

Primary Pleural Neurofibroma: Incidental Discovery of an Exceedingly Rare Neoplasm

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DOI: <https://doi.org/10.36347/sjmc.2026.v14i10.047>

| Received: 16.08.2026 | Accepted: 21.10.2026 | Published: 24.10.2026

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Abstract

Case Report

Introduction: Primary pleural neurofibromas (PPNF) are a type of benign pleural tumors that is very uncommon. In this case report, we describe our first case in our department which was a successful surgical removal of this entity by using U- VATS approach. **Case Presentation:** This is a 20-year-old woman, referred to our thoracic surgery department for a suspect right pleural lesion, the physical examination was normal without clinically palpable chest mass. The chest CT scan showed a large heterogeneous, hypodense right pleural mass next to the upper right lobe and fills the costo-vertebral groove suggesting a pleural liposarcoma, a chest MRI was then requested which showed a pleural lesion centered on the costovertebral groove at the height of (D2, D3, D4) well-limited with heterogeneous signal in T1 and T2 that measures (33 x53x55 mm), double fatty and tissue component, pushing the pulmonary parenchyma forward and backward in contact with the posterior arches of the 2nd, 3rd and 4th ribs and the corresponding costo-vertebral joints without intra spinal canal extension. After multidisciplinary concertation, a CT-guided biopsy of the lesion was performed, the histological examination of which revealed a Primary pleural neurofibromas (PPNF), then a complete resection of the lesion was performed under U-VATS approach and the histology of the specimen confirmed the diagnosis. The patient is currently in a good condition after two years of the operation and does not present any complications for the long-term follow-up. **Discussion:** Metastatic deposits are the most common cause of pleural masses, solitary or multiple. Primary pleuralneoplasms are rare entities that are occasionally encountered. Of these, the tumors of neurogenic origin are exceedingly rare with only a few cases reported in the literature, This is the 4th case in English literature to our knowledge [1]. Treatment of localized neurofibromas is complete surgical resection. Neurofibromas grow within and cannot be separated from normal nerve, and complete excision of the neoplasm requires sacrifice of the nerve. Local recurrence after complete excision is unusual with generally a good prognosis [2]. **Conclusion:** The Primary pleural neurofibromas (PPNF) are a very rare entity of pleural neoplasms. This is the first case in our department that demonstrates that the PPNF must be suspected in any pleural mass, especially in the absence history of cancerous illness or occupational exposure, and that complete surgical excision, when it's possible, is a safe and effective treatment.

Keywords: Primary Pleural Neoplasm, Primary Pleural Neurofibromas (PPNF), Benign Pleural Tumors, Thoracic Surgery.

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INTRODUCTION

Primary neoplasms of the pleura are exceedingly rare. Pleural neoplasms are more commonly metastatic than primary. They are more often malignant than benign, with malignant mesothelioma being the most common entity. Benign neoplasms include solitary fibrous tumor, lipomatous tumors, adenomatoid tumor, calcifying fibrous tumor, multicystic mesothelioma, and schwannoma. Other benign tumor-like lesions include

nodular pleural plaque and mesothelial cysts. Of these, solitary fibrous tumor is the most frequently encountered [3, 4].

Neurofibromas are benign neurogenic tumors that rarely involve the pleura our review of the literature found that there is a lack of similar cases, only 4 cases including ours were found.

Citation: Hafidi, S., Niyonkuru, E., Idelhaj, N., Boubia, S., Karkouri, M., & Ridai, M. (2026). Primary Pleural Neurofibroma: Incidental Discovery of an Exceedingly Rare Neoplasm. *Sch J Med Case Rep*, 14(10), 2325-2331. <https://doi.org/10.36347/sjmc.2026.v14i10.047>

This case report describes a case of Primary pleural neurofibroma (PPNF) completely removed by using U-VATS approach, in a 20-years-old woman. The patient was diagnosed and managed in our institution; a tertiary referral university teaching hospital. This work has been reported in line with the SCARE 2025 criteria [5].

CASE PRESENTATION

This is a 20 years old female patient, without significant past medical history and she was a non-smoker, in whom we discovered by chance a suspicious pleural mass, without functional signs during a chest x-ray requested as part of an assessment health with a view to applying for a job.

Following this, the patient was referred to the pulmonologist; the rest of the physical examination was unremarkable. For further evaluation, contrast-enhanced chest CT was done, which showed an heterodense right pleural mass measuring (55x49x29 mm) next to the right upper lobe and fills the costovertebral groove at the height of (D2, D3, D4) However, no obvious spinal extension was seen. (Fig.1)

For confirming the same, the patient underwent a chest MRI which revealed a pleural lesion centered on the costovertebral groove at the height of (D2, D3, D4) heterogeneous signal well limited in T1 and T2, hypointense on T1 weighted images and hyperintense on T2 weighted images with double component adipose and tissue, pushing the pulmonary parenchyma forwards and backwards into contact with the posterior arches of the 2nd, 3rd and 4th ribs and the corresponding costovertebral joints without extension of the intraspinal canal. (Fig.2)

After a multidisciplinary consultation, the decision was to do a CT-guided fine needle biopsy of the lesion, the histopathology of which showed a pleural neurofibromatous localization, then the patient was referred to our department for excision of the lesion, in a minimally invasive approach by using a right U-VATS.

Under general anesthesia and Selective intubation, in a left lateral position, the mass was removed through a right Uniportal VATS approach. A 3 cm incision was made at the level of the 4th intercostal space along the right medium axillar line and a wall protector was positioned, the exploration found an ovoid pleural mass measuring approximately 6 cm, encapsulated with well-circumscribed margins, It was well separated from the right lung parenchyma and centered on the costovertebral groove (D1, D2, D3) very adherent to the wall and intercostal spaces, especially to the posterior arches of the ribs (K2, K3, K4) and it was in a close contact with the stellate ganglion and the sympathetic chain, without spinal communication as suggested by imaging studies. The mass was completely dissected from the structures mentioned above (Fig. 3). The extraction was made successfully through an endobag. No perioperative complications occurred. A pleural drainage was performed. The post-operative period was uneventful. The pleural drainage was removed on the second day, and the patient was discharged from the Hospital. The histopathology confirmed a Primary pleural neurofibroma (PPNF) (Fig. 4). According to the analysis, its proper dimension was: 55 mm × 53mm × 49mm. Weight: 87 g. A control chest x-ray performed after 15 days was normal. The patient is currently in very good general condition after 2 years of the intervention, and does not present any complications or recurrence during the long-term follow-up.

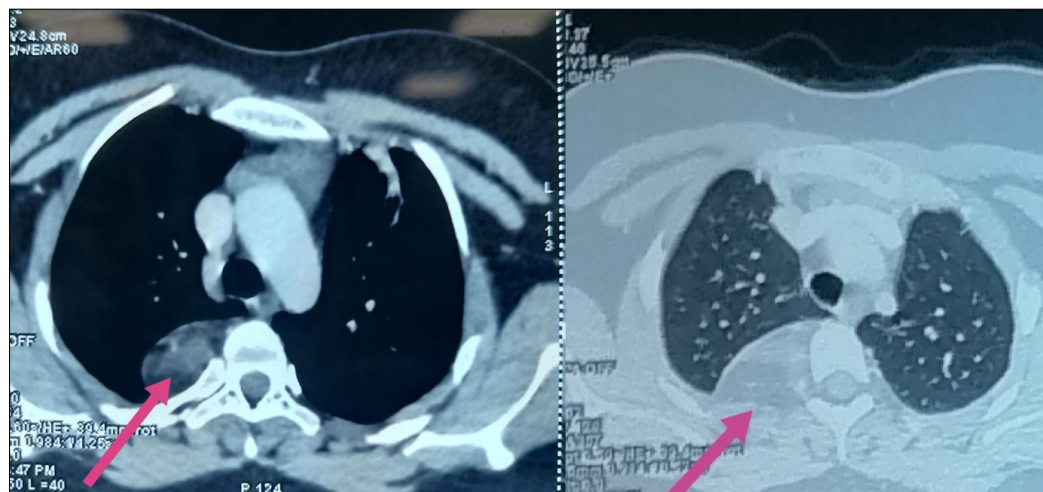


Figure 1: Chest CT Scan: heterodense right pleural mass measuring (55x49x29 mm) next to the right upper lobe and fills the costovertebral groove at the height of (D2, D3, D4) pushing the pulmonary parenchyma forwards and backwards into contact with the posterior arches of the 2nd, 3rd and 4th ribs and the corresponding costovertebral joints

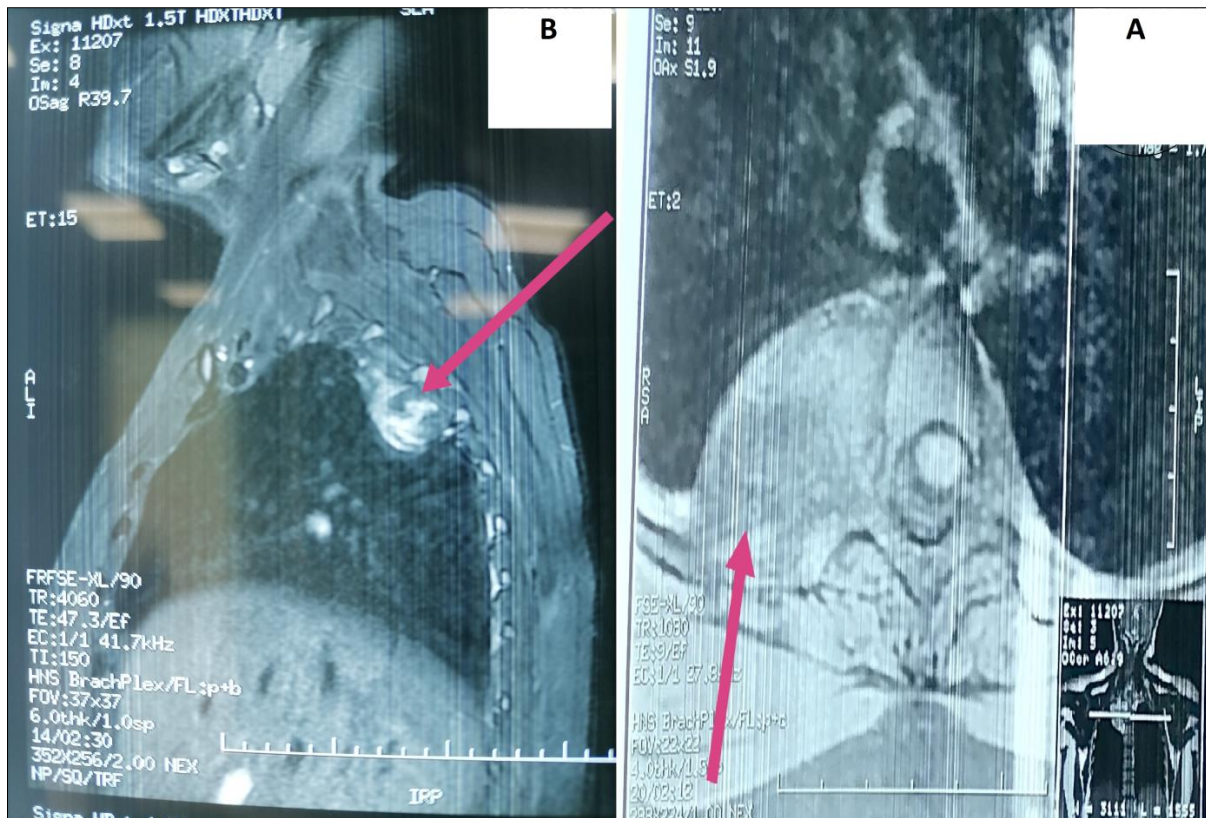


Figure 2: Chest MRI: pleural lesion with heterogeneous signal well limited in T1 and T2, hypointense on T1, hyperintense on T2 weighte with double component adipose and tissue without extension of the intraspinal canal

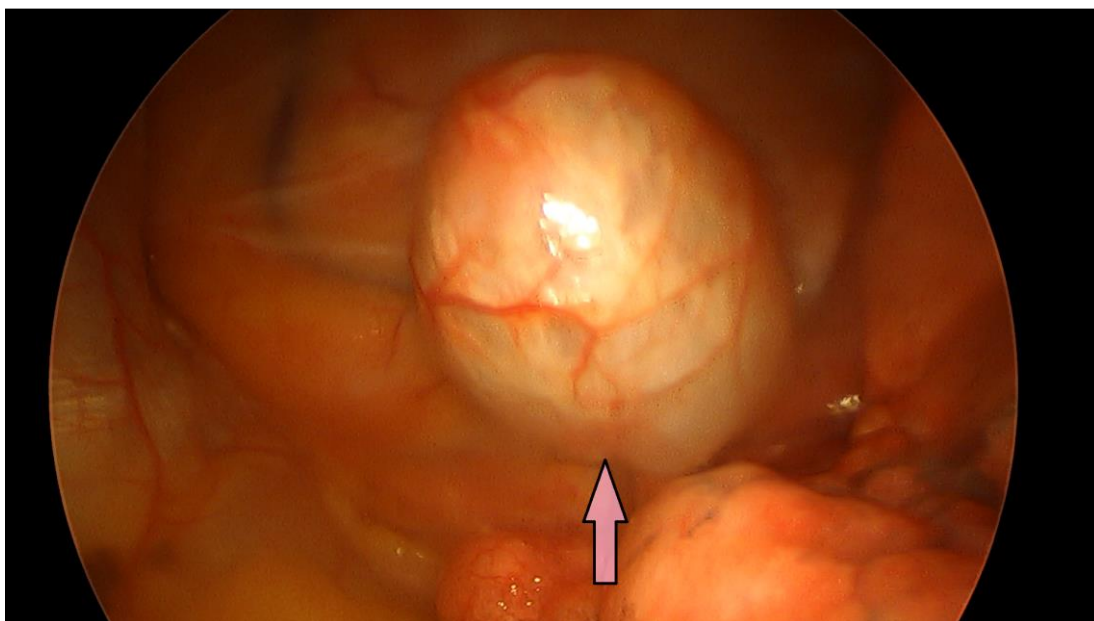


Figure 3: Per operative U-VATS image of the lesion

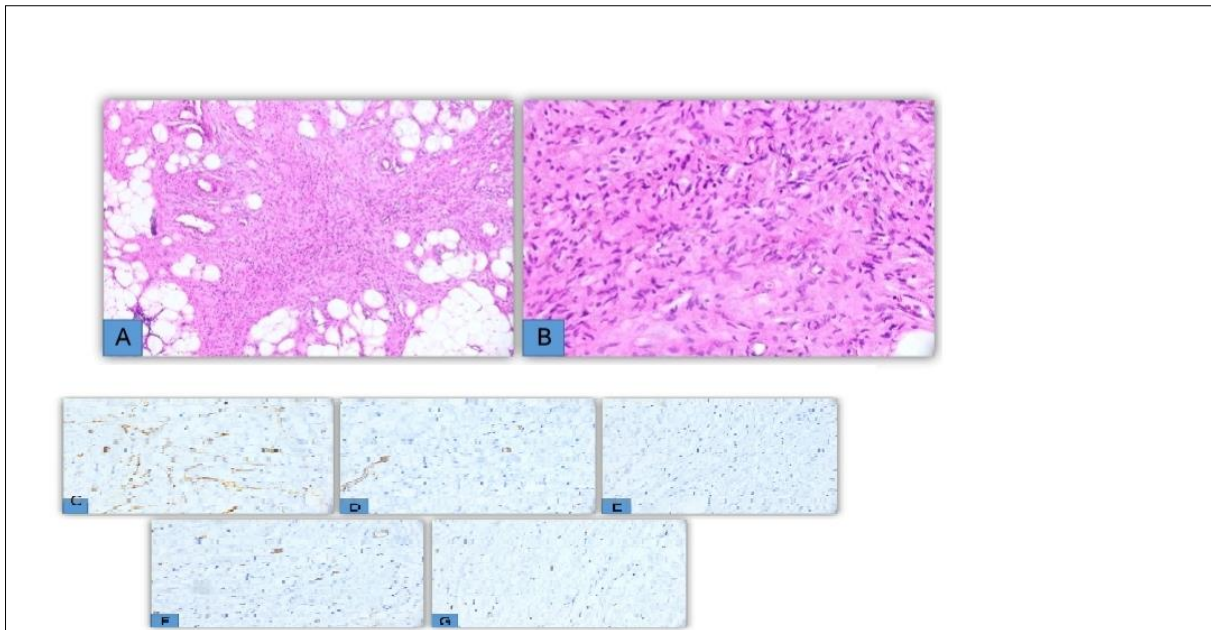


Figure 4: Histopathology and immunohistochemistry.: The specimen is composed of spindle cell tumor cells without cytonuclear atypia expressing protein S 100 in immunohistochemistry. (H&E, magnification×40)

DISCUSSION

The pleura is a serous layer that covers the thoracic structures and is divided into visceral and parietal pleurae. The visceral pleura is innervated by the vagus and sympathetic trunks, while the parietal pleura receives its nerve supply from somatic, sympathetic, and parasympathetic fibers via the intercostal nerves. The diaphragmatic pleura is supplied by the branches of the phrenic nerves. Nerve sheath tumors can arise from these nerves with direct pleural involvement.

Primary Pleural Neurofibromas (PPNF) originate from Schwann cells and fibroblasts and may arise in any peripheral nerve. They most often affect patients between 20 and 30 years of age and have no sex predilection [6].

Neurofibromas are the most common benign peripheral nerve sheath tumors. They can have localized, diffuse, or plexiform growth patterns. The localized form can occur sporadically. However, diffuse and plexiform forms are associated with neurofibromatosis type I. While mediastinal involvement is common in both adults and children, pulmonary and pleural involvement is rarely seen. Due to the presence of peribronchial and periarterial neural plexuses, few cases of intraparenchymal nerve sheath tumors have been reported without any chest wall or pleural connection. Approximately 90% of the neurofibromas are localised and not associated with neurofibromatosis. Localised neurofibromas tend to be slow growing and asymptomatic, although they may expand and cause symptoms due to compression of nerves or other adjacent structures [7].

Benign nerve sheath tumors encountered in the thorax include schwannomas, ganglioneuromas, and neurofibromas. Ratbi *et al.*, studied 17 patients who underwent resection of mediastinal benign neurogenic tumors and found schwannoma in 76.4% cases. The rarity of intrathoracic neurofibromas was further confirmed in a study of 149 intrathoracic neurogenic tumors by Bicakcioglu *et al.*, [4]. To date, three cases of localized benign PPNF have been reported in the English literature, 2 cases by Langman *et al.*, [8], including a known case of neurofibromatosis type I. Krishnamurthy *et al.*, [1], reported another case of isolated pleural neurofibroma in a 39-year-old woman who presented with worsening chest pain. Another case of a 31-year-old man with pleural neurofibroma was reported by Gupta *et al.*, [9]. The last case was reported by Sharma *et al.*, in 2021 in a 23-year-old man suffering from progressively worsening chest pain [10]. Our case is the 5th case in English literature and the last to our knowledge.

Differentiating a neurofibroma from a schwannoma and solitary fibrous tumor may sometimes be challenging. The presence of mast cells, which is integral in the development of neurofibromas, is one of the most specific histopathological features that help clinch the diagnosis of neurofibroma. Immunohistochemistry may also be helpful. Neurofibromas are invariably positive for S-100 protein and negative for STAT6 and bcl-2, in contrast to solitary fibrous tumors which are negative for S-100 protein and positive for STAT6 and bcl-2.

In our case, the tumor cells are positive for S-100 protein and negative for bcl-2 and STAT 6, ruling out solitary fibrous tumor. Schwannomas, like neurofibromas, are positive for S-100 protein and SOX-

10, and may show focal CD 34 positivity while being bcl-2 negative, thus making them harder to discern from neurofibromas [6-12]. Localisation of staining for neurofilament protein may be of some value in differentiating the two. Neurofibromas grow within the nerve of origin; accordingly neurofilament protein will stain axons entrapped within the tumor. Whereas, schwannomas grow peripherally and displace the nerve of origin, hence staining for neurofilament protein will fail to reveal any entrapped axons within the tumor though occasionally axons may be entrapped at the very periphery. However, some recent studies suggest that intralesional axons may be more frequently seen in schwannomas than previously reported. Pleural schwannomas are extremely rare neoplasms of the thoracic cavity, and less frequently in the pleura, with around 20 cases reported in the medical literature.

Unlike primary pleural neurofibromas (PPNF), which affect young patients and have no sex predilection, schwannomas tend to affect people aged 50 to 60 years. Additionally, men are affected more often than women [13].

Schwannomas grow slowly and, since they are generally asymptomatic, other differential diagnoses cannot be considered; however, larger tumours may compromise adjacent structures and cause cough, dyspnoea, and chest pain. Therefore, the vast majority of pleural schwannomas are discovered incidentally during investigations for other complaints [14].

Presenting symptoms largely depend on the size and location of these tumors and are non-specific (chest pain, dyspnea, and deformities). Those associated with neurofibromatosis will show other systemic manifestations of the syndrome. Pleural effusion mainly occurs due to mass effect and resultant lymphatic obstruction. Cross-sectional imaging plays a crucial role in the diagnosis and management of pleural masses. Careful assessment of the nature of the lesion, origin, and ancillary imaging findings are useful in narrowing the differentials.

The preferred treatment is surgical, consisting in local resection of the tumor. Stereotactic radiosurgery is useful if the growing tumour is near vital blood vessels or nerves [15, 16]. Different surgical approaches have been described for primary benign pleural tumors, according not only to the period and the availability of technologies, but also, especially in recent times, to the dimension and the organ infiltration. Open surgery has been considered the gold standard approach; thoracotomy, sternotomy, or even hemi or complete clamshell, have been described in the literature. Posterolateral was reported as the preferred access in different series in cases of larger tumors [17].

In minimally invasive surgery, both VATS and RATS have been gradually introduced, given the known

advantages in reducing postoperative pain and in-hospital stay. Minimally invasive surgery, even using a single port, Uniportal VATS for example can be offered in case of tumors of limited dimension, as it was done in our case which allow safe and proper intraoperative manipulation, but the indication for the use of a minimally invasive technique is strictly related to the skills and knowledge of surgeons.

In all recent literature, the authors found that patients treated with VATS had a significantly smaller diameter and a faster postoperative course compared to those treated with thoracotomy. Cardillo reported a conversion rate of 14.5% from VATS to open. It must be noted that over time, the use of minimally invasive surgery has largely developed, and the conversion rate might have dramatically reduced.

Nonetheless, patient safety and radical resection should be the priority for possible conversion from minimally invasive to open surgery [18–20]. Removal of the specimen in VATS and RATS must be performed with attention and by using a bag or by covering the utility incision with a soft tissue retractor in order to avoid possible contact metastases, which has been described by some authors. In cases of large pleural tumors, in order to avoid an enlargement of the minimally invasive surgical access and consequent injuries to the intercostal nerves, Hatooka suggests use of a subxiphoid access [20].

Prognosis is excellent and recurrence after resection is uncommon thanks to internal growth of PPNF, and complete excision of the neoplasm requires sacrifice of the nerve. This is acceptable in cases of pleural neurofibroma because they do not involve the major nerves [21].

CONCLUSION

Through this rare case, we emphasize that PPNF must be diagnosed in any spindle cell neoplasm of the pleura, regardless of sex, age, or the absence of symptoms. The main differential diagnosis includes schwannomas and solitary fibrous tumors which are excluded by histopathology supplemented by immunohistochemistry. Complete surgical resection is curative.

Abbreviations

PPNF: Primary pleural neurofibroma

U-VATS: Uniportal VATS

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflicts of Interest: The authors disclose no conflicts.

Sources of Funding: The authors disclose no sources of funding for research.

Provenance and Peer Review: Not commissioned, externally peer-reviewed.

Ethical Approval: N/A

Author Contribution

S. HAFIDI: study design, data collection, data analysis, and writing.

N. ID EL HAJ: study design, data analysis.

E.NIYONKURU: data collection, data analysis.

M.KARKOURI: study design, data collection, and analysis.

S.BOUBIA, M.KARKOURI, M.RIDAI: data analysis and reviewers.

Research Registration: N/A

Guarantor: Dr. Sara HAFIDI

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