Case Report

# Bilateral spontaneous pneumothorax in osteogenic sarcoma due to occult pulmonary metastases

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## ABSTRACT

Spontaneous pneumothorax (SPTX) is an uncommon complication of osteogenic sarcoma. Most of these SPTX associated with osteogenic sarcoma are due to detectable pulmonary metastasis. Development of SPTX without any evidence of pulmonary metastasis is extremely rare. Herein, we report a case of a young girl with osteosarcoma of the right femur that developed pneumothorax on both sides following treatment with combination chemotherapy without any obvious pulmonary metastases, bullae, or blebs.

**Key words:** Osteogenic sarcoma, pulmonary metastasis, spontaneous pneumothorax

## INTRODUCTION

Spontaneous pneumothorax (SPTX) is an uncommon complication of pulmonary metastasis. It represents <2% of whole cases of SPTX. It is more commonly occurring phenomenon in sarcomatous malignancies, particularly osteogenic sarcoma as compared to carcinomas. Development of pneumothorax is associated with impaired quality of life and increased risk of death within 2 years.[1] The majority of reported cases are secondary to detectable pulmonary metastatic lesions. Pneumothorax in the absence of pulmonary metastatic lesion is extremely uncommon and is limited to only few case reports[2,3] Here, we report a case of young girl with osteogenic sarcoma of the right femur that developed SPTX consecutively on both sides with no evidence of pulmonary metastasis or other parenchymal abnormalities. On follow-up after 5 months of development of pneumothorax, she developed bilateral metastases on follow-up.

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**Access this article online**

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.197862

## CASE REPORT

A 17-years-old school girl presented with complaints of pain and swelling at lower part of right thigh for 2 months, weight loss and loss of appetite for 1 month. Radiology (X-ray and magnetic resonance imaging) was suggestive of osteogenic sarcoma of lower third of right femur [Figure 1] which was further confirmed histopathologically [Figure 2]. She was planned for amputation of the primary tumor, but her parents refused for it. Then, she received the first cycle of combination chemotherapy (cisplatin and doxorubicin), and long-term central venous port (chemoport) was implanted in subcutaneous tissue of right chest wall for internal jugular venous access on following day of chemotherapy. She complained of acute onset dyspnea and left-sided chest pain on the next day of chemoport insertion. On examination, the intensity of breath sounds was significantly diminished over left hemithorax, and hyper-resonant note was elicited on percussion. Chest X-ray was advised which revealed pneumothorax on the left side with near complete collapse



**Cite this article as:** Kuwal A, Dutt N, Chauhan NK, Pareek P, Saini LK, Samdariya S. Bilateral spontaneous pneumothorax in osteogenic sarcoma due to occult pulmonary metastases. Clin Cancer Investig J 2016;5:483-5.

of left lung [Figure 3]. Intercostal tube thoracostomy was done which removed on the 10th day after complete expansion of the lung. Three days later, she again developed pneumothorax on the same side. Chest tube was re-inserted which results in partial re-expansion of the lung, and there was free air leak. Negative suction was applied, and chest tube remained *in situ* for 45 days. Pleurodesis was performed using doxycycline following complete expansion of the lung. Computed tomography (CT) scan of the thorax was performed which did not reveal any metastatic lesion.

After the second dose of chemotherapy, the patient again developed pneumothorax but this time was on right side. It was initially managed conservatively with high flow oxygen, but no improvement was observed. Tube thoracostomy was then performed and pleurodesis was also done in view of future recurrence.

She received three cycles of three weekly cisplatin and doxorubicin combination chemotherapy. It resulted in a good partial response. Further, she underwent mid-thigh amputation of the limb. Bone marrow margins were infiltrated by tumor. Then, she was started on external beam radiation therapy to the amputated stump by conventional fractionation.

On follow-up (after 5 months of first chemotherapy), contrast enhanced CT thorax revealed multiple metastatic lesions on both sides [Figure 4]. Because of poor performance status and general condition, she was discharged on best supportive care.

## DISCUSSION

SPTX is an uncommon complication of osteogenic sarcoma. It is frequently associated with increased risk of mortality as <10% of patients survive more than 2 years who developed SPTX. Barrin reported the first case of SPTX in a patient of bone sarcoma with pulmonary metastasis.[4] The true prevalence of SPTX in osteogenic sarcoma is difficult to establish as most of the literature is in the form of case

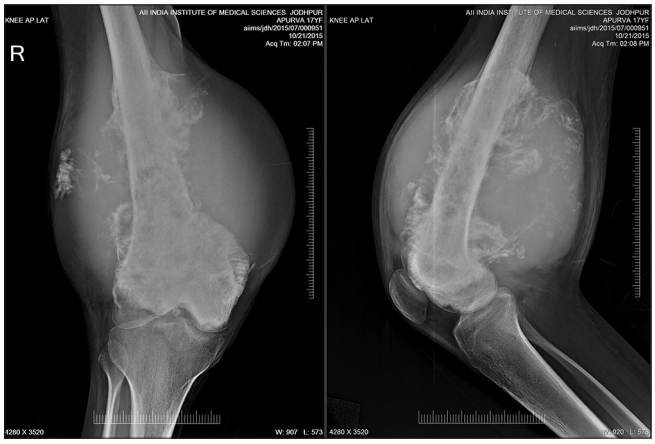


**Figure 1:** Anteroposterior and lateral view of lower thigh region shows large soft tissue mass lesion with ill defined, moth eaten destruction of lower femur having mixed sclerotic and lytic areas. There is marked periosteal reaction and new bone formation

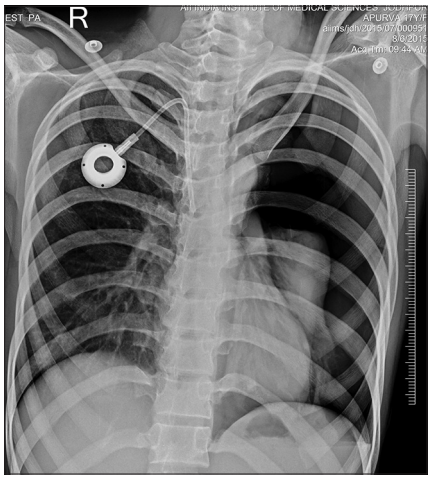


**Figure 3:** Chest radiograph posteroanterior view showing pneumothorax on left side and subcutaneously implanted chemoport on right

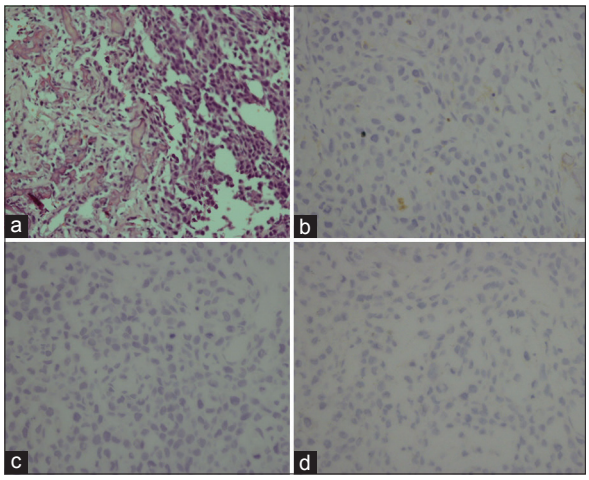


**Figure 2:** (a) Malignant osteoid surrounded by malignant cells with morphology of a small cell osteosarcoma (H and E × 20). (b-d) Immunohistochemistry with HMB-45 antibody, pan cytokeratin antibody, and CD99 antibody exhibiting tumor cells negativity, respectively (×40)

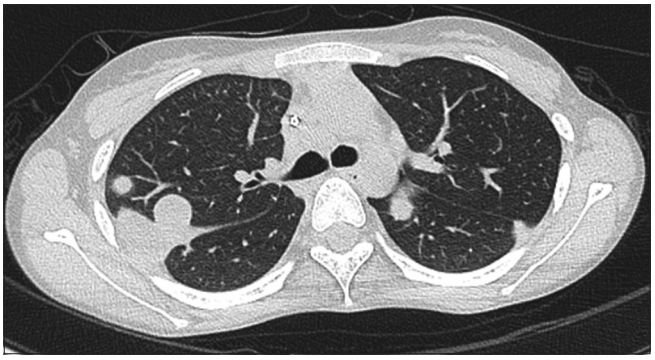

**Figure 4:** Contrast-enhanced computed tomography thorax showing well defined, multiple rounded nodular lesions over both lung fields with thickening of right major fissure (follow up)

reports or case series. However, Bergin has observed the prevalence of 5% among their series of twenty patients.[5] A systemic review on SPTX complicating sarcoma reported the prevalence of 1.9% and osteogenic sarcoma was the most common cell type associated with SPTX among these cases.[1]

Our case developed pneumothorax on the left side following insertion of chemoport on contralateral side and received combination chemotherapy (cisplatin and doxorubicin) 1 day before developing pneumothorax. Postchemoport pneumothorax usually develops on the side of central venous placement. Although there is a single reported case of bilateral spontaneous pneumothoraces which is at least theoretically possible due to contralateral bullae rupture or congenital intrapleural communication, the development of contralateral pneumothorax has neither been reported nor seems logical.[6] Thus, we can safely assume that the present pneumothorax was not a complication of placement of chemoport.

Several pathogenic mechanisms have been postulated regarding the development of SPTX in patients with osteogenic sarcoma which includes direct pleural involvement by tumor, cavitation of peripheral metastatic lesion and treatment with chemotherapeutic agents. A retrospective study of patients with the osteogenic sarcoma and testicular tumor observed the risk of SPTX of 7% which was doubled after introduction of chemotherapy.[7] Yamamoto and Mizuno reported a case SPTX in a young girl with osteogenic sarcoma femur without any detectable pulmonary metastases. On follow up, she developed metastatic lesions 6 months following surgery of the limb.[2] Another case series observed the development of SPTX after receiving chemotherapy even though none of the patients had prior evidence of pulmonary involvement.[3] The present case also had no radiological pulmonary involvement at the time of development of pneumothorax. On follow-up, the patient developed metastatic lesions in both the lungs so the necrosis of peripherally located micro-metastatic lesion following chemotherapy could be the appropriate explanation for the development of SPTX rather than direct toxicity of chemotherapeutic agent.

Management of SPTX in these cases is difficult as they have increased the frequency of recurrence. Tube thoracostomy alone is an ineffective mean for prevention of recurrence in these cases and results only partial re-expansion of the lung. This is probably because of the development of bronchopleural fistula (BPF) secondary to tumor necrosis following chemotherapy and rupture into pleural space. In our case also, the patient developed SPTX with BPF on right side and required chest tube placement for long duration. Surgical closure can be an option for management of BPF, but these patients are usually poor surgical candidates which further ads morbidity. Pleurodesis with negative suction through chest tube is an acceptable approach in these patients. Our patient suffered a recurrence of SPTX on both sides requiring tube thoracostomy and pleurodesis every time.

## CONCLUSION

SPTX is an uncommon complication of osteogenic sarcoma that is associated with recurrence and increased morbidity. SPTX can be developed even in the absence of detectable pulmonary metastasis. Therapeutic options are also limited in such cases because of poor general condition of these patients. Pleurodesis should be performed in the first episode as they suffered a recurrence. Further prospective studies are needed to address risk factors, prognosis, and appropriate treatment modality in such cases.

### Acknowledgment

We are grateful to Dr. Shilajit Bhattacharya, Professor and Head, Department of Pathology All India Institute of Medical sciences, Jodhpur for providing their valuable support.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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