CASE REPORT

Rare Case of Primary Pleural Spindle Cell Sarcoma in a Young Patient

Palaniappan Inbamuthiah[1], Rajesh Gupta[2], Mohamed Alhaj Mustafa[3], Desh Deepak Idnani[4], Imad Eldin Ahmed Hamed[5], Anandi Damodaran[6]

Received on: 14 December 2022; *Accepted on:* 31 March 2023; *Published on:* 12 May 2023

Abstract

Aim and background: This study presents a rare interesting case of primary pleural sarcoma in a young patient.

Case description: A 22-year-old Ethiopian male, with no significant past medical history, presented with fever, dry cough, and right-sided chest pain of 4-day duration. The chest X-ray showed moderate right-side effusion. Pleural tap revealed yellowish slightly turbid fluid that was lymphocyte predominantly exudate with high lactate dehydrogenase (LDH), normal glucose and negative malignant cell cytology. Computerized tomography (CT) chest showed mild-to-moderate right-sided pleural effusion with subsequent underlying right basal lung collapse associated with multiple scattered calcified right pleural plaques. Laboratory investigations interestingly showed high alkaline phosphatase. He presented again after 10 days with a massive right pleural effusion. Diagnostic thoracoscopy was done and a pleural biopsy revealed malignant spindle cells in a haphazard pattern, forming neoplastic bone and osteoid with foci of necrosis. Immunohistochemistry was consistent with malignant spindle cell sarcoma of pleura. Follow-up revealed the patient succumbed to illness within a few weeks before further evaluation and chemotherapy could be done.

Conclusion and clinical significance: Primary pleural sarcoma is a rare tumor with a poor prognosis and has little past literature review. Diagnosis might be missed and early thoracoscopic biopsy and immunohistochemistry are usually necessary for diagnosis. Presentation in a young patient, presence of pleural calcification in CT scan, elevated alkaline phosphatase, and evidence of new bone formation in histopathology were interesting and unique features seen in this case.

Keywords: Pleural malignancy in young, Pleural tumors with calcification, Primary pleural sarcoma, Rare pleural malignancies, Spindle cell sarcoma.

Bengal Physician Journal (2023): 10.5005/jp-journals-10070-8000

Introduction

Malignant pleural effusion is a common clinical occurrence, the diagnosis of which in some cases may prove challenging. It may be the presenting symptom in many cases.[1] They can be associated with a wide variety of malignancies and the metastatic pleural effusions being more common. Lung carcinoma is the most common primary tumor, followed by breast and stomach carcinomas.[2,3] Metastatic pleural effusion can remain as unknown primary in about a tenth of cases. Primary pleural malignancies are less common and malignant mesothelioma is the most well known and studied. Other primary pleural tumors are rare. We present a rare case of primary pleural spindle cell sarcoma occurring in a young patient.

Case Description

Our patient was a 22-year-old Ethiopian national, male, with no significant past medical history, who presented with fever, dry cough, and right-side chest pain of 4-day duration. There was no history of weight loss or recent travel. He was an occasional smoker and alcoholic, and driving was his occupation.

Initial clinical examination and chest X-ray showed moderate right-side effusion. Pleural tap revealed yellowish slightly turbid fluid that was lymphocyte predominant, exudative and with high lactate dehydrogenase (LDH), and normal glucose. Malignant cell cytology was negative. A computerized tomography (CT) chest was done which showed mild-to-moderate right-sided pleural effusion with subsequent underlying right basal lung collapse associated with multiple scattered calcified right pleural plaques (Fig. 1). Lab investigations interestingly showed high alkaline phosphatase.

In about 10-day time, the patient presented again with a massive right pleural effusion (Fig. 2). Diagnostic thoracoscopy

[1,2]Department of Internal Medicine, Kuwait Hospital, Sharjah, United Arab Emirates

[3]Department of Thoracic Surgery, Kuwait Hospital, Sharjah, United Arab Emirates

[4]Anatomical Pathology Laboratory (Pure Health), Al Qassimi Hospital, Sharjah, United Arab Emirates

[5]Department of Internal Medicine, Kuwait hospital, Sharjah, United Arab Emirates

[6]Department of Neurology, Kuwait Hospital, Sharjah, United Arab Emirates

Corresponding Author: Palaniappan Inbamuthiah, Department of Internal Medicine, Kuwait Hospital, Sharjah, United Arab Emirates, Phone: +0568431509, e-mail: drpalanipulmo@gmail.com

How to cite this article: Inbamuthiah P, Gupta R, Mustafa MA, *et al.* Rare Case of Primary Pleural Spindle Cell Sarcoma in a Young Patient. Bengal Physician Journal 2023;10(1):18–21.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

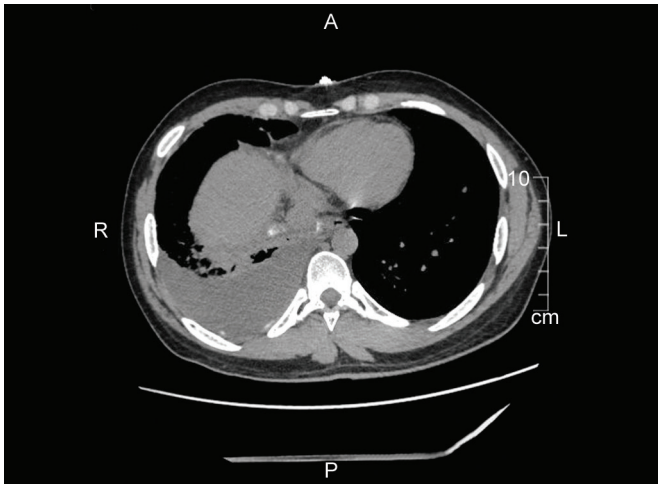

Fig. 1: CT chest cut showing moderate right pleural effusion with underlying lung collapse

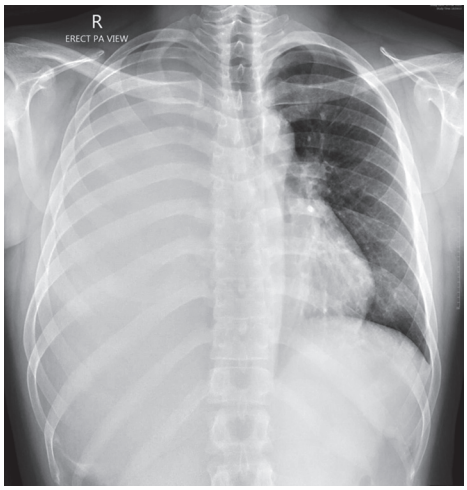

Fig. 2: Chest X-ray showing massive right pleural effusion

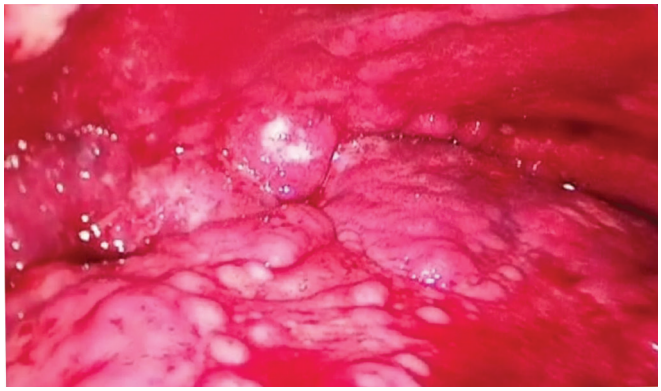

Fig. 3: Diagnostic thoracoscopy showing multiple pleural nodules and plaques and some of them were calcified

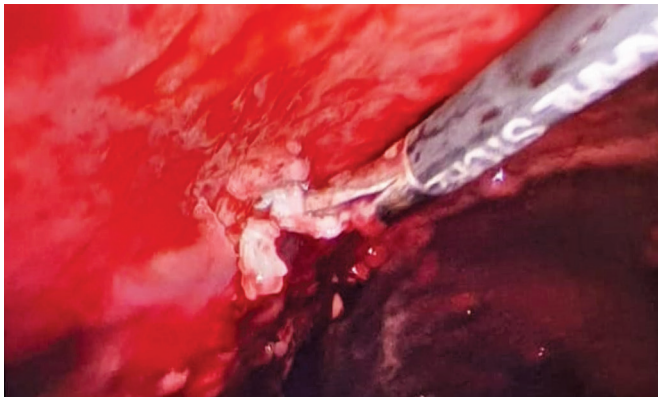

Fig. 4: Biopsy being taken from a pleural lesion

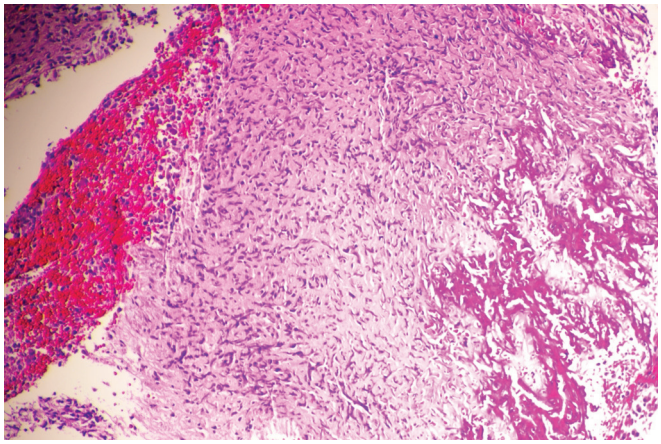

Fig. 5: Malignant spindle tumor cells in 10×—haphazardly arranged with new bone formation and foci of hemorrhage

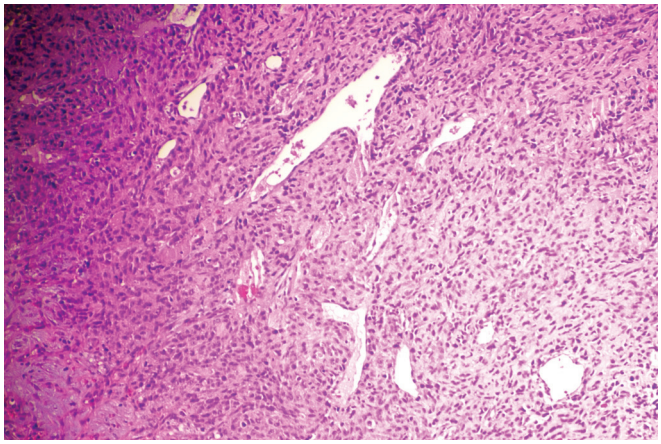

Fig. 6: Malignant spindle tumor cells in 10× (low power view)—haphazardly arranged

was done, which showed multiple nodules and plaques over the parietal and visceral pleurae, and some of them were calcified (Fig. 3). Pleural biopsies were obtained (Fig. 4). Histopathology revealed malignant spindle cells in a haphazard pattern, with hyperchromatic irregular nuclei, indistinct cytoplasm, and foci of malignant cells forming neoplastic bone and osteoid with foci of necrosis (Figs 5 to 10).

Immunohistochemistry Dako stains with adequate controls were done for calretinin/sma/cd34/s100/bcl2/ttf1/ck5/6/ck7/ckae1/ae3/wt 1. They showed negative staining, while those done with

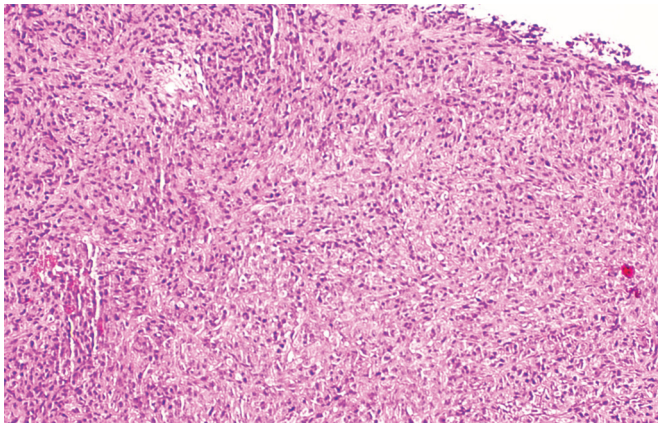

Fig. 7: Malignant spindle tumor cells in 10× (low power view)—haphazardly arranged

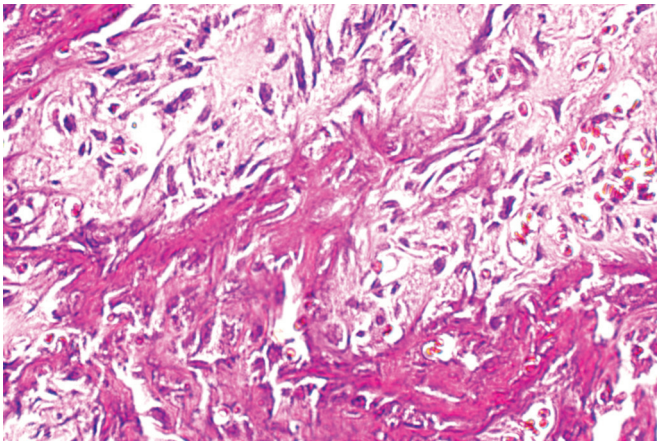

Fig. 9: Malignant new bone formation in 40× (high power view)

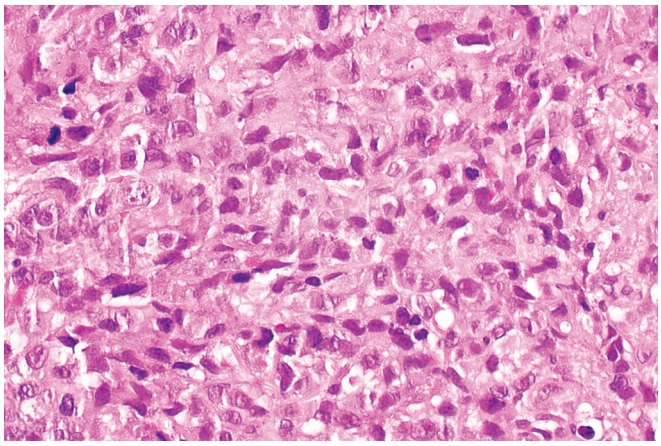

Fig. 8: Malignant spindle tumor cells in 40× (high power view)—haphazardly arranged with many abnormal mitosis

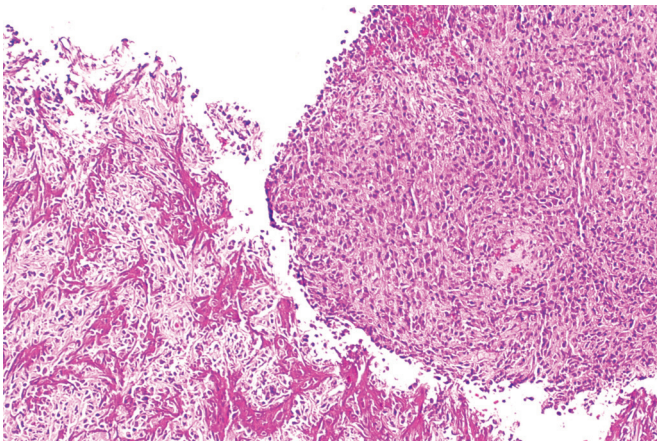

Fig. 10: Malignant spindle tumor cells in 10× (low power view)—haphazardly arranged with a new bone formation on the left side

vimentin/cd99 showed positive staining. KI67—high proliferation index was >70%—consistent with malignant spindle cell sarcoma of pleura. Samples were negative for acid-fast bacilli (AFB) staining and tuberculosis polymerase chain reaction (TB PCR). The patient was referred to the oncology center for further management. Follow-up revealed the patient succumbed to illness within a few weeks before further evaluation and chemotherapy could be done.

Discussion

Primary pleural malignancies other than malignant mesothelioma are rare and not well known. According to Christopher et al.,[4] malignant mesothelioma constitutes >90% of the primary malignant pleural tumors. Other reported primary pleural tumors include pleural liposarcoma[5] and localized fibrosarcoma. Most of these tumors present with pleural effusion, which may progress to massive effusion rapidly. Primary spindle cell tumors of pleura are a rare occurrence and need to be differentiated from tumors like spindle cell carcinoma, sarcomatoid mesothelioma, spindle cell melanoma, fibroma, and synovial sarcoma.[6–8] Diagnosis will usually need a thoracoscopic biopsy and immunohistochemistry.

Spindle cell sarcomas are extremely rare tumors that originate from mesenchymal cells and usually present as painless mass lesions in extremities or in the head and neck. Less commonly they are known to arise in the breast, reproductive system, viscera, lungs, and nervous system. They can occur in all age groups, but more commonly in late middle and old age groups. According to Feng and Wang[9] the mean age of diagnosis is 61. The treatment modalities include surgery, adjuvant radiotherapy, and chemotherapy with surgical resection being the mainstay treatment modality. Poor prognostic factors include age >64, primary tumor site being an internal site or viscera, and advanced stage.[9]

In our patient, spindle cell sarcoma had an uncommon presentation at a young age. Also, histopathology revealed malignant cells showing a new bone formation and osteoid, which correlated with elevated alkaline phosphatase and pleural calcification. The patient had an aggressive and rapidly progressive course with a poor prognosis.

Conclusion

Rapidly progressive and/or recurrent pleural effusions even in young patients should arouse suspicion and make us consider thoracoscopic biopsy and immunohistochemistry.

Primary spindle cell tumors are a rare entity. Usually they occur later in age, in the sixth decade. Primary pleural spindle cell sarcoma presenting at a young age with features of new bone formation and the aggressive course was a unique clinical presentation, and as

per our literature review so far, no similar case has been reported. This case is presented for its rarity and uniqueness.

References

1. Monte SA, Ehya H. Positive effusion cytology as the initial presentation of malignancy. Acta Cytol 1987;31(4):448–452. PMID: 3604540.
2. DiBonito L, Falconieri G. The positive pleural effusion. A retrospective study of cytopathologic diagnoses with autopsy confirmation. Acta Cytol 1992;36(3):329–332. PMID: 1580116.
3. Ong KC, Indumathi V. The diagnostic yield of pleural fluid cytology in malignant pleural effusions. Singapore Med J 2000;41(1):19–23. PMID: 10783675.
4. Christopher T Erb, Kelsey M Johnson. Rare pleural tumors. Clin Chest Med 2013;34(1):113–136. DOI: 10.1016/j.ccm.2012.12.001.
5. Prabhakar N, Vaiphei K. Primary pleural liposarcoma: A rare entity. Lung India 2019;36(5):438–440. DOI: 10.4103/lungindia.lungindia_246_18.
6. Boyraz B, Hung YP. Spindle cell tumors of the pleura and the peritoneum: Pathologic diagnosis and updates. APMIS 2022;130(3):140–154. DOI: 10.1111/apm.13203.
7. Travis WD. Sarcomatoid neoplasms of the lung and pleur. Arch Pathol Lab Med 2010;134(11):1645–1658. DOI: 10.5858/2010-0086-RAR.1.
8. Rdzanek M, Fresco R. Spindle cell tumors of the pleura: differential diagnosis. Semin Diagn Pathol 2006;23(1):44–55. DOI: 10.1053/j.semdp.2006.06.002.
9. Feng L, Wang M. Spindle cell sarcoma: A SEER population-based analysis. Sci Rep 2018;8(1):5024. DOI: 10.1038/s41598-018-23145-4.