

Case Report

A rare case of giant intercostal nerve schwannoma

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ABSTRACT

Schwannomas are benign peripheral nerve sheath tumours that most commonly affect spinal and cranial nerves, while their occurrence along intercostal nerves is exceedingly rare. Giant schwannomas, those exceeding 5 cm in diameter, can exert significant mass effect on adjacent structures, causing pain, neurological symptoms, or respiratory compromise. We present the case of a 33-year-old woman referred to the Royal National Orthopaedic Hospital with a symptomatic 10 cm soft tissue mass arising from the left 7th intercostal nerve. Preoperative magnetic resonance imaging (MRI) revealed an intramuscular, septated lesion with atypical imaging characteristics, prompting differential diagnoses including sarcoma. Core biopsy confirmed a benign schwannoma. The patient reported persistent pain, hypersensitivity, and early satiety. Surgical excision was performed via an anterolateral chest wall approach with meticulous dissection to preserve surrounding nerves and pleura. Histopathology confirmed a benign schwannoma with characteristic spindle cell architecture and S100 protein positivity. Postoperative recovery was uneventful, with resolution of symptoms and no neurological deficit. This case highlights the diagnostic challenge posed by giant intercostal schwannomas and the importance of thorough imaging, histological evaluation, and surgical planning. Despite their benign nature, giant schwannomas can cause significant morbidity, warranting timely surgical intervention. Multidisciplinary management involving orthopaedic surgeons, radiologists, and histopathologists is essential to optimise outcomes. Our report adds to the limited literature on giant intercostal nerve schwannomas and underscores the need for heightened clinical suspicion when evaluating large intrathoracic soft tissue masses with atypical radiographic features.

Keywords: Schwannoma, Intercostal nerve, Nerve sheath tumour, Chest wall mass, Benign

INTRODUCTION

Schwannomas are benign peripheral nerve sheath tumours originating from Schwann cells, and they are the most common type of peripheral nerve sheath tumours.¹ They account for approximately 48% of benign neurogenic tumours and are most frequently found along cranial, spinal, and peripheral nerves.¹ While schwannomas are typically small, slow-growing, and asymptomatic, large tumours - commonly referred to as giant schwannomas - can cause significant mass effect, compressing adjacent structures and leading to clinical symptoms.²

Schwannomas arising from intercostal nerves are uncommon.³ When they reach substantial sizes, they may

present with respiratory symptoms, chest pain, or neurological deficits due to mass effect on the surrounding tissues.⁴ Although benign, giant schwannomas require surgical excision to prevent progressive symptoms and complications such as pulmonary compression, spinal cord displacement, or vascular compromise.^{2,5} The surgical approach is influenced by tumour size, anatomical location, and involvement of surrounding structures.⁴

Here, we present a rare case of a giant schwannoma arising from the intercostal nerve, successfully excised through a tailored surgical approach. We discuss the imaging characteristics, intraoperative challenges, and histopathological findings, placing our case within the broader context of existing literature.

CASE REPORT

A 33-year-old female midwife with a background of asthma managed with Ventolin was referred to the Sarcoma Unit at the Royal National Orthopaedic Hospital on April 29, 2024, following the identification of a suspicious 10 cm soft tissue mass in the left 7th intercostal space on a computed tomography (CT) abdomen and pelvis (CTAP) performed on 28 April 2024 by the referring unit. Her clinical presentation was characterized by a three-month history of recurrent viral upper respiratory tract infections (URTI), with a one-month history of progressive left upper quadrant (LUQ) pain and epigastric discomfort. She also reported early satiety, though she denied constitutional symptoms such as fever, night sweats, or weight loss.

Her past medical history was notable for an episode of Epstein-Barr virus (EBV)-induced jaundice and splenomegaly in November 2022, for which she had been evaluated at Princess Alexandra Hospital. However, she had been lost to gastroenterology follow-up. At the time of her current presentation, there was no clinical jaundice, and her stool and urine color remained normal.

Imaging findings

A magnetic resonance imaging (MRI) thorax with contrast was performed on 14 May 2024, providing characterisation of the lesion. A 95×58 mm (axial)×82 mm (craniocaudal) soft tissue mass (Figure 1a) was identified in the left lateral thoracic wall, with imaging features suggesting an intramuscular origin. The external intercostal muscle was stretched and expanded, while the internal intercostal muscle was not visualized, raising suspicion of infiltration. The innermost intercostal muscle was displaced medially due to the expansile nature of the lesion (Figure 1b).

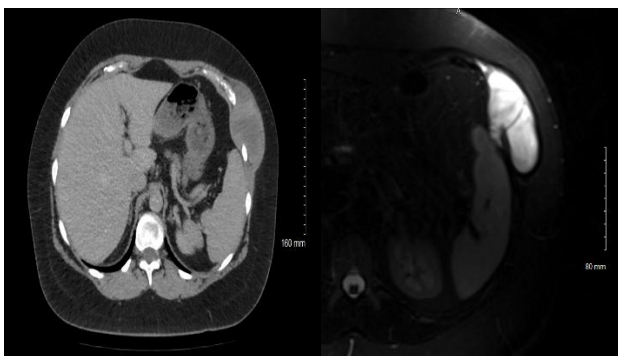


Figure 1: MRI findings.

The lesion demonstrated increased T2 signal intensity with septations and fibrous strands traversing the tumour (Figure 2a), and areas of low-grade restricted diffusion (Figure 2b), particularly in the dependent portions, which showed prominent postcontrast enhancement. Notably, there was no evidence of direct infiltration into the thoracic

or peritoneal cavity. However, the imaging characteristics were not entirely classical for a nerve sheath tumour, raising concerns for an alternative diagnosis, such as an intramuscular metastasis or soft tissue sarcoma.



Figure 2: MRI findings.

Histopathology and diagnosis

Given the atypical radiological features, an ultrasound-guided needle biopsy was performed on 31 May 2024, and histological analysis revealed a nerve sheath tumor composed of spindle cells randomly arranged within a fibromyxoid stroma, with tactile differentiation. There was no tumor necrosis, and occasional mitoses were observed.

Immunohistochemistry confirmed diffuse positivity for S100 protein, consistent with a Schwann cell origin, while epithelial membrane antigen (EMA) was negative. H3K27me3 was retained, effectively excluding a diagnosis of malignant peripheral nerve sheath tumor (MPNST). The Ki-67 proliferative index was less than 5%, indicative of low mitotic activity.

Based on these findings, a diagnosis of a benign schwannoma was established, with no histological evidence of malignancy.

Clinical course and examination

Following the confirmed benign nature of the lesion, the patient was referred to the peripheral nerve injury unit for further assessment and management. She reported persistent discomfort, hypersensitivity, and pain over the lower left chest wall, exacerbated by wearing clothes or a bra. She also described localized tenderness and a shooting pain upon palpation.

On clinical examination, a mobile, subcutaneous mass, measuring approximately 12×4 cm, was palpable in the lower anterior left chest wall. The lesion was tender to palpation, with percussion eliciting discomfort.

Sensory examination revealed hypoesthesia (4/10 compared to the contralateral side), but there were no motor deficits or features of neuropathic pain extending beyond the lesion itself.

Surgical decision and management

Given the progressive symptoms, size of the lesion, and its effect on daily function and quality of life, surgical excision was deemed appropriate. The patient was counselled regarding the risks of surgery, including pain, bleeding, infection, injury to adjacent nerves or vascular structures, and the possibility of recurrence.

Considering the lesion's expansile nature and potential infiltration into the intercostal muscles, the primary surgical objective was to achieve complete resection while preserving nerve integrity.

Surgical technique

Under general anaesthesia, the patient was supine with full exposure to the area of focus on the left side of antero-lateral side of the chest wall. Transverse straight incision was made over the mass measuring around 10-12 cm (Figure 3).

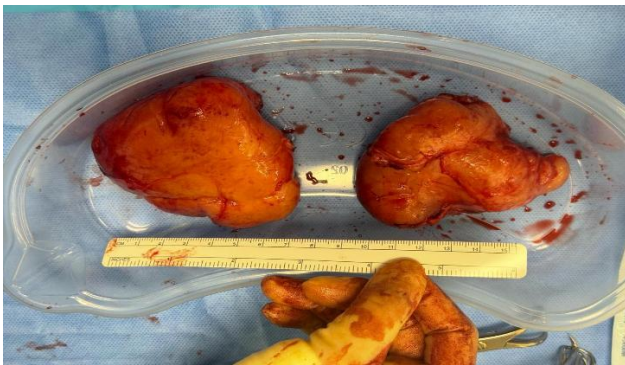


Figure 3: MRI findings.

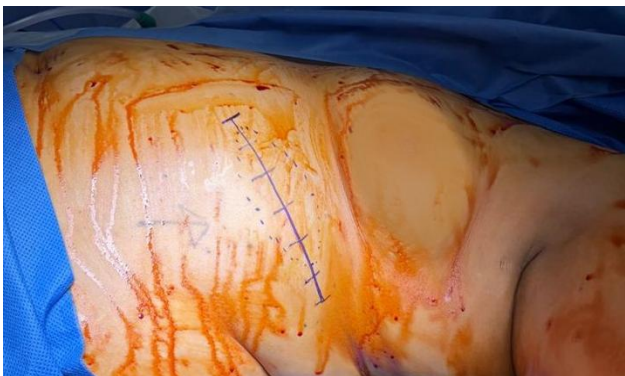


Figure 4: MRI findings.

Skin incised sharply with meticulous subcutaneous layer dissection down to fascia with hemostasis secured. The tumor was identified and was attached to adjacent ribs and extended into intercostal space. Layer by layer dissection done isolating the healthy nerves with forceps and freer and protecting any damage to intrathoracic structures. The tumor was found divided by thick layer into two encapsulated masses. Both were excised completely

(Figure 4) and leaving healthy nerve intact. Proper irrigation and hemostasis were done. Valsalva maneuver conducted to exclude any pleural injury during dissection, which was intact. Deep layer closure done, then skin closed in two layers. No drain was needed to be used. Skin glue and semi-occlusive dressing was applied.

Post-surgery

The patient was discharged on the same day. Two weeks postoperatively the patient was seen in outpatient clinic for wound check - wound was clean and healed. There were no neurological symptoms detected. Despite mild pain, there was no collection, sensory or respiratory dysfunction noted.

Final histopathology

Macroscopic examination of the larger tumor measuring 80×73×50 mm was partly covered by a thin layer of translucent membrane, while the smaller one measuring 80×55×40 mm, completely covered. Microscopic examination revealed a nerve sheath tumor composed of spindle cells within a fibro-myxoid stroma with focal vague nuclear palisding. There was focal mild cytological atypia with hyper chromatic nuclei. Perivascular cellular condensation was focally noted. There were no identified mitotic figures or tumor necrosis. These findings confirmed the diagnosis of benign schwannoma.

DISCUSSION

Schwannomas are well-encapsulated, slow-growing tumours that arise from Schwann cells and typically present as solitary lesions along peripheral nerves.¹ They are histologically characterized by Antoni A and Antoni B areas, with Antoni A areas being cellular with nuclear palisading, and Antoni B areas being myxoid and hypocellular. Immunohistochemical staining for S100 protein is a hallmark feature, aiding in the confirmation of diagnosis.¹

Giant schwannomas and intercostal nerve involvement

While schwannomas of the spinal nerves, brachial plexus, and sciatic nerve are well-documented, intercostal nerve schwannomas are rare.³ Case reports describe these tumours as well-tolerated for long periods, often discovered incidentally on imaging or presenting with dyspnoea, chest pain, or neurological symptoms when they enlarge.^{4,6} In the present case, the tumour's large size necessitated an individualized surgical approach to ensure safe excision while preserving surrounding structures.

Surgical considerations and challenges

Complete surgical resection is the gold standard for treating schwannomas, particularly when they reach significant size.^{2,5} The choice of surgical approach depends on tumour location and potential involvement of adjacent

structures.⁴ Preoperative imaging, particularly MRI and contrast-enhanced CT, is crucial for delineating the tumour's origin, extent, and relationship with nearby anatomical structures.⁴

Several studies have highlighted the importance of preoperative embolisation in cases of highly vascular tumours, as this reduces intraoperative blood loss and facilitates en-bloc resection.⁷ In cases where tumour adherence to the nerve is significant, microsurgical dissection techniques help preserve nerve function.⁵ Given that intercostal schwannomas are often in close proximity to the pleura, care must be taken to avoid pleural breach or pneumothorax.⁴

Histopathology and differential diagnosis

The diagnosis of schwannoma is confirmed histologically, with characteristic biphasic Antoni A and Antoni B areas and strong S100 positivity.¹ The differential diagnosis includes - neurofibromas: lack complete encapsulation and are often associated with neurofibromatosis type 1 (NF1), malignant peripheral nerve sheath tumours (MPNSTs): rare but aggressive; characterized by higher mitotic activity and necrosis, and soft tissue sarcomas: show heterogeneous enhancement and infiltrative growth on imaging.^{1,8,10}

Importance of early intervention

While schwannomas are benign, delayed intervention can lead to complications due to progressive tumour growth and compression of adjacent structures.^{2,9} Cases of respiratory failure secondary to giant intrathoracic schwannomas have been reported, emphasizing the need for timely diagnosis and treatment.⁵

CONCLUSION

Giant schwannomas of the intercostal nerve are exceptionally rare, and their slow-growing nature often results in delayed diagnosis and treatment. Despite their benign histology, large schwannomas can cause significant mass effect, leading to symptoms that warrant surgical excision.

This case highlights the importance of early identification, meticulous surgical planning, and intraoperative techniques to achieve complete tumour resection while preserving nerve function. Given the challenges associated with large schwannomas, a multidisciplinary approach involving neurosurgeons, thoracic surgeons, and radiologists is essential for optimizing patient outcomes.

Regular follow-up is also crucial to detect recurrence or malignant transformation, although the latter is rare.

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