, including surgical, orthopedic, medical, and radiation oncology.

The treatment of choice is complete surgical resection [2, 5]. The role of adjuvant therapies such as radiotherapy and chemotherapy are not clear. Histologically, MPNSTs are radioresistant and chemoresistant. Nevertheless, post-surgery radiotherapy has been reported to provide some benefit in high-grade, large, or deep MPNSTs or in case of margin invasion [3, 5].

Even with aggressive surgical and radiation treatment, the prognosis is poor. Poor prognostic signs include tumors exceeding 5 cm in size, high tumor grade, association with NF1, older age, distant metastases at the time of diagnosis, and inability to achieve tumor-free margins. The rate of distant metastases is round 40 %. Metastases have been reported all over the body. Common sites are the lungs, liver, bone, and subcutaneous tissue. Less common sites are the endocrine organs, heart, and brain. The 5-year survival ranged from 34–64 % [9].

Conclusion

MPNSTs are rare tumors that mostly affect patients with NF1. They also occur sporadically or in patients who have undergone radiation therapy. The clinical presentation consists of a painfully rapidly enlarging mass accompanied by neurological deficits mainly in the extremities, trunk, head, and neck. A biopsy is required for a definitive diagnosis, and surgery is the treatment of choice. The role of adjuvant treatments is not well established, and the prognosis is poor with a 5-year survival rate ranging from 34–64 %.

Compliance with ethical standards

Conflict of interest

I. Sepúlveda, A. Compan, C. García, E. Platin, C. Delgado, F. Mucientes, and F. Fredes declare that there are no actual or potential conflicts of interest in relation to this this article.

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