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Case Report

Radiologic manifestation of the malignant peripheral nerve sheet tumor involving the brachial plexus

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ABSTRACT

A 63-year-old African American female with history of bilateral breast cancer status after lumpectomy and radiation therapy presented with right hand, wrist, and arm pain. She was found to have a right axillary mass and a large lesion in the right brachial plexus. A biopsy of the brachial plexus mass came back as a malignant peripheral nerve sheath tumor. This case report illustrates the critical value of multiple imaging modalities in definitive diagnosis of this rare pathologic entity.

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are rare, aggressive, and have poor prognosis. These tumors' usual involvement of the sacral plexus with brachial plexus lesions occurring significantly less frequently. Patients typically present with nonspecific symptoms with imaging playing an essential role in early diagnosis. The purpose of this case report is to describe characteristic imaging findings, review the differential diagnostic considerations, and discuss the role of image-guided tissue sampling in the diagnosis of MPNST.

Case report

Patient is a 63-year-old African American female with history of bilateral breast cancer (pathology proven, invasive ductal carcinoma, tubular type) status after lumpectomy of right and left breast in 2008 and 2005, respectively. In 2009, patient presented with right wrist and hand pain. Initially, her symptoms were attributed to mild osteoarthritis of the interphalangeal joints seen on the radiograph and was treated conservatively without significant improvement. Further evaluation revealed a possibility of cubital tunnel syndrome for which the patient underwent a surgical release with no

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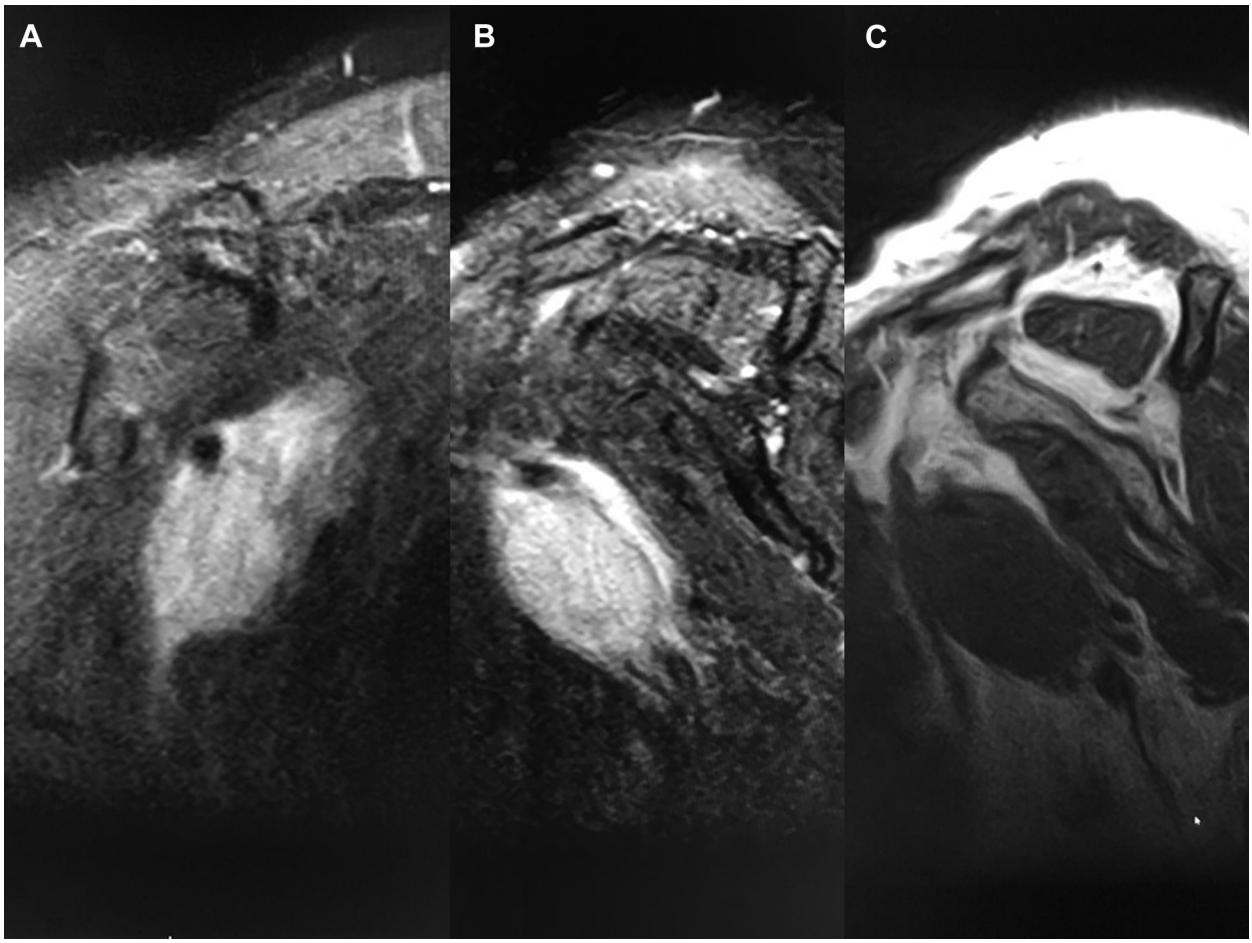

Fig. 1 – The noncontrast magnetic resonance imaging (MRI) of the shoulder demonstrate heterogenous, solid mass with hyperintense signal on sagittal (A), and coronal fat-saturated T2-weighted (B) sequences and hypointense signal on T1-weighted sequence (C).

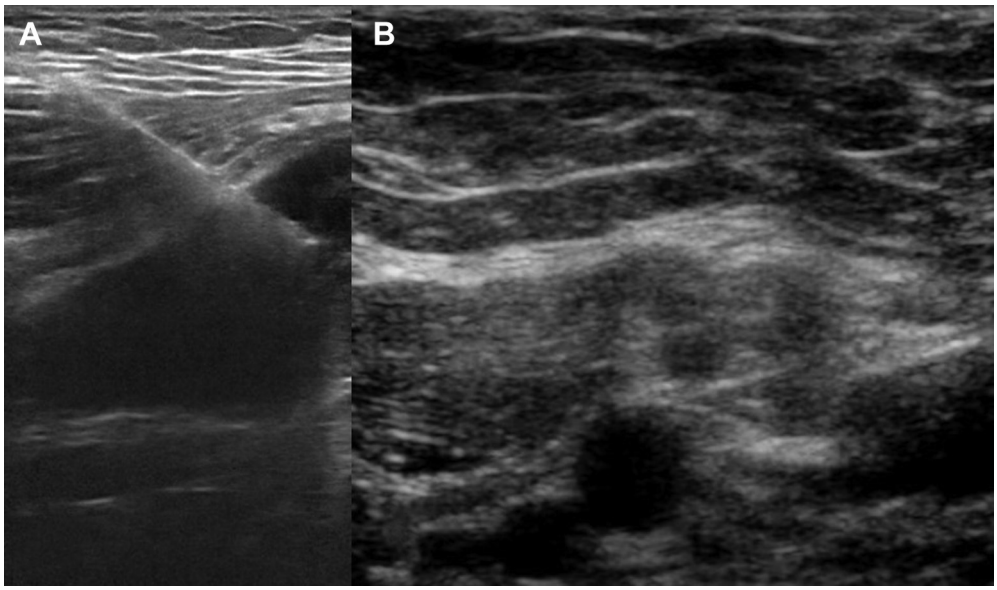

Fig. 2 – Image (A) demonstrates an ultrasound-guided biopsy. The mass appears heterogeneously hyperechoic on ultrasound (B).

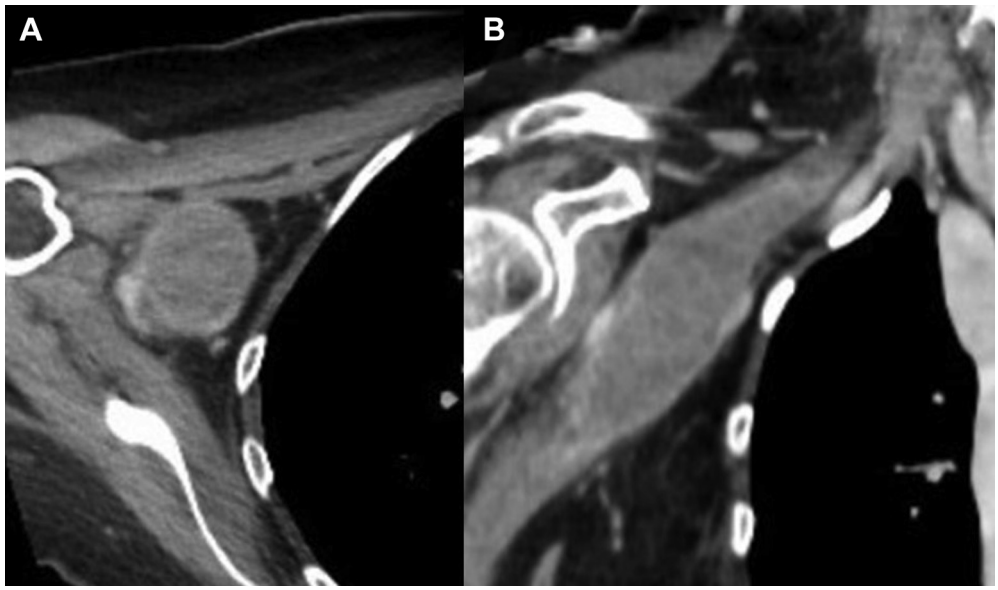

Fig. 3 – Postcontrast axial (A) and coronal (B) images show heterogeneously enhancing solid mass involving the brachial plexus and splaying the vessels.

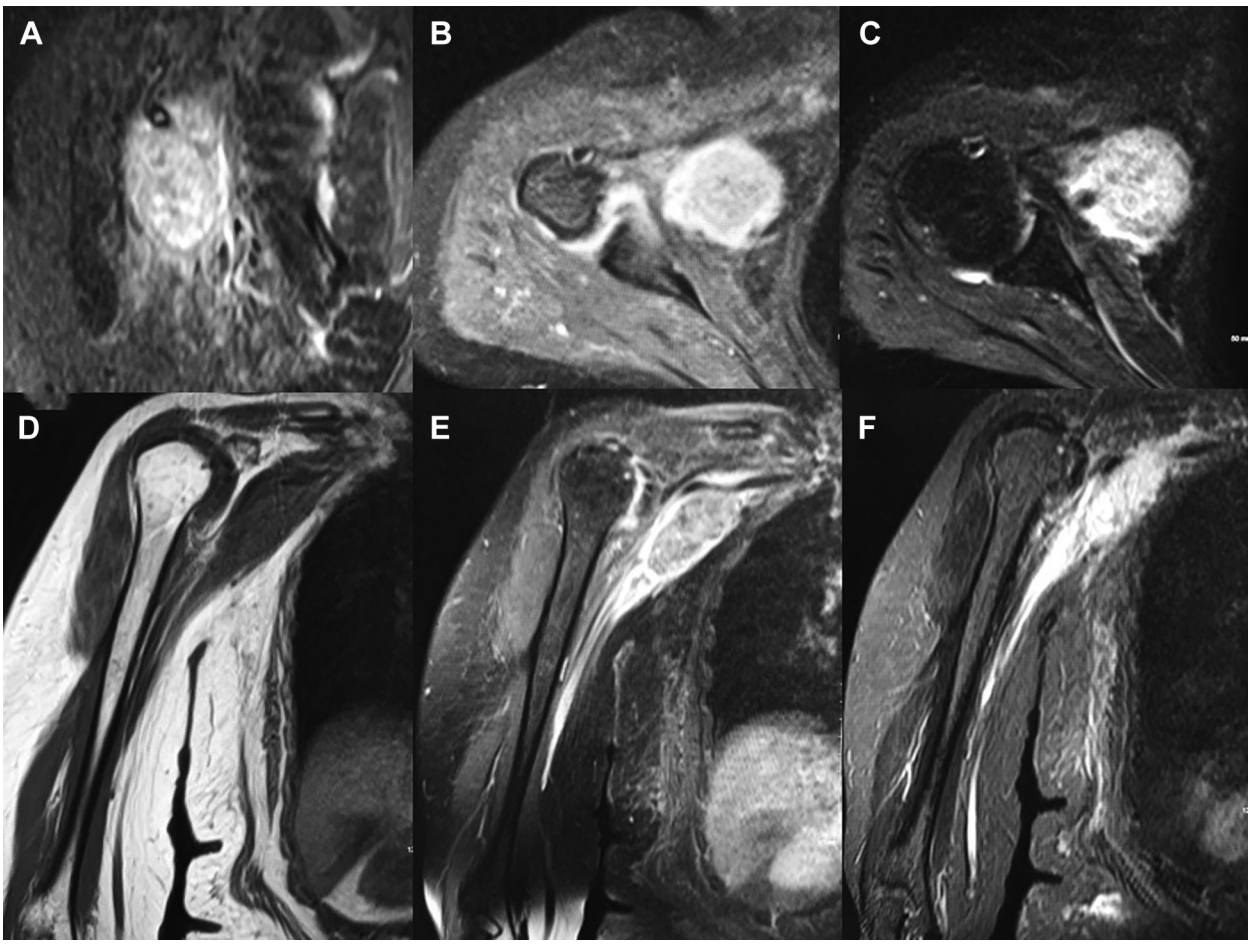

Fig. 4 – MRI images of the shoulder with and without gadolinium were obtained at multiple sequences and various planes. The sagittal short T1 inversion recovery (STIR) (A), axial T1 fat-saturated postcontrast (B), axial T2 fat-saturated (C), coronal T1 (D), coronal T1 postcontrast fat-saturated (E), and coronal STIR (F) sequences are demonstrated. The malignant peripheral nerve sheath tumor appears as a heterogenous solid mass with T1 hypointense, T2 hyperintense, and avid enhancement on postcontrast images.

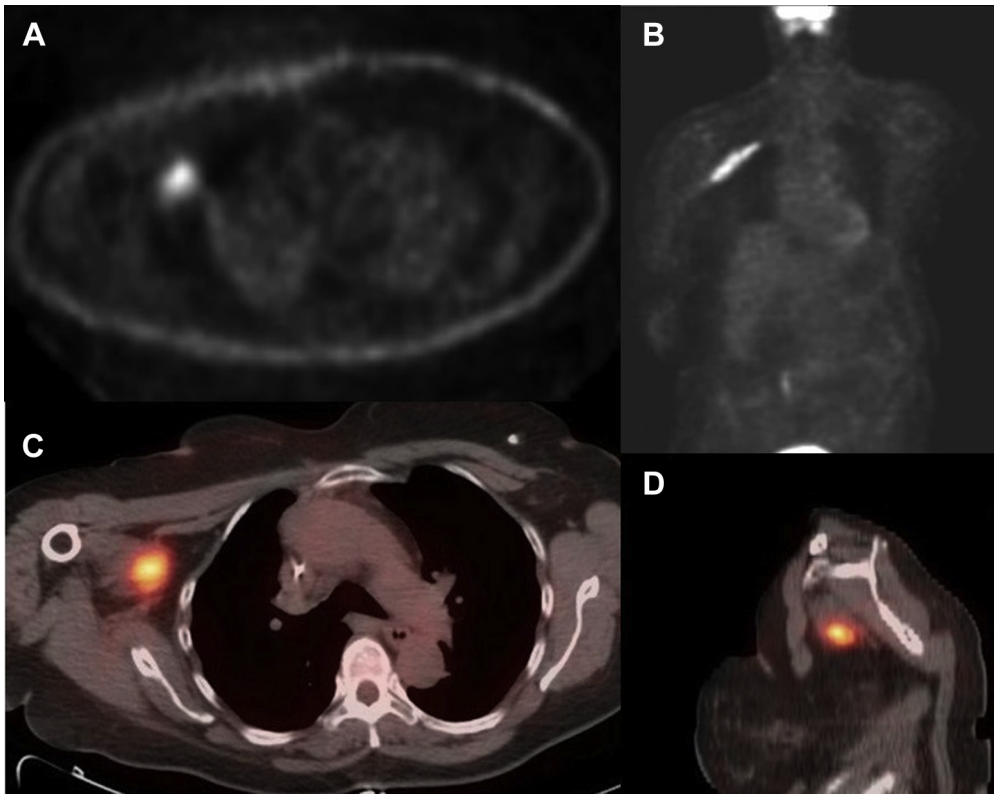

Fig. 5 – The axial nonattenuated corrected (A), coronal attenuated corrected (B), axial fused positron emission tomography-computed tomography (PET-CT) (C), and sagittal fused PET-CT (D) images demonstrated a very hypermetabolic mass involving the right brachial plexus.

appreciable symptomatic amelioration. Later, the patient developed neck and right upper extremity pain and weakness, which prompted radiographic and magnetic resonance imaging (MRI) evaluation of the cervical spine, shoulder, and brachial plexus.

Although the cervical spine MRI showed degenerative cervical spondylosis, the shoulder and brachial plexus MRI revealed a heterogeneously enhancing, T1 hypointense and T2 hyperintense soft tissue mass within the axilla and along the right brachial plexus. The lesion involved the neurovascular bundle displacing adjacent vascular structures and measuring 2.4 × 3.9 × 5.5 cm (Fig. 1).

This finding prompted an ultrasound-guided biopsy of the right axillary mass, which revealed the diagnosis of MPNST (Fig. 2). Staging computed tomography (CT) of the chest, abdomen, and pelvis did not identify any additional sites of metastatic disease. On CT, the mass was characterized as heterogeneously enhancing solid mass involving the right brachial plexus, splaying the vessels (Fig. 3). A follow-up MRI of the shoulder was obtained to evaluate extension of brachial plexus involvement which remonstrated the mass as shown in Figure 4.

Positron emission tomography/CT showed an fludeoxyglucose (FDG) avid heterogenous mass in the right brachial plexus with standardized uptake values of 9.49, consistent with known MPNST (Fig. 5).

The treatment plan was quickly developed and included surgical management after neoadjuvant chemoradiation to decrease the risk of neurovascular damage. Eventually, the patient's right arm was amputated.

Discussion

MPNSTs are rare with reported incidence of 0.001%-0.1% [1]. Nearly, 50% of MPNSTs are found in patients with neurofibromatosis type 1 [2]. Given our patient's history of malignancy, a connection between breast malignancy and MPNST was questioned. According to Redzepagic et al [3], no clear association has been reported in literature between MPNST and breast cancer. However, radiation therapy has been shown to be a risk factor for the development of MPNST, which was present in our case [4].

Histologically, MPNSTs may be further subdivided into neurofibrosarcoma, neurogenic sarcoma, malignant schwannoma, and malignant neurilemmoma because of their ectomesenchymal origin [4]. These tumors tend to originate in the nerve roots of the sacral plexus, with less than 6% arising from the brachial plexus [5–7].

Radiologically, these tumors have a nonspecific but aggressive appearance. Sonographically, these lesions typically appear heterogeneously echogenic with posterior

acoustic enhancement. On MRI, they tend to be T1 isointense to muscle and hyperintense on fluid sensitive sequences. As is characteristic of many malignant tumors, MPNSTs enhance avidly on postcontrast CT and MRI, and have increased metabolic activity on Tc-99m and F-18 FDG positron emission tomography imaging [6,8]. Definitive diagnosis, however, requires tissue sampling that is commonly performed under imaging guidance.

The signs and symptoms are also nonspecific, usually including pain, palpable mass, numbness, and paresthesia, which tends to be progressive as opposed to insidious, as in this case [3,6]. Tumor complications include neurovascular, soft tissue and osseous invasion, and metastasis. Morphologically, they have irregular borders and tend to be larger than 5 cm [7].

Differential consideration for an enhancing, FDG avid, mass within the brachial plexus include metastases, primary nerve sheath tumors (benign or malignant), various sarcomas, radiation fibrosis, and neurolymphomatosis [9]. Metastases tend to have lower signal intensity on T2 relative to soft tissue and occur via direct spread (ie, Pancoast tumors) or by lymphohematogenous means (breast, lung or head, and neck squamous cell carcinoma) [10]. Radiation fibrosis exhibits low T1 and T2 signal intensity with variable to low enhancement. Benign and malignant nerve sheath tumors have greater overlap in appearance; however, malignant nerve sheath tumors tend to be larger, with irregular margins and heterogeneous enhancement [10,11].

Treatment includes surgical excision with wide margins. Regional nodal resection is not indicated because of hematogenous spread of this malignancy. Other treatment options include radiation, chemotherapy, and/or limb amputation.

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