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Case Report

Massive exophytic malignant peripheral nerve sheath tumor

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ABSTRACT

We present a case of a solitary neurofibroma involving the right posterior shoulder of a 69-year-old man with degeneration into a massive, malignant peripheral nerve sheath tumor measuring more than 3 times the average reported size. The radiographic, magnetic resonance imaging, and computed tomographic features are compared with the gross appearance and pathology.

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Introduction

We present a case of a solitary neurofibroma involving the right posterior shoulder of a 69-year-old man that over the span of approximately 3 years underwent degeneration into a massive, malignant peripheral nerve sheath tumor (MPNST), ultimately measuring more than 3 times the average reported size. The radiographic, magnetic resonance imaging (MRI), and computed tomographic (CT) features are described and demonstrated, as is the gross appearance and pathology.

Case report

A 69-year-old Caucasian man initially noticed a painful, right posterior shoulder mass in 2012, at which time he presented to an outside institution for evaluation. Shoulder radiography was performed and demonstrated a large soft-tissue mass

without appreciable calcification or tumor matrix (Fig. 1). An MRI was also obtained and more clearly defined a 10-cm by 5-cm by 5-cm homogeneous lesion within the posterior and superior soft tissues of the right shoulder that appeared hypointense on T1-weighted imaging, hyperintense on fluid sensitive sequences, and with homogenous internal enhancement after the administration of intravenous gadolinium-based contrast (Figs. 2A–D). The mass was subsequently biopsied under image guidance, and pathology was consistent with a neurofibroma.

The patient presented to our institution approximately 3 years removed from initial work-up, reporting the mass had undergone a recent rapid increase in size and with worsening associated pain. The patient's history was otherwise noncontributory; specifically, there was no history of neurofibromatosis or report of prior malignancy, surgery, or trauma to the affected region. On physical examination, the mass appeared superficial to the right trapezius muscle, in keeping

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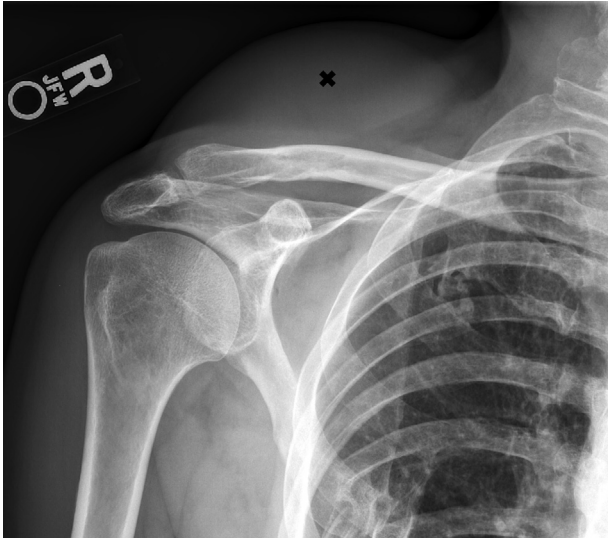


Fig. 1 – Initial presentation. Frontal radiograph demonstrates large soft-tissue density mass superior to the trapezius without appreciable calcification or tumor matrix.

with the prior MRI, and the overlying skin was attenuated but without fungation (Figs. 3A and B). His right shoulder range of motion was preserved, and his right upper extremity was neurovascularly intact.

A repeat MRI was obtained at our institution and demonstrated a 20-cm by 18-cm by 15-cm mass hypointense on T1-weighted imaging, heterogeneously hyperintense on fluid sensitive sequences, with heterogeneous internal enhancement after the administration of intravenous gadolinium-based contrast, and with numerous flow voids attributable to multiple large vessels (Figs. 4A–D). Computed tomographic imaging performed at an outside institution in 2015 just before his referral was reviewed, and notable for a lack of soft-tissue calcification or appreciable internal tumor matrix (Fig. 5). Given the increase in size and progressive pain, an ultrasound-guided biopsy was performed, and pathology confirmed a spindle cell sarcoma consistent with MPNST (Figs. 6A and B).

The patient was discussed at the multidisciplinary tumor board at our institution after biopsy. This panel includes the department of pathology, oncology, orthopedic oncology, and radiology. Consensus regarding management was established, and included neoadjuvant radiation, followed by tumor

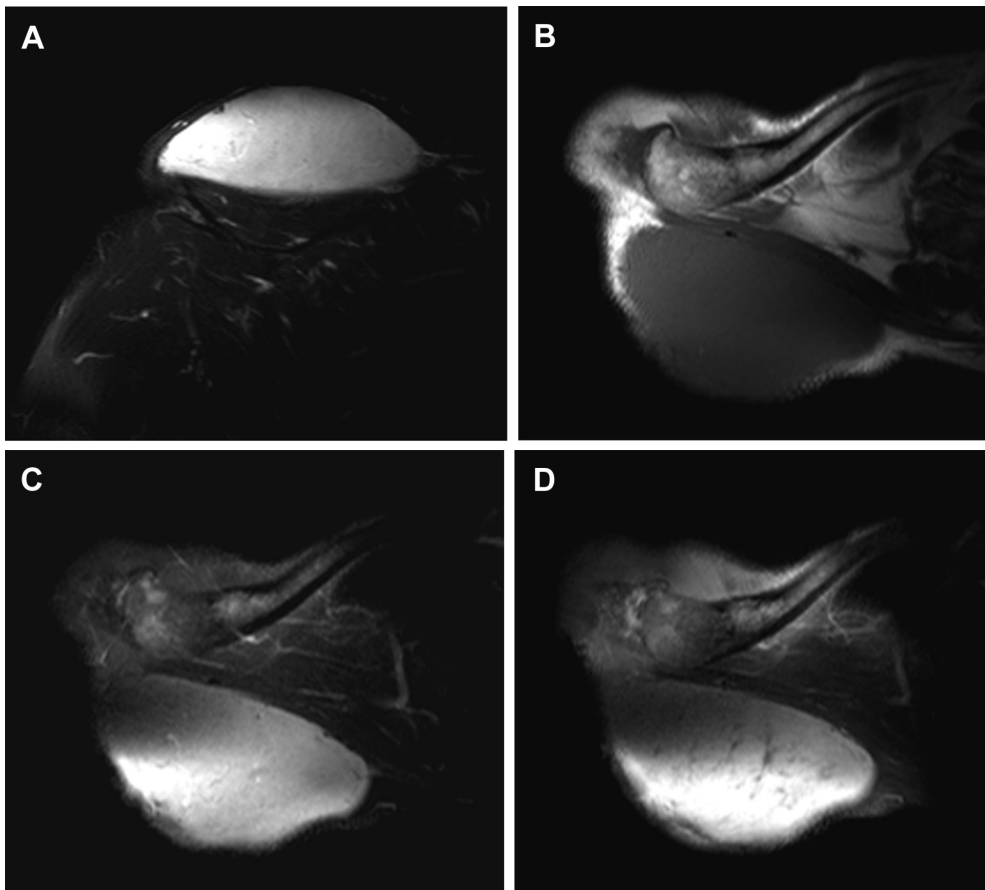


Fig. 2 – Initial presentation. (A) Coronal short-tau inversion recovery (STIR) MRI demonstrates a large hyperintense ovoid mass superior to the trapezius. (B) Axial T1-weighted MRI demonstrates a large hypointense ovoid mass superficial to the trapezius. (C) Axial T2-weighted fat-suppressed MRI demonstrates a large hyperintense ovoid mass superficial to the trapezius. (D) Axial T1-weighted fat-suppressed post-intravenous gadolinium-based contrast MRI demonstrates a large ovoid mass with homogeneous internal enhancement.

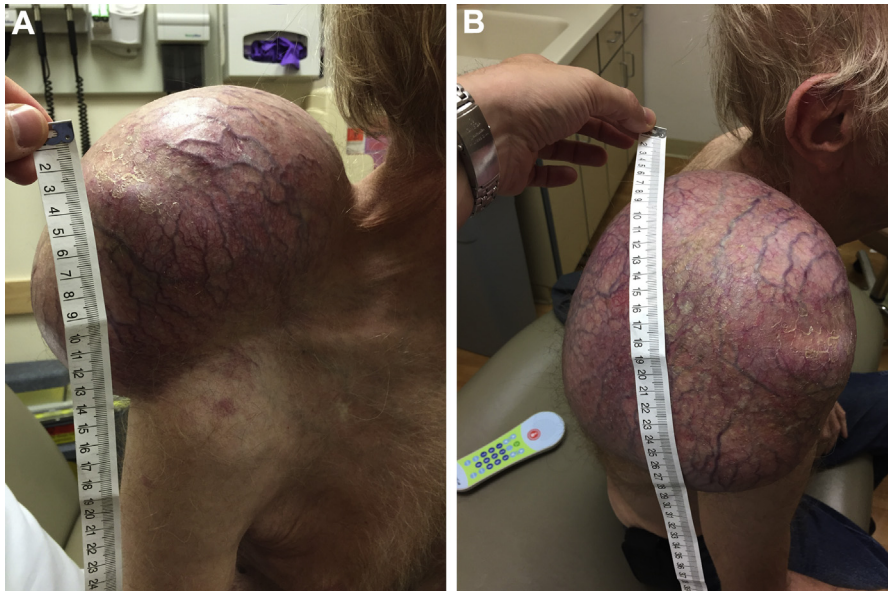


Fig. 3 – Second presentation, 3 years later. (A) Clinical photograph shows large mass involving the superior shoulder with overlying attenuated skin. (B) Clinical photograph shows large mass involving the superior shoulder with overlying attenuated skin.

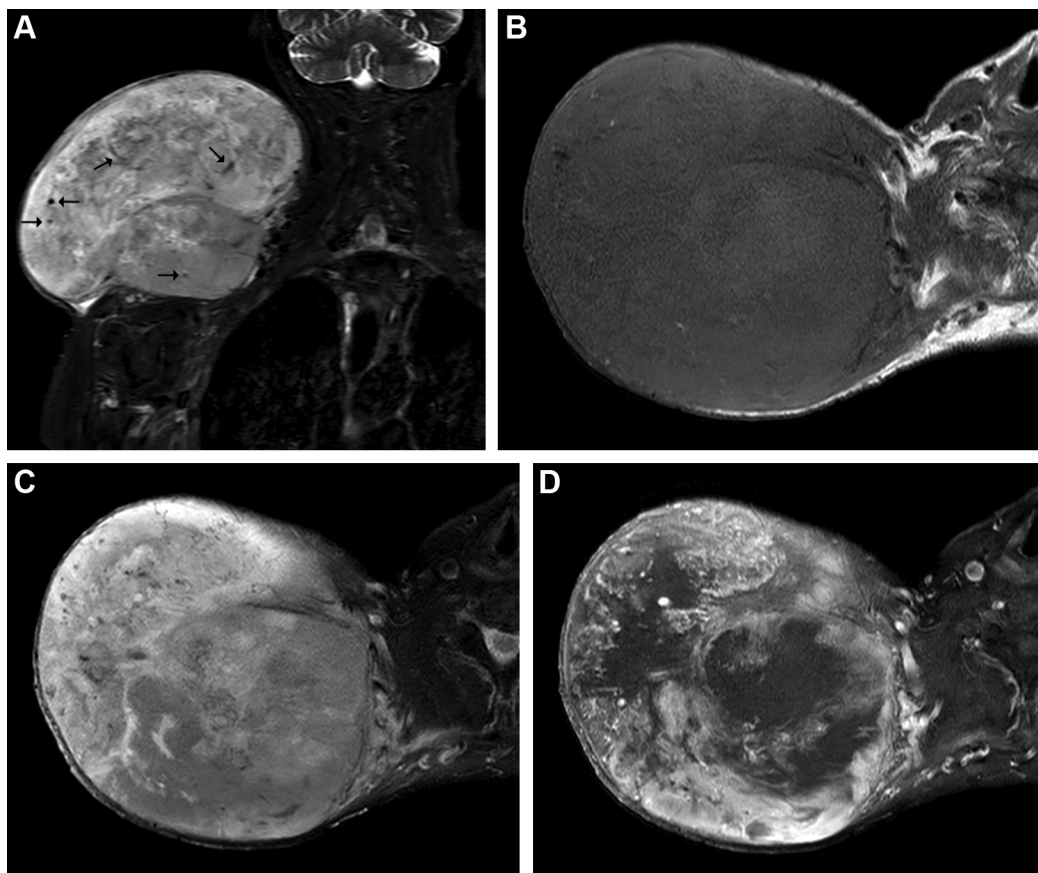


Fig. 4 – Second presentation, 3 years later. (A) Coronal short-tau inversion recovery (STIR) MRI demonstrates significant interval size increase of a heterogeneously hyperintense shoulder mass (vascular flow voids denoted by arrows). (B) Axial T1-weighted MRI demonstrates significant interval size increase of a hypointense shoulder mass. (C) Axial T2-weighted fat-suppressed MRI demonstrates significant interval size increase of a heterogeneously hyperintense shoulder mass. (D) Axial T1-weighted fat-suppressed post-intravenous gadolinium-based contrast MRI demonstrates heterogeneous enhancement of a right shoulder mass.



Fig. 5 – Second presentation, 3 years later. Intravenous iodinated contrast-enhanced CT coronal soft-tissue window shows no soft-tissue calcification or tumor matrix within the mass.

resection. Over the course of 42 days, the patient received a total dose of 5,000 cGy of radiation in 25 fractions to his right shoulder mass. A follow-up postradiotherapy MRI demonstrated a marginal increase in tumor size, with a concomitant decrease in internal enhancement, suggesting a modest positive therapeutic response with treatment-related necrosis.

After completion of radiation therapy, the mass was surgically resected by the orthopedic oncology service at our institution and weighed 4.5 kg (Figs. 7A-C). The plastic surgery

service closed the large resultant wound with a pedicled latissimus dorsi flap and split-thickness skin graft.

Discussion

Neurofibromas are benign peripheral nerve sheath tumors composed of Schwann cells, fibroblasts, and perineurial cells. Most neurofibromas are solitary and occur in individuals 20-30 years of age and without a history of neurofibromatosis, although there is a strong association between neurofibromatosis and the development of neurofibromas [1]. Specifically, nearly 100% of patients with multiple neurofibromas have neurofibromatosis type 1 (NF1) [1].

Neurofibromas are classified as either cutaneous or intraneural, with the former arising from small intradermal nerves. The plexiform neurofibroma refers to a subtype of neurofibroma with a unique pathologic growth pattern that may be grossly apparent (pedunculated dermal and subcutaneous tumors that appear thick, wormy, and baggy) or only evident on microscopic examination [2]. This specific subset of neurofibroma is considered pathognomonic for NF1, and carries a relatively high risk of malignant degeneration, occurring in up to 13% [2,3].

On MRI, neurofibromas typically appear isointense to peripheral nerves on T1-weighted imaging and demonstrate peripheral hyperintensity with central hypointense or isointense signal on T2-weighted imaging. Contrast enhancement, if present, is typically homogeneous and mild.

Malignant degeneration of a neurofibroma is often clinically manifest by a rapid increase in tumor size or sudden onset of pain or neurovascular symptoms in a previously asymptomatic patient. On average, MPNSTs measure 6 cm; this is in contrast to our case in which the massive tumor measured up to 20 cm in maximal dimension [4]. The most common location of a MPNST is the supraclavicular brachial plexus [3]. Notably, approximately 50% of MPNSTs arise in patients without NF1 [3,5].

On MRI, MPNSTs appear heterogeneously hyperintense on fluid sensitive sequences, with intense heterogeneous enhancement after the administration of gadolinium-based intravenous contrast, and exhibit other features typical of a

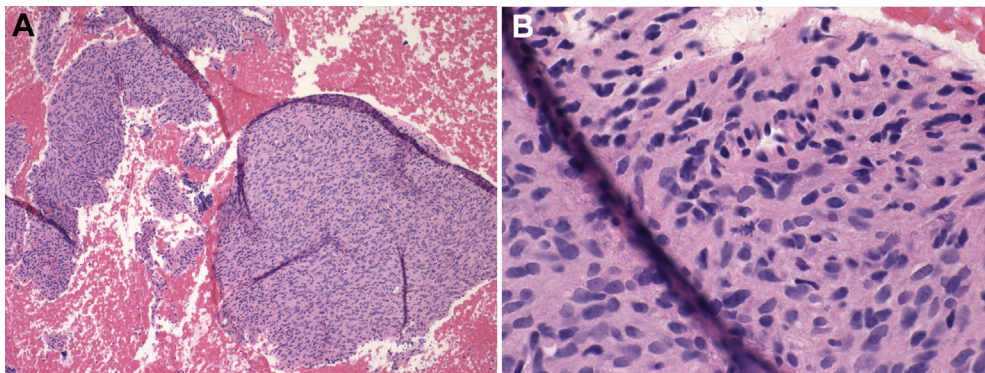


Fig. 6 – Second presentation, 3 years later. (A) Photomicrograph at 10× magnification with hematoxylin and eosin stain shows pleomorphic spindle cells arranged in a vague storiform pattern. (B) Photomicrograph at 60× magnification with hematoxylin and eosin stain demonstrates rare mitoses.

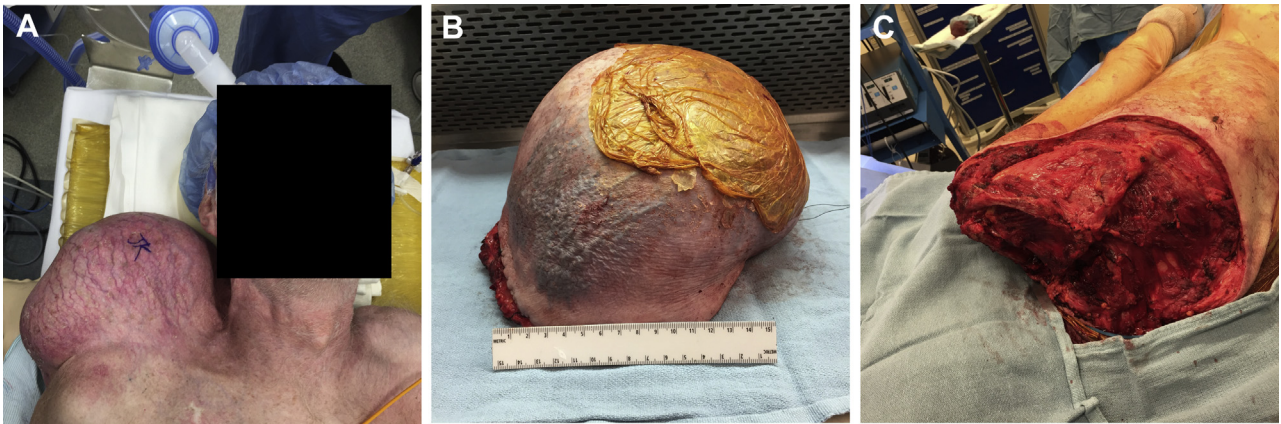


Fig. 7 – Intraoperative imaging. (A) Large mass involving the superior right shoulder with overlying attenuated skin acquired just before surgical resection. (B) Large MPNST weighing 4.5 kg. (C) Large defect at the right shoulder after resection of a massive malignant peripheral nerve sheath tumor. The defect was subsequently closed with a pedicled latissimus dorsi flap and split-thickness skin graft.

locally aggressive lesion, such as invasion of adjacent structures and indistinct margins [6].

Treatment of a MPNST is dependent on tumor location, with peripheral-extremity lesions typically requiring local excision alone. More proximal lesions may require amputation, although there is a recent trend toward limb sparing resection [3]. Adjuvant therapies such as external beam radiation, brachytherapy seeds, or chemotherapy may be performed dependent on final pathology, margins, and location. Prognosis is variable, but a review of 221 patients who underwent local resection for MPNST demonstrated a 10-year disease-specific mortality rate of 43% [7].

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