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Raven Dunn

Matthew J. Marget

Suhael R. Momin

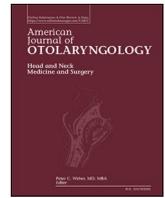
Laura Garcia-Rodriguez

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# American Journal of Otolaryngology–Head and Neck Medicine and Surgery

journal homepage: [www.elsevier.com/locate/amjoto](http://www.elsevier.com/locate/amjoto)

## Recurrent malignant peripheral nerve sheath tumor of the parietal scalp<sup>☆</sup>

Raven Dunn<sup>a,\*</sup>, Matthew J. Marget<sup>b</sup>, Suhael Momin<sup>b</sup>, Laura Garcia-Rodriguez<sup>b</sup>

<sup>a</sup> Wayne State University School of Medicine, 540 E. Canfield Avenue, Detroit, MI 48201, United States of America

<sup>b</sup> Department of Otolaryngology-Head & Neck Surgery, Henry Ford Health System, 2799 W Grand Blvd, Detroit, MI 48202, United States of America

### ARTICLE INFO

#### Keywords:

Malignant peripheral nerve sheath tumors  
Sarcoma  
Neurofibromatosis  
Pediatric  
Facial plastics

### ABSTRACT

Malignant peripheral nerve sheath tumors (MPNSTs) are a rare and aggressive subtype of sarcomas defined by their neural origin. Head and neck manifestations are particularly uncommon. Challenges exist in diagnosis, management, and recurrence. Achieving local control, particularly in the head and neck region, is difficult. We present a patient with a rapidly enlarging MPNST on the right parietal calvarium shortly after resection of a right vagus nerve MPNST. Recommendation was made for excision and reconstruction with a local advancement flap followed by radiation therapy. Local control with good aesthetic outcome was achieved by applying recommended surgical and oncologic principles.

### 1. Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are a rare and aggressive subtype of sarcomas. These tumors are defined by their Schwann cell origin and arise from a peripheral nerve or from benign peripheral nerve sheath tumors. Nearly half of MPNSTs are associated with neurofibromatosis type 1, with very few sporadic occurrences in the general population [1]. This paper will discuss a case of a pediatric patient with a sporadic recurrence of MPNST in an unusual location on the parietal scalp 7 months after treatment of the primary tumor.

### 2. Case report

We present a patient with a rapidly growing MPNST near the occipitoparietal region of the calvarium shortly after resection of a right vagus nerve MPNST.

The patient is a 16-year-old male who initially presented with a 6-cm neck mass. The mass was located at the right angle of the mandible and extended inferiorly to the right neck, level III. The mass was noted to be relatively firm, slightly mobile, and nontender. Magnetic resonance imaging of the soft tissue of the neck demonstrated an approximately 6.8 × 4.2 × 2.9 cm mass deep to the sternocleidomastoid and posterolateral to the carotid sheath. The neck mass was biopsied with pathology consistent with a high-grade nerve sheath tumor. The mass was then surgically excised. Pathology demonstrated a plexiform neurofibroma (staining for S-100 and CD34) with malignant transformation and

consequent loss of S100, high Ki-67 expression, and necrosis. Postoperatively the patient underwent adjuvant proton beam radiation therapy to the neck.

Seven months later, the patient presented with an enlarging, mobile, and painless exophytic soft tissue mass on the right parietal calvarium. A computed tomography of the head with contrast demonstrated concern for a neoplasm and the fine-needle aspiration biopsy suggested a benign adnexal mass, such as a trichilemmal cyst. Recommendation was made for excision and reconstruction with a local advancement flap. At the time of excision, the mass had increased in size to 5.5 × 5.5 cm (Fig. 1). In the operating room, an incision was made circumferentially around the mass with a 1 cm margin. This incision was then carried further down into the galea aponeurotica, where the mass could then be dissected away from the scalp in a subgaleal plane. The resultant defect is shown in Fig. 2. The mass was excised en bloc. Then, an incision was made along the occipitoparietal scalp and a rotational flap was created in the subgaleal plane to reapproximate the defect. Postoperative pathology was positive for a high-grade spindle cell sarcoma with features of MPNST. The patient was later presented to our institution's Multidisciplinary Head and Neck Tumor Board, where treatment recommendation was again made for proton beam therapy.

### 3. Discussion

MPNSTs are rare tumors that can develop sporadically with a low life-time risk of 0.001% [1]. These tumors may also occur in association

<sup>☆</sup> All authors have approved the final manuscript and attest to the integrity of the original data and the analysis reported in the manuscript.

\* Corresponding author.

E-mail address: [rtd@med.wayne.edu](mailto:rtd@med.wayne.edu) (R. Dunn).

<https://doi.org/10.1016/j.amjoto.2020.102812>

Received 19 October 2020;

Available online 24 October 2020

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Fig. 1. Scalp mass preoperatively.



Fig. 3. Reconstruction demonstrating maintained frontal hairline.

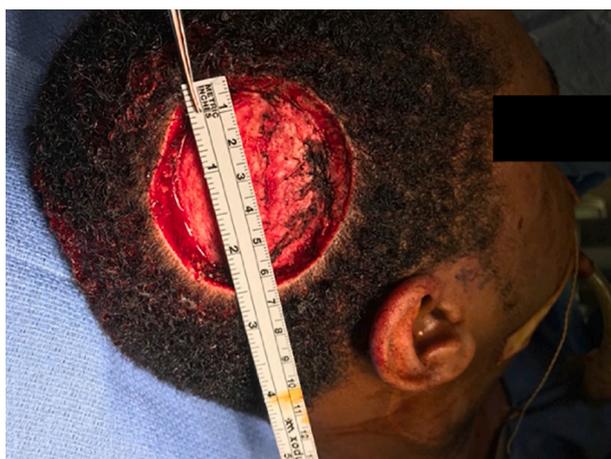


Fig. 2. Resection defect.

with neurofibromatosis, where the risk can be as high as 8%–13% [1]. MPNSTs that arise in the head and neck region constitute only 8%–16% of all MPNSTs and are less than 4% of all soft tissue sarcomas [2]. While initial incidence is exceedingly rare in the general population, recurrence is common. Local 5-year recurrence rates are between 18%–22% if treated with a combination of surgery and radiation therapy but are significantly higher with surgery alone [3]. Distal 5-year recurrence rates are between 36%–42%, with the lungs being most common site of metastasis [2,3]. Minimizing local recurrence can be accomplished through negative surgical margins, radiation, and early detection before the tumor can grow greater than 2 cm in size or spread to local nodes. Achieving clear margins in head and neck MPNSTs can be challenging due to frequent involvement of vital structures.

In our patient, the initial presentation was a right vagus nerve MPNST that had clear margins and no nodal involvement on resection. The patient underwent postoperative radiation therapy. The patient is not known to have NF-1, and he does not meet the major criteria for diagnosis [4]. Genetic testing for neurofibromatosis has been recommended in patients with MPNSTs.

Final pathology of the scalp lesion was positive for loss of H3K27me3 expression. This finding is highly sensitive with moderate specificity for

either sporadic or radiation therapy related MPNSTs. Over 95% of sporadic cases demonstrate a loss of expression for this protein [5].

Proper reconstruction of the scalp is essential for good cosmesis and reducing wound healing complications such as desiccation or dehiscence. This type of reconstruction may be indicated following resection, as seen in our patient, or following traumas such as infection or burns [6]. The anatomy of the scalp is often described by the SCALP mnemonic: S (skin), C (subcutaneous tissue), A (aponeurotic layer), L (loose areolar tissue), and P (pericranium). The loose areolar tissue, or the subgaleal fascia, is what gives the scalp mobility. In this patient, resection of the scalp mass through the subgaleal plane created a defect approximately 33 cm<sup>2</sup> in the parietal region. Tissue in this area is amenable to tissue rearrangement because of the continuity of the aponeurotic layer and the temporoparietal fascia, which overlies the deep temporal fascia rather than the periosteum. This gives more flexibility in advancement of tissue [7].

Approaching repairs requires assessment of size, anatomy, risks of outcome, and affected aesthetics. There are numerous options to close, ranging from primary closure for small defects to free tissue transfers for areas with radiation damage or areas larger than 30 cm<sup>2</sup>, depending on the location of the defect and the patient's individual history. Following the principle of replacing like tissue with like, an understanding of the described algorithmic approaches to parietal scalp defects was used to guide management [6,7]. A local rotational flap was chosen both to improve cosmetic outcomes in a pediatric patient and to minimize concerns for morbidity. Stretch of the flap was aided by intraoperative scoring of the galea for tissue expansion. The patient's outcome was excellent with successful resection and maintained aesthetics as seen in Fig. 3 at the patient's 1-week postoperative visit.

This case demonstrates the importance of screening and monitoring for recurrence in patients with MPNSTs, as the risk of recurrence is quite high. As discussed previously, the patient received proton beam therapy for both presentations, as it has been shown to significantly reduce the risk of local recurrence, likely owing to better local control [3]. The median disease-free state of head and neck MPNST was found to be 9 months, with a 50% 2-year disease-specific survival. Disease-specific survival further declines to 30% at 5 years [8]. This case also exhibits the unusual ways this type of sarcoma can recur. Initial biopsy showed what was believed to be a benign adnexal tumor, possibly a trichilemmal cyst, which was consistent with the patient's clinical presentation of a growing scalp mass without neurological defects. However, upon final pathologic evaluation of the surgical specimen, it was demonstrated that the mass was in fact a recurrent peripheral nerve sheath tumor. If intervention had been delayed, the aggressive nature of the tumor could easily lead to suboptimal surgical margins with a more complex reconstruction needed. Fortunately, our patient was able to have surgical

resection of the recurrent MPNST with negative margins and a favorable cosmetic outcome for reconstruction.

#### Declaration of competing interest

None.

#### References

- [1] Ferrari A, Bisogno G, Carli M. Management of childhood malignant peripheral nerve sheath tumor. *Pediatr Drugs* 2007;9:239–48.
- [2] Owosho AA, Estilo CL, Hury JM, Chi P, Antonescu CR. A clinicopathologic study of head and neck malignant peripheral nerve sheath tumors. *Head Neck Pathol* 2018; 12:151–9.
- [3] Bishop AJ, Zagars GK, Torres KE, Bird JE, Feig BW, Guadagnolo BA. Malignant peripheral nerve sheath tumors: a single institution's experience using combined surgery and radiation therapy. *Am J Clin Oncol* 2018;41:465–70.
- [4] Ferner RE, Huson SM, Thomas N, et al. Guidelines for the diagnosis and management of individuals with neurofibromatosis 1. *J Med Genet* 2006;44:81–8.
- [5] Prieto-Granada CN, Wiesner T, Messina JL, Jungbluth AA, Chi P, Antonescu CR. Loss of H3K27me3 expression is a highly sensitive marker for sporadic and radiation-induced MPNST. *Am J Surg Pathol* 2016;40:479–89.
- [6] Desai SC, Sand JP, Sharon JD, Branham G, Nussenbaum B. Scalp reconstruction: an algorithmic approach and systematic review. *JAMA Facial Plast Surg* 2015;17: 56–66.
- [7] Leedy JE, Janis JE, Rohrich RJ. Reconstruction of acquired scalp defects: an algorithmic approach. *Plast Reconstr Surg* 2005;116:54e–72e.
- [8] Patel TD, Shaigany K, Fang CH, Park RC, Baredes S, Eloy JA. Comparative analysis of head and neck and non-head and neck malignant peripheral nerve sheath tumors. *Otolaryngol Head Neck Surg* 2016;154:113–20.