Axillary Malignant Peripheral Nerve Sheath Tumor With Aggressive Clinical Course: A Case Report

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AIM: Malignant peripheral nerve sheath tumor (MPNST) involvement of the axillary region is exceptionally uncommon and poses significant diagnostic and therapeutic challenges.

CASE PRESENTATION: We report the case of a 41-year-old woman who presented with a left axillary mass initially detected during pregnancy. Imaging revealed a large axillary lesion closely associated with neurovascular structures. Histopathological and immunohistochemical analyses supported an MPNST diagnosis. Despite multimodal management, including partial surgical resection, combination chemotherapy with ifosfamide/doxorubicin-based regimens, radiotherapy, repeated surgical interventions, and reconstructive procedures, the disease demonstrated aggressive behavior with multiple local recurrences, nodal and cutaneous metastases, and an eventual mass in the gluteal region.

RESULTS: MPNSTs account for 3–10% of soft tissue sarcomas and are frequently associated with poor outcomes, particularly in tumors larger than 5 cm, high-grade lesions, and those associated with neurofibromatosis type 1. Diagnosis relies on a combination of histopathology, immunohistochemistry, and advanced imaging modalities. Complete surgical resection with negative margins remains the only potentially curative treatment; however, this is often not feasible due to tumor location and extent. Radiotherapy may improve local control, while chemotherapy has limited efficacy in advanced disease.

CONCLUSIONS: Axillary MPNST is an exceedingly rare entity with an aggressive clinical course and poor prognosis. This case highlights the challenges in management and underscores the need for early diagnosis, aggressive multimodal treatment, and the development of more effective systemic and targeted therapies.

Keywords: malignant peripheral nerve sheath tumor; axillary; neurogenic tumor; case report

Introduction

Malignant peripheral nerve sheath tumors (MPNSTs) are rare, highly aggressive mesenchymal sarcomas characterized by rapid growth, a high rate of local recurrence, and significant resistance to chemotherapy, which together make management particularly challenging [1,2]. These tumors most frequently occur in the extremities (approximately 45%), followed by the trunk (34%) and the head and neck region (19%) [3,4]. In contrast, involvement of the brachial plexus is uncommon [5].

Definitive diagnosis of MPNSTs often requires histopathological and immunohistochemical evaluation. Management remains challenging due to high recurrence rates, metastatic potential, and limited responsiveness to systemic therapy.

Clinically, patients with MPNST typically present with a rapidly enlarging mass, pain, or progressive neurological symptoms resulting from nerve compression or invasion. Because imaging findings may overlap with those of benign peripheral nerve sheath tumors, diagnosis is often delayed and requires correlation of radiological, histopathological, and immunohistochemical findings. Current treatment strategies rely primarily on complete surgical excision with negative margins whenever feasible. Radiotherapy is frequently used to improve local disease control, whereas chemotherapy may be considered in selected patients with advanced, unresectable, or metastatic disease, although therapeutic responses remain limited.

MPNSTs arising in the axillary region are exceedingly rare. This report describes a case of axillary MPNST in the context of previously reported cases.

Case Presentation

A 41-year-old woman first noted a mass in her left axilla during pregnancy in 2020 but did not seek medical evaluation at that time. She subsequently delivered by cesarean section. Approximately one month postpartum, clinical evaluation and imaging were performed. Imaging examinations, including contrast-enhanced CT, contrast-enhanced MRI, and 18F-FDG PET/CT, were performed to evaluate tumor extent, local invasion, and possible metastatic le-

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sions. Physical examination revealed a firm, mobile mass measuring approximately 5 × 5 × 5 cm in the left axillary region. Thoracic computed tomography demonstrated a lesion measuring 77 × 88 mm in the left axilla, characterized by mild peripheral enhancement and areas of fluid attenuation (Fig. 1).



Fig. 1. Preoperative contrast-enhanced computed tomography of the thorax demonstrating a large left axillary soft-tissue mass. The arrow indicates the left axillary mass. The lesion appears relatively well circumscribed, measuring approximately 77 × 88 mm, with heterogeneous attenuation and areas of cystic degeneration. The mass occupies the left axillary compartment and displaces adjacent soft tissues, with no evidence of calcification.

Contrast-enhanced magnetic resonance imaging (MRI) of the left shoulder revealed a relatively well-defined mass measuring approximately 8.5 × 7.5 cm in the anterior aspect of the left axillary region. The lesion caused marked compression of adjacent neural structures and surrounding vascular elements (Fig. 2).

Histopathological evaluation of a tru-cut biopsy obtained from the a mass in the left axilla revealed areas of necrosis and focal fibroadipose tissue. The patient was subsequently referred to a university hospital, where surgical intervention was undertaken. Partial tumor resection was performed due to neurovascular involvement. Intraoperatively, a cystic mass measuring approximately 10 cm was identified. Complete excision was not achievable because of the lesion's proximity to the axillary vascular structures. Based on the available operative information from the referring center, the lesion appeared intimately associated with major axillary vascular structures and adjacent brachial plexus elements, limiting the feasibility of safe radical excision. Detailed information regarding the specific neurovascular structures involved and whether vascular reconstruction was considered was not available in the operative records. Consequently, partial resection was performed, accompanied by drainage of the cystic contents during dissection. Additionally, enlarged lymph nodes measuring 2–3 cm were dissected from the surrounding tissue and excised en bloc with the specimen.

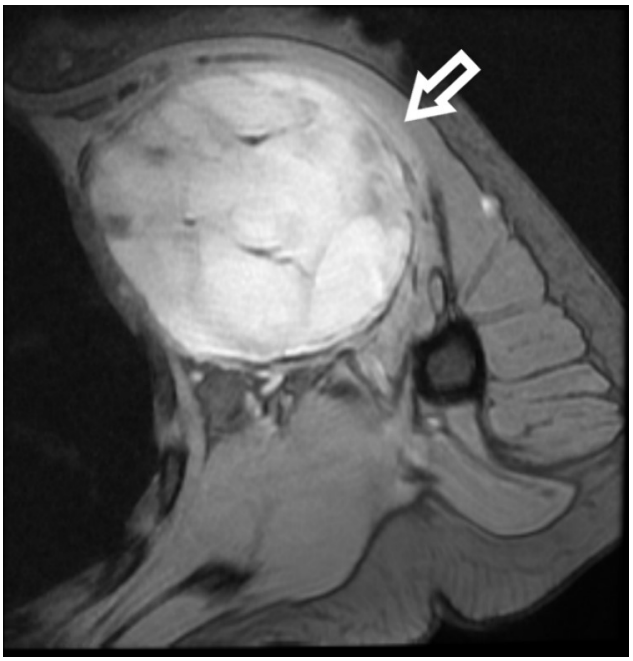


Fig. 2. Preoperative contrast-enhanced magnetic resonance imaging of the left axillary region. The arrow indicates a large heterogeneous mass measuring approximately 8.5 × 7.5 cm. The lesion demonstrates mixed signal intensity with internal cystic-necrotic areas and causes compression of adjacent neurovascular structures within the axilla.

Liquid-based cytological analysis using centrifugation microscopy demonstrated features suspicious for malignancy. Immunohistochemical evaluation, performed with a panel of 16 markers, revealed tumor cell positivity for S-100, vimentin, and SRY-box transcription factor 10 (SOX10), along with weak positivity for transducin-like enhancer of split 1 (TLE1). The neoplastic cells were negative for actin, desmin, human melanoma black 45 (HMB45), Melan-A, cluster of differentiation (CD)68, pancytokeratin, CD34, CD21, CD23, CD35, and anaplastic lymphoma kinase (ALK). The Ki-67 proliferation index was estimated at approximately 20%. Overall pathological findings were consistent with a malignant mesenchymal neoplasm, accompanied by reactive lymphoid hyperplasia in the axillary lymph nodes. Based on the combined histomorphological and immunohistochemical features, MPNST was considered the most likely diagnosis.

Following pathological diagnosis, the patient was scheduled to receive combination chemotherapy consisting of ifosfamide and doxorubicin hydrochloride, with mesna administered for uroprotection. Enoxaparin sodium, antiemetic agents, analgesics, and pantoprazole sodium were administered as supportive medications as clinically indicated. The chemotherapy regimen was administered over three consecutive days per cycle. Subsequently, adjuvant radiotherapy was administered (total dose 66 Gy). The patient underwent adjuvant external beam radiotherapy. A

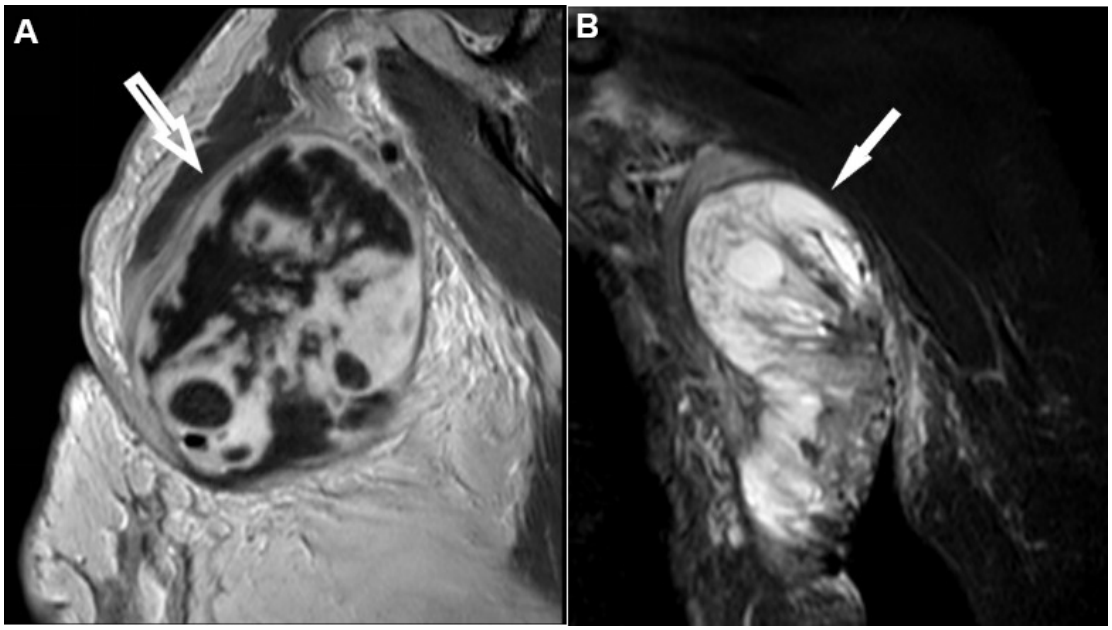


Fig. 3. Post-treatment magnetic resonance imaging (MRI) of the left axillary region following chemotherapy and radiotherapy. (A) Axial T1-weighted contrast-enhanced image demonstrating a heterogeneous soft-tissue mass in the left axilla (arrow), with irregular enhancement and internal low-signal areas corresponding to cystic-necrotic degeneration. (B) Coronal T2-weighted image showing a residual/recurrent tumor measuring approximately 90 × 66 mm (arrow), characterized by heterogeneous high signal intensity and prominent cystic-necrotic components. The lesion extends toward the serratus anterior musculature and remains in proximity to the axillary artery and vein, with partial circumferential encasement of the adjacent vascular structures.

total radiation dose of 6600 Gy was delivered, consisting of 5000 Gy administered in daily fractions of 200 Gy using 6 MV X-ray beams generated by a linear accelerator, followed by a boost dose of 1600 Gy directed to the residual mass in the left axillary region. Persistent residual/recurrent disease was detected in the follow-up imaging. Post-radiotherapy magnetic resonance imaging (MRI) revealed a mass lesion in the left axillary region measuring approximately 90 × 66 mm at its greatest dimension. The lesion was hypointense on T1-weighted images and mildly hyperintense on T2-weighted images, with heterogeneous contrast enhancement and prominent cystic–necrotic components. Extension toward the serratus anterior muscles was also observed (Fig. 3A,B). The lesion was in proximity to the axillary artery and vein, with evidence of circumferential extension around these vascular structures.

Thoracic computed tomography of the left axillary region demonstrated a mass measuring 66 × 49 mm, characterized by central cystic–necrotic components, irregular margins, and soft-tissue density. The lesion was also associated with disruption of the anterior skin and subcutaneous fat planes (Fig. 4A,B).

Positron emission tomography–computed tomography (PET/CT) demonstrated increased fluorodeoxyglucose (FDG) uptake within the irregular soft-tissue mass, with a maximum standardized uptake value (SUVmax) of 10.4, decreased from the prior value of 27.4. FDG uptake in the remaining imaged regions was within physiological limits. Compared with the previous PET/CT examination, these findings were consistent with partial metabolic regression.

Despite this multimodal treatment, the patient was subsequently readmitted to the oncology clinic because of recurrence of the axillary mass. Despite this multimodal approach, she was subsequently readmitted to the oncology clinic with recurrence of the axillary mass, accompanied by arm contracture that limited abduction. Biopsies obtained from the recurrent axillary mass demonstrated histopathological features consistent with malignant peripheral nerve sheath tumor (MPNST), epithelioid type, compatible with schwannian differentiation.

Immunohistochemical analysis (16 markers in total) demonstrated positivity for S-100, vimentin, and CD99, while negative staining was observed for pan-cytokeratin (PanCK), epithelial membrane antigen (EMA), Melan-A, HMB45, CD138, actin, desmin, myogenin, CD68, CD30, CD20, CD3, and CD5.

During chemotherapy, thoracic computed tomography of the left axillary region demonstrated a cavitary lesion with a thin wall and central necrosis, measuring approximately 30 × 16 mm. Additionally, multiple semisolid mass lesions were identified in the left breast, the largest measuring approximately 26 mm. These lesions were characterized by irregularly contoured soft-tissue components associated with thin-walled cavitary structures (Fig. 5A,B). The breast lesions were considered separate benign lesions rather than metastatic breast lesions.



Fig. 4. Recurrent left axillary MPNST on thoracic computed tomography. (A) Axial contrast-enhanced image demonstrating an irregular soft-tissue mass with central cystic-necrotic degeneration (arrow). (B) Coronal reconstruction showing the extent of the recurrent lesion within the axillary region and its close relationship to surrounding soft tissues. The lesion demonstrates irregular margins and disruption of adjacent subcutaneous tissue planes. MPNST, malignant peripheral nerve sheath tumor.

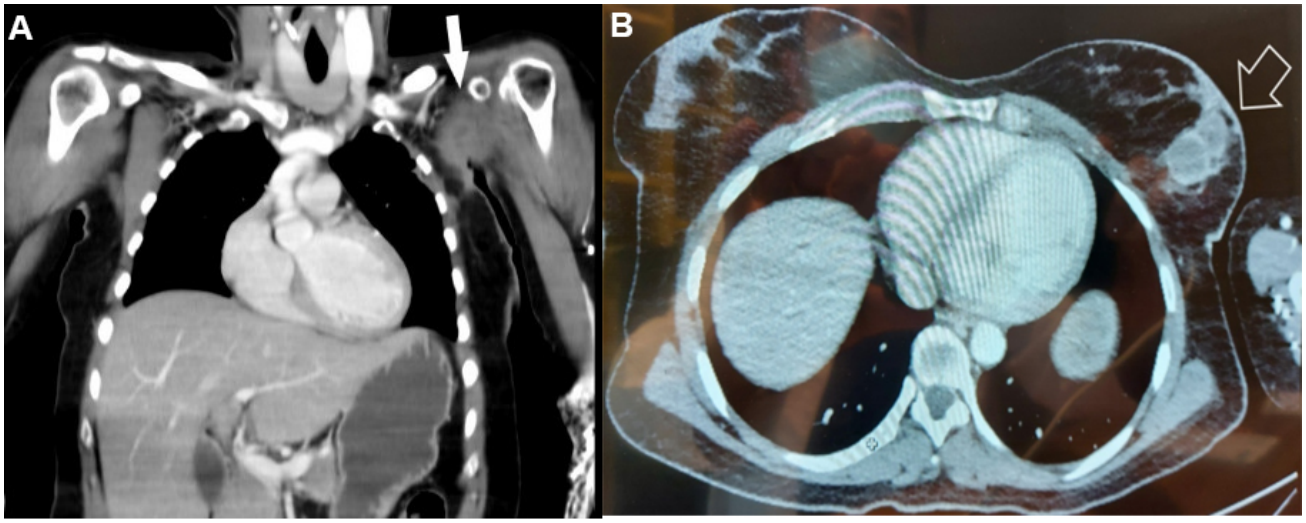


Fig. 5. Preoperative thoracic computed tomography demonstrating recurrent disease. (A) Coronal image showing recurrent tumor involvement in the left axillary region (arrow), characterized by cavitary changes and central necrosis. (B) Axial image demonstrating multiple irregular semisolid lesions within the left breast (open arrow), representing contiguous local tumor extension rather than distant breast metastases.

At our institution, modified radical mastectomy with axillary tumor excision and Limberg flap reconstruction was performed. Intraoperatively, a cavitary defect was identified in the axillary region, with the tumor fixed to the thoracic wall and encasing the axillary vascular structures. Reconstruction of the axillary defect was performed using a Limberg flap. Owing to significant axillary contracture, the dissection was technically challenging.

Histopathological examination of the mastectomy specimen revealed multiple gray–white nodular lesions, the largest measuring 2.6 cm, consistent with plexiform schwannoma. Additionally, a gray–brown mass measuring 3.5 × 2.5 × 1.5 cm excised from the axillary tissue was diagnosed as MPNST (Fig. 6A–C).

Postoperatively, the patient continued systemic chemotherapy with Ifosfamide/Mesna/Adriamycin (IMA) in combination with gemcitabine. Eight months after the initial axillary tumor resection, while the patient was undergoing chemotherapy, wound recurrence was confirmed by biopsy; repeat tumor excision and Z-plasty reconstruction were performed. The axillary wound dehisced, and a biopsy obtained from the involved tissue demonstrated histopatho-

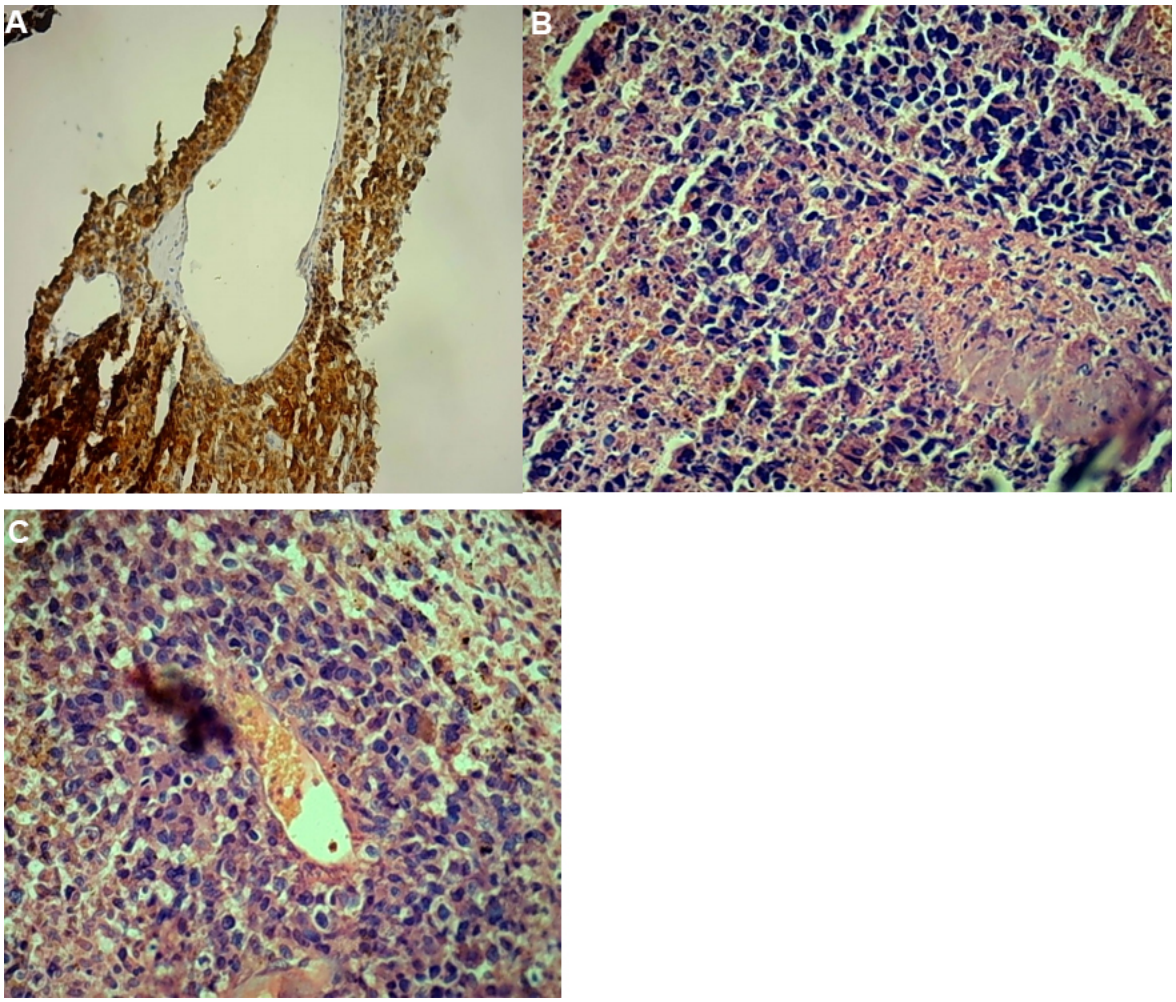


Fig. 6. Histopathological and immunohistochemical findings of the tumor. (A) Immunohistochemical staining demonstrating S-100 positivity in spindle- to oval-shaped atypical tumor cells (×200). (B,C) Histopathological examination demonstrating spindle- to oval-shaped atypical tumor cells on hematoxylin and eosin (H&E) staining (×400).

logical findings consistent with MPNST. The patient subsequently underwent reoperation, during which the recurrent axillary tumor was excised and the resulting defect reconstructed using a Z-plasty technique. Pathological examination of the resected specimen again confirmed the diagnosis of MPNST.

Five months after the repeat surgery, rapid local recurrence with suspected nodal and cutaneous metastases, as well as gluteal metastasis, was detected. The patient experienced rapid recurrence of the tumor in the axillary region, accompanied by nodal and cutaneous metastases, as well as distant metastasis to the gluteal region. Magnetic resonance imaging of the hip demonstrated a mass measuring approximately 43 × 91 × 96 mm within the right gluteus medius muscle. The lesion appeared heterogeneously hyperintense on T2-weighted sequences and hyperintense on T1-weighted sequences (Fig. 7).

During chemotherapy, the patient experienced respiratory deterioration, poor treatment tolerance and severe fatigue.

This case has been reported in line with the Case Report (CARE) Guidelines to ensure the accuracy and completeness of the report (**Supplementary Material**).

Discussion

MPNST was defined by the World Health Organization in 2013 as a malignant neoplasm that originates either from a peripheral nerve, from a pre-existing benign nerve sheath tumor, or in the setting of neurofibromatosis type 1 (NF1). When these defining clinical contexts are absent—particularly in sporadic de novo cases or tumors arising after radiotherapy—the diagnosis relies on the integration of histopathological, immunohistochemical, and ultrastructural findings that support Schwann cell differentiation [4,6].

MPNST is a rare and highly aggressive sarcoma of the peripheral nerve sheath, accounting for approximately 3–10% of all soft tissue sarcomas [7]. A summary of previously reported axillary region MPNST cases, including demographic characteristics, tumor features, and treatment approaches, is presented in Table 1 (Ref. [4,5,7,8,9,10]).

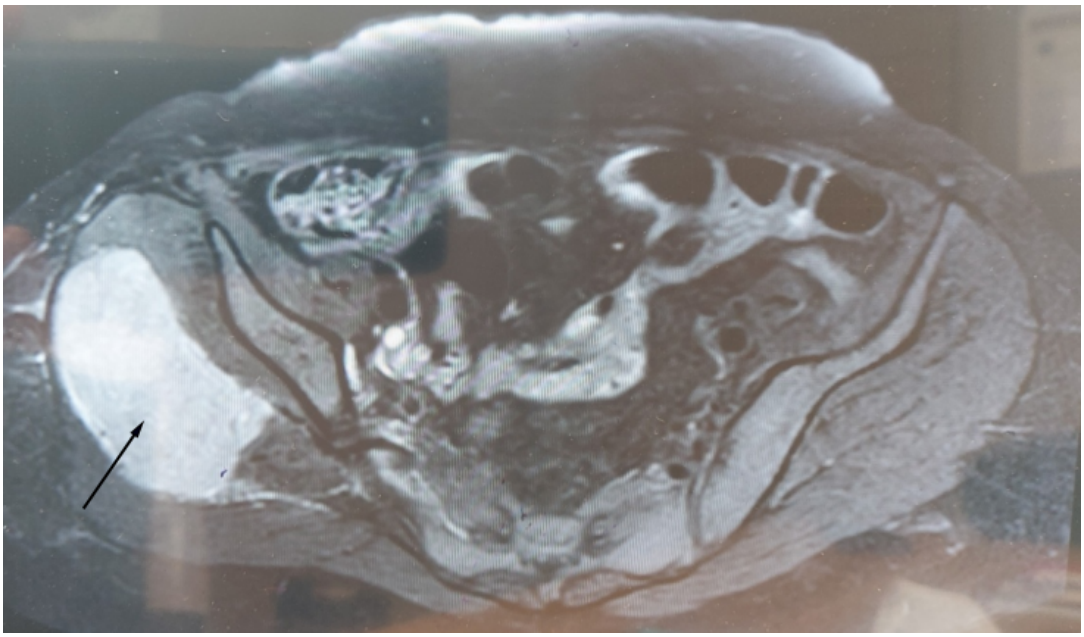


Fig. 7. Magnetic resonance imaging of the pelvis demonstrating a mass in the right gluteal region. The arrow in indicates the metastatic mass in the right gluteal region.

Comparison with previously reported axillary MPNST cases demonstrates several noteworthy differences. Most published patients were older adults (45–82 years), whereas our patient was relatively young at 41 years of age. Similar to the cases reported by Aran et al. [5] and Kusumoto et al. [7], the tumor showed close association with major neurovascular structures, which significantly limited the possibility of complete surgical excision. However, unlike most previously reported cases that were managed with a single definitive operation, our patient experienced multiple local recurrences despite repeated surgery, chemotherapy, and radiotherapy. Furthermore, the disease demonstrated an unusually aggressive clinical course with nodal involvement, cutaneous metastases, and subsequent distant metastasis to the gluteal region. Although local recurrence and distant spread are well-recognized features of MPNST, the combination of repeated recurrences, extensive local progression, and distant metastatic dissemination observed in the present case appears more aggressive than in most previously reported axillary cases. These findings further emphasize the poor prognosis associated with large, incompletely resectable axillary MPNSTs involving critical neurovascular structures. These tumors most commonly arise in the extremities—particularly in proximal locations—followed by the trunk and the head and neck region. The majority originate from major nerve trunks, most frequently the sciatic nerve. Clinically, patients often present with a painful or rapidly enlarging mass, which may be accompanied by neurological deficits due to nerve involvement [11].

The differential diagnosis of epithelioid MPNST includes metastatic carcinoma, malignant melanoma, and clear cell sarcoma. Accurate diagnosis relies primarily on detailed histopathological evaluation supported by immunohistochemical analysis and, when necessary, comparative genomic hybridization to identify characteristic chromosomal abnormalities [7].

Neurofibromatosis type 1 (NF1), also known as von Recklinghausen disease, is an autosomal dominant genetic disorder affecting approximately 1 in 3500 newborns. It results from germline inactivation of the NF1 tumor suppressor gene located on chromosome 17q11.2, with nearly 50% of cases arising from de novo mutations. NF1 is characterized by a wide spectrum of clinical manifestations, including pigmented skin lesions (such as café-au-lait macules, Lisch nodules, and axillary freckling), skeletal dysplasias, learning disabilities, and an increased predisposition to various neoplasms, including optic pathway gliomas, high-grade astrocytic tumors, and pheochromocytomas.

Patients with NF1 have a substantially increased risk of developing soft tissue sarcomas compared with the general population, and MPNST represents the most common malignancy associated with this condition. The lifetime risk of developing MPNST in individuals with NF1 is estimated to be approximately 5–10%, compared with about 0.001% in the general population. Multiple studies have demonstrated that NF1-associated MPNSTs are associated with significantly poorer outcomes compared with sporadic cases [11]. Reported outcomes for malignant peripheral nerve sheath tumor indicate a poor prognosis, with a 5-year overall survival rate of approximately 49% and a 5-year event-free survival rate of 37%. Local recurrence occurs in about 38% of cases, and nearly 39% of patients develop distant metastases, most commonly involving the lungs [8].

Table 1. Reported cases of malignant peripheral nerve sheath tumors (MPNSTs) involving the axillary region.

Author (Ref.)	Age / Sex	Location	Size	Findings	Treatment
Val-Bernal JF et al. [4]	45 / Female	Right axilla & breast	14.5 cm	Ovoid, high-density mass adjacent to pectoral muscle, no calcifications.	Total mastectomy with removal of pectoralis major, minor, and 2 axillary LNs
Shima Aran et al. [5]	63 / Female	Right axilla	2.4 × 3.9 × 5.5 cm	Soft tissue mass involving neurovascular bundle; prior bilateral breast cancer	Neoadjuvant chemoradiation; right arm amputation due to extent
Shuayb and Begum [10]	16 / Female	Left breast/ axillary region	Multiple lesions up to 11 × 11 × 6 cm	Recurrent malignant phyllodes tumor with mesenchymal features	Modified radical mastectomy, chemotherapy (IVA [ifosfamide, vincristine, and actinomycin D] protocol), 50 Gy radiotherapy with a 10 Gy boost
Kusumoto et al. [7]	70 / Male	Left axilla	12–21 cm	Epithelioid MPNST (vimentin, S-100, NSE positive); rapid growth	Surgical excision with lymphadenectomy; patient died 11 weeks post-op
Zhou et al. [8]	60 / Female	Left axilla	5.7 × 5.7 × 7.7 cm	Spindle cells in staggered bundles (H&E ×10)	Complete surgical excision
Na and Kim [9]	82 / Male	Right axilla	13.2 × 12.5 × 16.7 cm	High-grade MPNST (recurrence of undifferentiated pleomorphic sarcoma)	Palliative wide excision

LNs, lymph nodes; NSE, neuron-specific enolase.

Primary malignant peripheral nerve sheath tumor is characterized microscopically by densely cellular proliferations of spindle-shaped or epithelioid cells exhibiting nuclear atypia and increased mitotic activity. Tumor cells are often arranged in intersecting fascicles or, less commonly, in a herringbone pattern. Epithelioid variants typically demonstrate strong immunohistochemical positivity for S-100 protein, vimentin, and neuron-specific enolase (NSE) [7]. MPNSTs show a strong association with neurofibromatosis type 1 (NF1), primarily due to loss of neurofibromin, which results in activation of the RAS/MAPK/mTOR signaling pathways. Molecular studies have highlighted significant biological heterogeneity among MPNSTs, including aggressive variants such as malignant triton tumors, which are characterized by rhabdomyoblastic differentiation and are associated with a particularly poor prognosis [8].

Definitive diagnosis of MPNST relies on a combination of histopathological examination and immunohistochemical profiling. For early detection, magnetic resonance imaging (MRI) and tissue biopsy should be performed promptly when malignant transformation is suspected. However, differentiating MPNSTs from benign plexiform neurofibromas on MRI can be challenging, as both entities may demonstrate low-to-intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images with heterogeneous enhancement. Patients with MPNSTs generally have giant masses, peripheral enhancement patterns, perilesional edema-like zones, and intratumoral cystic lesions [12,13]. MRI can therefore aid in distinguishing MPNSTs from benign nerve sheath tumors when these characteristics are present. In addition, FDG PET/CT is valuable for staging and follow-up, as local recurrence and distant metastases frequently demonstrate increased metabolic activity.

The primary treatment for malignant peripheral nerve sheath tumors (MPNSTs) is wide surgical resection to achieve clear negative margins. Complete surgical excision remains the only potentially curative modality; however, this is frequently challenging because of tumor size, anatomical location, neurovascular involvement, or the presence of metastatic disease [14]. Radiotherapy is commonly used to improve local disease control in patients with large, high-grade tumors or positive surgical margins; however, it has not been shown to confer a benefit in overall survival [15]. Evidence supports that neoadjuvant chemotherapy, particularly with agents like doxorubicin and ifosfamide, might render some locally advanced primary MPNSTs operable, thereby improving disease-free survival [16]. Despite these interventions, objective response rates to chemotherapy in advanced MPNST remain disappointingly low, ranging from 20% to 30% [17]. Consequently, there are no definitively effective treatment options for patients with recurrent, unresectable, or metastatic MPNST, underscoring the urgent need for enrollment in clinical trials investigating novel targeted therapies. Unfortunately, the clinical efficacy of targeted, non-cytotoxic treatments has been limited, with stable disease achieved in fewer than 25% of patients and a median progression-free survival of less than two months [18]. MPNSTs are associated with a poor prognosis and, in many cases, represent highly aggressive and frequently fatal malignancies [18].

Local recurrence and distant metastases occur commonly, particularly in tumors larger than 5 cm in diameter, and are strongly associated with adverse clinical outcomes.

The present case demonstrates several aggressive features associated with poor prognosis in MPNST, including large tumor size, close neurovascular involvement, incomplete initial resection, repeated local recurrence, nodal and cutaneous metastases, and eventual distant metastasis. The axillary location further complicated surgical management because of the tumor's intimate relationship with major vascular and brachial plexus structures, limiting the feasibility of complete excision. Despite multimodal treatment consisting of repeated surgery, chemotherapy, radiotherapy, and reconstructive procedures, the disease demonstrated rapid progression, highlighting the aggressive biological behavior of axillary MPNST. However, this report has several limitations. As a single-case report, the findings should be interpreted cautiously and cannot establish causal relationships or be generalized to broader patient populations. In addition, incomplete information regarding the initial treatment performed at the external hospital and limited follow-up data may have affected the comprehensive interpretation of the disease course and outcomes.

Conclusions

Axillary MPNST is an exceptionally rare and highly aggressive soft tissue sarcoma associated with substantial diagnostic and therapeutic challenges. This case highlights the importance of early recognition, multidisciplinary management, and close follow-up because of the high risk of recurrence and metastatic progression despite aggressive multimodal treatment.

Availability of Data and Materials

The data analyzed are available from the corresponding author upon reasonable request.

Author Contributions

YY is the sole contributor to this manuscript. The author confirms sole responsibility for the conception and design of the study; the acquisition, analysis, and interpretation of data; the preparation of the manuscript; and for being accountable for all aspects of the work.

Ethics Approval and Consent to Participate

Written informed consent for operations and the case details was obtained from the patient. The study was conducted in accordance with the Declaration of Helsinki. Ethical approval was not required because this study was a single case report involving retrospective analysis of routinely collected clinical data without additional intervention. All images have been fully anonymized to protect patient privacy.

Acknowledgment

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Funding

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Conflict of Interest

The author declares no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at https://doi.org/10.62713/aic.4628.

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