Non-Neural Sheath Lesions of Brachial Plexus Region: Case Series and Literature Review

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Abstract

Objectives Symptomatic non-neural sheath lesions in the brachial plexus region are rare. However, surgical excision in some cases becomes challenging owing to the secondary involvement of neural elements, encasement of the vital neurovascular structures, and involvement of multiple anatomical compartments.

Material and Methods A retrospective search of surgically treated brachial plexus region non-neural sheath lesions (BPNNSLs) was performed, and 15 patients were included in the analysis. The benign BPNNSLs included 4 lipomas, 2 desmoid tumors, 1 meningioma, and 1 arteriovenous malformation. The malignant BPNNSLs included two patients, each with synovial sarcomas, non-Hodgkin's lymphomas, and metastasis. In our series, one patient had recurrent myxoid sarcoma.

Results The patient demographics, clinical, radiological, and operative details, treatment options, and outcomes were analyzed and compared with the available literature. The study highlighted the challenges and outcomes associated with the surgical management of these rare lesions.

Conclusions Complete surgical excision of benign BPNNSLs usually produces favorable outcomes with rare new deficits. Malignant lesions often require adjuvant therapy, and the prognosis varies based on the type and extent of the lesion. A multidisciplinary approach with insight into differentiating features helps in successfully treating these lesions.

Keywords

- non-neural sheath lesions
- peripheral nerve
- brachial plexus
- supraclavicular
- neck
- axilla
- surgical excision

Introduction

Peripheral nerve sheath tumors are the most common lesions in the brachial plexus region, while non-neural sheath lesions involving the brachial plexus are rare.[1,2] Desmoid tumors are often reported among these non-neural lesions, especially in the neck and axilla area.[3,4] Such lesions may adhere to, involve, or encase neurovascular structures or spread across multiple anatomical compartments, complicating excision.[4,5] Our study retrospectively reviewed surgically treated brachial plexus region non-neural sheath lesions (BPNNSLs) and highlighted the challenges faced and outcomes achieved.

Material and Methods

We retrospectively reviewed 15 cases of BPNNSLs treated surgically at P.D. Hinduja Hospital between 2011 and 2023. Benign lesions included 4 lipomas, 2 desmoid tumors, 1

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meningioma, and 1 arteriovenous malformation (AVM). Malignant cases comprised 2 synovial sarcomas, 2 non-Hodgkin's lymphomas (NHLs), 2 metastases, and 1 recurrent myxoid sarcoma in the brachial plexus region.

Results

►**Tables 1** and **2** detail the demographic features; clinical, radiological, and operative findings; treatment options; and outcomes for each patient.

Discussion

Benign Lesions

Lipoma

The four cases of lipoma presented as slowly enlarging masses, primarily in the neck and axilla. Magnetic resonance imaging (MRI) typically showed well-defined, hyperintense lesions on T1-weighted imaging (►**Fig. 1A**). Complete excision using intraoperative nerve stimulation resulted in no postoperative deficits, with follow-ups showing no recurrence (►**Fig. 1B, 1C**). Lipomas are one of the rare causes of compressive brachial plexopathy, with only 25 cases identified in the recent literature. Because of the slow growth rate, patients present with thoracic outlet syndrome only when lesions attain significant size. Complete removal aided with intraoperative electrophysiological monitoring leads to excellent outcomes with rare recurrence. Large untreated lipomas can lead to malignant transformation.[6]

Desmoid Tumor

Patients with desmoid tumors, also called aggressive fibromatosis, presented with rapidly growing masses in the neck and axilla. One had features of ipsilateral Horner's syndrome. A trial of imatinib, a chemotherapeutic agent, did not help, and the tumor progressed. MRI showed a homogenous enhancing solid lesion in a right supraclavicular compartment with extension into the mediastinum (►**Fig. 1D**). Sternotomy was performed by a cardiovascular surgeon. The tumor was encasing the internal jugular vein and the vertebral artery, which were sacrificed. The subclavian and carotid arteries that were compressed by the tumor were preserved. Complete excision with negative margins was performed (►**Fig. 1E, 1F**). He developed hoarseness of voice probably due to insult of the recurrent laryngeal nerve. The hoarseness was initially managed by injection laryngoplasty. At the 24-month follow-up visit, he had no recurrence, and the hoarseness of his voice had completely recovered. The second patient with recurrent axillary region desmoid tumor, operated previously at some other center, had shoulder and hand grip weakness, which improved after tumor excision and remained recurrence free at 12 months of follow-up. Although classified as benign, these tumors exhibit locally invasive behavior, making treatment difficult, especially when affecting the brachial plexus. Their aggressive nature often leads to high recurrence rates (up to 80%) if margins are positive after surgery. The proximity to neural elements results in functional challenges and complicates achieving clear surgical margins without risking additional neurological deficits. Adjuvant treatments such as radiotherapy, chemotherapy, and hormonal therapy are commonly used postsurgery to manage residual disease. Radiotherapy has shown efficacy, achieving local control in 79 to 82% of cases, although young patients are more prone to radiation-induced side effects. While surgery remains a first-line option for localized control, multimodal treatment is essential for durable disease control, especially given the high recurrence rates. Progression-free survival rates are typically favorable with combined therapies, achieving 90% at 2 years.[7] Periodic and long-term postsurgical assessments are mandatory for desmoid tumors.

Meningioma

A case of meningioma presented with a slowly enlarging neck mass and mild upper limb pain. MRI showed a homogenous supraclavicular mass (►**Fig. 1G**). Positron emission tomography (PET) scan did not reveal hypermetabolism in the lesion. Radical excision was performed, sparing neural elements. The patient remained asymptomatic at 36 months of follow-up. In the latest systematic review of 213 primary extracranial meningiomas (PEMs), only 3 cases of neck meningioma were identified. Like our case, these three tumors also remained stable after radical to complete resection. PEMs are thought to arise from ectopic arachnoid cells from nerve sheaths or neural crest cells, and mechanisms involving trauma or raised intracranial pressure could stimulate migration, causing cells to settle in atypical locations like the neck. Surgical resection is the primary treatment and has shown favorable outcomes with low recurrence. The PEMs' benign nature and careful resection generally lead to positive long-term recovery.[8]

Arteriovenous Malformation

One case of AVM presented with slowly enlarging neck mass and mild upper limb pain, confirmed by MRI and computed tomography angiography (CTA; ►**Fig. 1H**). Complete excision was achieved without any neurological deficit. At 24 months of follow-up, there was no recurrence, and the patient remained asymptomatic. High-quality imaging modalities like MRI, CTA, or digital subtraction angiography help identify feeding arteries, draining veins, and the extent of extremity AVM to critical structures, including the brachial plexus. Embolization of feeding arteries within 24 to 72 hours before surgery can reduce blood flow to the AVM and minimize intraoperative blood loss. Polyvinyl alcohol particles or similar embolic materials are often used, and careful catheter positioning ensures precise occlusion of feeding vessels without affecting surrounding structures. At surgical resection, the standard approach is to work distal to proximal along the feeding arteries, ligating them individually to reduce AVM pressure and lessen the chance of sudden blood loss. After arterial control, drainage veins are clamped and ligated. Regular imaging, typically using CTA or MRI, is crucial to detect residual or recurrent AVM, as re-expansion of incompletely resected AVMs can occur through neovascularization from adjacent vessels.[9,10]

Table 1 Key features of benign brachial plexus region non-neural sheath lesions

Patient	Diagnosis	Age (y)	Sex	Clinical features	PET/CT scan findings	Preoperative biopsy performed	Brachial plexus elements	Extent of resection	Adjuvant treatment	Postoperative outcome	Last follow-up (mo)	Outcome at last follow-up	Recurrence
1	Lipoma	42	M	Slowly enlarging mass in the neck, and mild upper limb pain for the past 1 y	Not done	Yes	Compression of the C5–C6 nerve roots and upper trunk	Complete excision	Not advised	No new deficit; pain resolved	18	Asymptomatic	No
2	Lipoma	36	F	Slowly enlarging mass in axilla for the past 1.5 y	Not done	No	Compression of cords and branches	Complete excision	Not advised	No new deficit	12	Asymptomatic	No
3	Lipoma	50	F	Slowly enlarging mass in neck for the past 1 y	Not done	No	Compression of the C5–C7 nerve roots, upper trunk, and middle trunk	Complete excision	Not advised	No new deficit	27	Asymptomatic	No
4	Lipoma	45	M	Slowly enlarging mass in neck for the past 1 y	Not done	No	Compression of the nerve C5–C6 nerve roots and upper trunk	Complete excision	Not advised	No new deficit	34	Asymptomatic	No
5	Desmoid tumor	49	M	Rapidly enlarging mass in neck over 1 mo, ipsilateral ptosis and miosis (Horner's syndrome). Started on imatinib (CT) but tumor continued to progress	Not done	Yes	Compression of the C5–C7 nerve roots, upper trunk, and lower trunk	Complete excision	Not advised	Hoarseness of voice	24	Asymptomatic	No
6	Recurrent desmoid tumor	23	F	Rapidly enlarging mass in axilla for 3 mo; operated at another center. Biopsy: benign neurogenic tumor (faulty?). After surgery: progressive enlargement of swelling over 1 y, and right shoulder abduction and hand grip weakness for the past 3 mo	Not done	No	Compression of cords and branches	Complete excision	Not advised	No new deficit; shoulder abduction and hand grip improved	12	Asymptomatic	Yes (in the supraclavicular region)
7	Meningioma	37	F	Slowly enlarging mass in neck for 1 y and mild upper limb pain for 1 y	No hypermetabolism	Yes	Involvement of the perineurium of the C5–C6 nerve roots and upper trunk	Radical excision, small part of the tumor attached to the neural elements left behind	Not advised	No new deficit	36	Asymptomatic	No
8	Arteriovenous malformation	22	M	Slowly enlarging mass in neck for 2 y, and mild upper limb pain	Not done	No	Compression of the C5–C6 nerve roots and upper trunk	Complete excision	Not advised	No new deficit	24	Asymptomatic	No

Abbreviations: CT, chemotherapy.

Table 2 Key features of malignant brachial plexus region non-neural sheath lesions

Patient	Diagnosis	Age (y)	Sex	Clinical features	PET/CT scan findings	Preoperative biopsy performed	Brachial plexus elements	Extent of resection	Adjuvant treatment	Postoperative outcome	Last follow-up (mo)	Outcome at last follow-up	Recurrence
9	Synovial sarcoma	40	M	Rapidly enlarging mass over 6 mo, upper limb pain, paresthesia, and weakness for 4 mo	No metastasis, hypermetabolism of lesion	Yes	Perineurium of the upper trunk and C5–C7 nerve roots	Complete excision with external neurolysis of neural elements	Received adjuvant RT and CT	No new deficit	24	Asymptomatic	No
10	Synovial sarcoma	38	M	Rapidly enlarging mass over 4 mo; upper limb pain and paresthesia for 3 mo	Lung metastasis, hypermetabolism of lesion	Yes	Perineurium of the C5–C7 nerve roots	Complete excision along with excision of neural elements	Received adjuvant RT and CT	Shoulder abduction and elbow flexion weakness	6	Persistent shoulder abduction and elbow flexion weakness	Yes
11	Recurrent myxoid sarcoma	28	F	Rapidly enlarging mass in the neck, operated outside and CT given, recurrence with mass, pain, paresthesia shoulder abduction and elbow flexion weakness	Not done	Yes	Perineurium of the C5–C7 nerve roots	Complete excision with external neurolysis of neural elements	Received adjuvant RT	No new deficit	12	Asymptomatic	No
12	Non-Hodgkin's lymphoma	42	M	Pain and paresthesia in the upper limb	Hypermetabolism	No	Encasement of the C5–T1 nerve roots and trunks	Subtotal excision	Received adjuvant CT	No new deficit	36	Asymptomatic	No
13	Non-Hodgkin's lymphoma	32	M	Pain in the upper limb	Hypermetabolism	No	Encasement of the nerve roots (C5–T1) and trunks	Subtotal excision	Received adjuvant CT	No new deficit	24	Asymptomatic	No
14	Metastasis from breast carcinoma	56	F	Pain, paresthesia and hand grip weakness for 3 mo	Hypermetabolism	No	Involvement of the C5–T1 nerve roots and trunks	Radical excision	Received adjuvant CT	No new deficit,	12	Persistent hand grip weakness	No
15	Metastasis from breast carcinoma	48	F	Severe neurogenic pain, and shoulder and hand grip weakness	Hypermetabolism	No	Involvement of the C5–T1 nerve roots and trunks	Radical excision with external neurolysis of neural elements	Received adjuvant CT	No new deficit	6	Death due to spread of breast carcinoma	No

Abbreviations: CT, chemotherapy; PET/CT, positron emission tomography computed tomography; RT, radiotherapy.

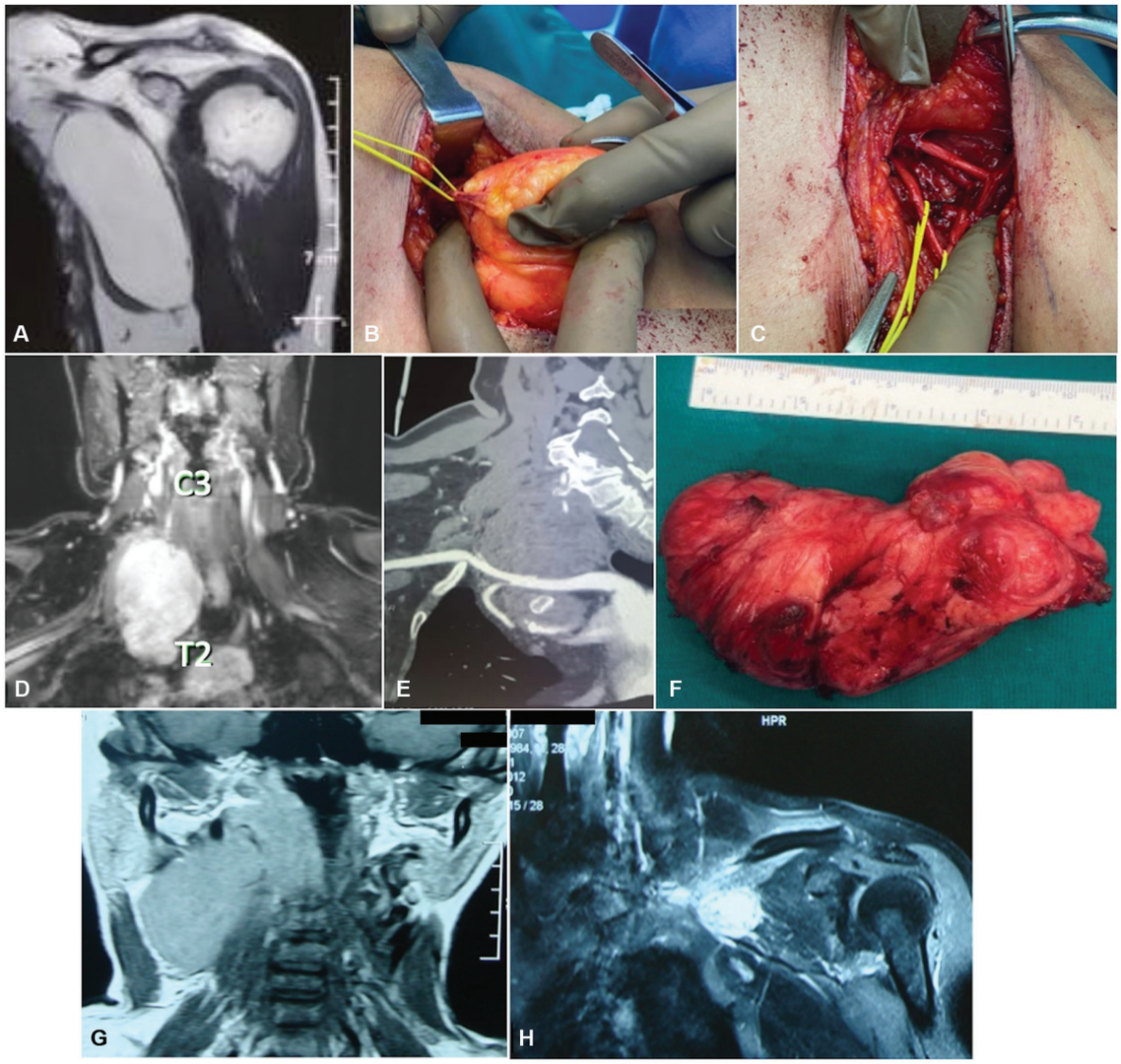

Fig. 1 Benign brachial plexus region non-neural sheath lesions. Lipoma: (**A**) Magnetic resonance imaging (MRI) scan of the left axilla, T1-weighted image, showing homogenous, hyperintense, well-defined lesion. (**B**) Intraoperative image showed a lipomatous lesion in the axilla closely related to the divisions and cords of brachial plexus. (**C**) Intraoperative image showing complete excision of the lipoma with preserved infraclavicular brachial plexus. Desmoid tumor: (**D**) Contrast-enhanced MRI scan of the brachial plexus showing a large solitary homogenously enhancing lesion in the right supraclavicular region with extension in the mediastinum. (**E**) Computed tomography angiography showed inferior displacement of the right subclavian artery and complete encasement of the vertebral artery. (**F**) The operative specimen after complete tumor excision. Meningioma: (**G**) MRI scan of the brachial plexus, T1-weighted image showing a homogenous lesion in a right supraclavicular compartment with involvement of the roots and trunks of the brachial plexus. Arteriovenous malformation**:** (**H**) MRI scan of the brachial plexus showing brilliantly enhancing left supraclavicular lesion.

Malignant Lesions

Synovial and Myxoid Sarcomas

Two patients with synovial sarcoma and one patient with recurrent myxoid sarcoma presented with rapidly enlarging masses, severe upper limb pain, paresthesia, and motor weakness. MRI scan revealed heterogeneous solid-cystic giant supraclavicular lesions in relation to major neurovascular structures (►**Fig. 2A**). CTA of the neck vessels showed that the lesions were displacing the major neck vessels, including the subclavian vessels (►**Fig. 2B**). PET scan in two patients showed hypermetabolism in the lesion, and in one patient, there was metastatic spread to the lungs. Preoperative biopsy in these cases helped in formulating aggressive surgical plans. Intraoperatively, the perineurium of the brachial plexus was secondarily involved by the lesions. Complete tumor excision with external neurolysis was performed. The patient subsequently received adjuvant radiotherapy and/or chemotherapy. One of the patients encountered an early recurrence at 6 months of follow-up. The neural involvement in synovial sarcoma is not commonly reported in the literature.[11] The initial imaging with MRI and biopsy (preferably core needle over fine-needle aspiration) is essential for an accurate diagnosis. Immunohistochemical

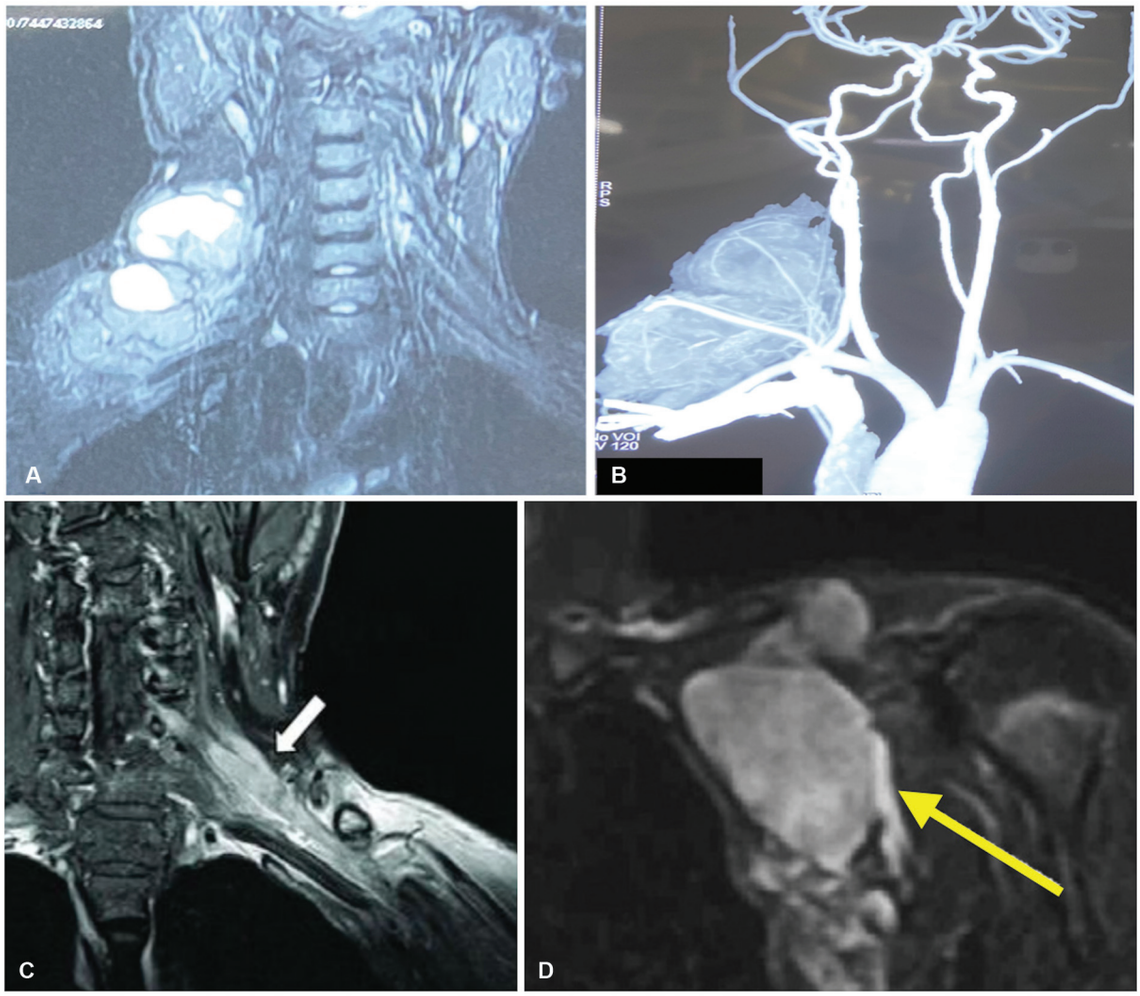

Fig. 2 Malignant brachial plexus region non-neural sheath lesions. Synovial sarcoma: (**A**) Contrast-enhanced magnetic resonance imaging (MRI) scan of the brachial plexus showing a heterogeneously enhancing solid cystic lesion in the right supraclavicular compartment involving the roots of the brachial plexus. (**B**) Computed tomography angiography showed medial and inferior displacement of the common carotid and subclavian vessels, respectively. Non-Hodgkin's lymphoma: (**C**) Contrast-enhanced MRI scan of the left brachial plexus showed enhancement and thickening of the brachial plexus elements (*white arrow*). Metastasis from breast carcinoma: (**D**) MRI scan of the brachial plexus showing a homogenously enhancing solitary left supraclavicular lesion (*yellow arrow*).

analysis also helps in confirming the diagnosis.[12] The surgery involves an individualized approach and an extensive dissection to avoid compromising the brachial plexus and surrounding structures. The role of adjuvant therapies (chemotherapy and radiotherapy) is debatable. Synovial sarcomas generally benefit from radiotherapy, especially in cases with positive margins. Some studies suggest preoperative chemotherapy for tumors over 5 cm or those with high metastatic potential. However, its impact on survival rates is inconclusive. Also, few reported cases have shown long-term recurrence-free rates without the adjuvant treatment, suggesting a variable disease course. Long-term follow-up with imaging is essential, as synovial sarcomas have a high risk of local recurrence and metastasis, often to the lungs. The prognosis depends on factors like tumor size, resection margins, and the patient's age.[13]

Non-Hodgkin's Lymphoma

Two cases of NHL presented with pain and paresthesia in the upper limb. MRI showed diffuse thickening of the neural elements in the supraclavicular compartment (►**Fig. 2C**). PET scan showed hypermetabolism suggestive of the malignant nature of the lesions. The tumors were seen to encase the brachial plexus; subtotal excision was performed aiming at biopsy and debulking, followed by adjuvant chemotherapy. The patients remained asymptomatic at the 24- and 36-month follow-ups. NHLs, especially diffuse large B-cell lymphoma (DLBCL), are the commonly described brachial plexus neurolymphomatosis. Infiltration into the brachial plexus affects the nerve roots to the branches. The extent of involvement can range from localized mononeuropathy to widespread plexopathy.[14] MRI and PET scans are pivotal for visualizing neural enlargement, fascicular disorganization,

and enhancement, which are indicative of nerve infiltration. CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) combined with rituximab is the standard regime protocol for systemic control of DLBCL, often augmented by high-dose methotrexate to penetrate the blood–nerve barrier and prevent relapses. Cases with severe neuropathic pain or focal nerve infiltration respond well to radiotherapy. However, the prognosis in neurolymphomatosis tends to be poorer than in other DLBCL cases due to the challenges in treatment response and high relapse rates.[15]

Metastasis from Breast Carcinoma

In our series, both cases of metastasis were from breast carcinoma. They presented with severe neurogenic pain and motor weakness. MRI showed localized, solid, supra- and infraclavicular lesions involving all brachial plexus elements (►**Fig. 2D**). PET scan showed hypermetabolism with increased intake. Radical excision and adjuvant chemotherapy were given. One patient had persistent hand grip weakness at the 12-month follow-up, and the other patient died after 6 months from the time of surgery due to the spread of carcinoma to multiple organs. Breast carcinoma spreads locally or via lymph nodes in the axillary or supraclavicular regions and is most common to metastasize to the brachial plexus. Lung carcinoma, particularly the Pancoast tumor involving the upper lobe of the lung, is the second most common primary to spread. Other less common lesions that spread to the brachial plexus are melanomas, lymphomas, renal carcinomas, and prostate carcinomas.[16] Complete excision is rarely performed because of the complex anatomy and potential for significant nerve damage. Surgical decompression followed by adjuvant treatment is the common surgical protocol. Prognosis is generally poor, given that brachial plexus metastasis often occurs in the late stages of cancer. Pain management and physical therapy are important supportive measures for improving patient comfort and function.[17]

Other Rare Lesions

The literature also describes rarer non-neural sheath brachial plexus region lesions, including venous angiomas, hemangiopericytomas, glomus tumors, capillary hemangiomas, neurovascular hamartomas, hemangioblastomas, myositis ossificans, osteochondromas, ganglioneuromas, cystic hygromas, myoblastomas, granular cell tumors, epidermoid cysts, Ewing's sarcoma, and extrarenal rhabdoid tumors.[5]

Conclusions

Management of BPNNSLs requires a strategic approach tailored to the lesion's benign or malignant nature. For benign BPNNSLs, complete surgical excision is generally effective, often yielding favorable outcomes with minimal recurrence. Techniques like intraoperative nerve stimulation and precise dissection minimize postoperative neurological deficits. Regular follow-up is essential, especially for aggressive benign lesions like desmoid tumors, which carry a high risk of recurrence despite complete resection.

Malignant BPNNSLs often require a multidisciplinary approach, integrating surgery with adjuvant therapies. For sarcomas and lymphomas, the combination of surgical excision with radiotherapy and chemotherapy is vital to control local and systemic disease. In metastatic cases, palliation becomes the primary goal, with treatments focusing on decompression, pain relief, and quality-of-life enhancement.

Overall, managing BPNNSLs benefits from a deep understanding of tumor biology, careful surgical planning, and collaboration among surgical, radiological, and oncological teams. Long-term imaging and clinical follow-up are essential to detect early recurrences and guidance for further treatment.

Authors' Contributions

A.K. and V.K. contributed to the concept, design, definition of intellectual content, literature search, clinical studies, data acquisition, manuscript preparation, manuscript editing, and manuscript review. B.J., G.M., and K.D. contributed to the concept, design, definition of intellectual content, literature search, clinical studies, manuscript editing, and manuscript review.

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Conflict of Interest

None declared.

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