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Malignant fibrous histiocytoma presenting with complete opacification of the hemithorax: A case report



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ABSTRACT

BACKGROUND: Malignant fibrous histiocytoma, a subtype of primary lung sarcoma is a very rare disease. It usually presents as a lung nodules and the final diagnosis is made by immunohistochemical studies.

METHODS: A 45-year-old patient presented with progressive dyspnea, dry cough and right shoulder pain. Chest X-ray revealed complete opacification of the right hemithorax. Chest computed tomography confirmed the presence of a heterogeneous lesion occupying the whole right hemithorax causing a mass effect on the trachea. Ultrasound guided biopsy was done and final pathology was suggestive of malignant fibrous histiocytoma.

CONCLUSION: Progressive dyspnea in young otherwise healthy patients should be investigated early on. In our case the presence of right shoulder pain indicates advance disease illustrated by the singular imaging findings.

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1. Introduction

Malignant fibrous histiocytoma (MFH) of the lung, a subtype of primary lung sarcomas, is a very rare tumor with only 50 cases reported in the literature.¹ It is an aggressive tumor with high potential of local relapse and distant metastases. The cell origin of MFH remains controversial.¹ The majority of MFH of the lung described presented with a solitary pulmonary mass or nodule.² To the best of our knowledge this is the first case to be reported with a tumor occupying the entire hemithorax. The final diagnosis is based on microscopic and immunohistochemical studies. The treatment of choice is surgical resection with negative microscopic margins.³ The role of adjuvant radiotherapy and chemotherapy has not definitely been defined yet.³

2. Case presentation

A 45-year-old female patient, presented to our institution with a 3-month history of right shoulder pain, dry cough and progressive dyspnea. She denied any fever, chills or episodes of hematemesis. No recent weight loss was reported, no smoking. At admission, the patient's vital signs were stable with an oxygen saturation of 94% on

room air. Physical examination revealed decreased air entry over the right hemithorax with dullness to percussion.

Chest X-ray (Fig. 1) showed complete opacification of the right hemithorax with consolidation in the left lower lobe. A chest computed tomography (Figs. 2 and 3) confirmed the presence of a heterogeneous lesion causing a mass effect on the trachea with

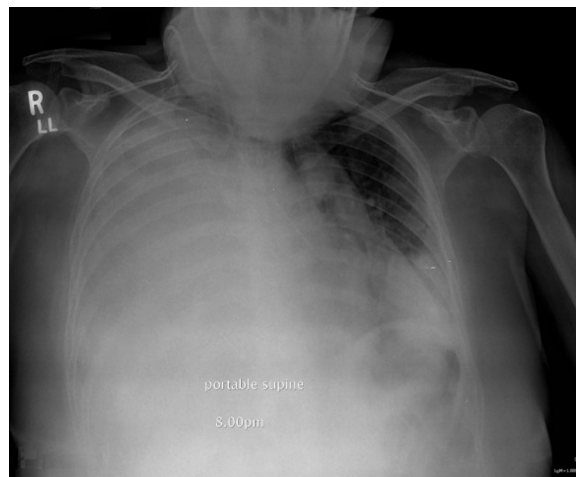


Fig. 1. Chest X-ray in supine position showing complete opacification of the right hemithorax with consolidation in the left lower lobe.

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Fig. 2. Computed tomography of the chest in coronal view showing a heterogeneous lesion in the right hemithorax causing total lung collapse with mass effect on the trachea with areas of fluid attenuation and other areas of soft tissue attenuation.



Fig. 3. Computed tomography of the chest in axial view showing the tumor on the right side and a left pleural effusion.

areas of fluid attenuation and other areas of soft tissue attenuation. Soft tissue density is seen along the right hilum representing collapsed lung. An ultrasound guided biopsy was performed and pathology revealed high grade sarcoma with positive smooth

muscle actin (SMA) and muscle specific actin (MSA) on immunohistochemistry. These findings were in favor of a malignant fibrous histiocytoma (MFH). The case was discussed in a multidisciplinary meeting and it was agreed that this tumor was unresectable and decision was to go for chemotherapy (adriamycine and ifosfamide). Unfortunately she did not proceed with her treatment and was lost for follow-up.

3. Conclusion

This case illustrates the importance of early work up of progressive dyspnea especially in young healthy patients since early diagnosis of any lung malignancy can lead to definitive treatment. Patients with suspected primary MFH of the lung must be carefully evaluated to rule out a metastatic origin, as MFH is primarily a soft tissue tumor of the extremities and is extremely rare to be found as primary tumor in the lung like in our case.

Conflict of interest

The authors declare that they have no conflict of interest.

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Ethical approval

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.

Author contributions

Jean Salem is the first author. Ali Shamseddine and Deborah Mukherji are co-authors who helped in designing the manuscript with Walid Faraj who is a senior author. Mohamad Khalife is a co-senior author who edited the manuscript along with Ghina El Nounou and Abdallah Abou EL Naaj. Ali Haydar is a radiologist and helped in image editing.

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