

Spontaneous massive hemothorax related to a neurofibroma: A case report

Slim Charfi¹ , Abdesslem Hentati² , Walid Abid² , Imed Frikha² 

¹Department of Pathology, CHU Habib Bourguiba, Sfax, Tunisia

²Department of Cardiovascular and Thoracic Surgery, CHU Habib Bourguiba, Sfax, Tunisia

Received: January 29, 2018 Accepted: May 21, 2018 Published online: April 24, 2019

ABSTRACT

A 33-year-old woman was admitted due to sudden-onset dyspnea and right-sided chest pain. Imaging studies revealed a right-sided hemothorax and an hyperdense mass of the posterior mediastinum. A *monobloc* and complete tumor resection was done by thoracotomy. The diagnosis of a neurofibroma was confirmed by the pathological exam. A careful examination of the patient did not suggest any signs of von Recklinghausen's disease. To the best of our knowledge, this is the first case of spontaneous massive hemothorax secondary to a neurofibroma in non-von Recklinghausen's disease.

Keywords: Hemothorax; neurofibroma; neurofibromatosis.

Spontaneous massive intra-thoracic bleeding is rare and life-threatening complication. It usually occurs due to vasculopathy. A concomitant mediastinal neoplasm related to spontaneous hemothorax is a rare finding.^[1] Herein, we report a case of spontaneous hemothorax secondary to a neurofibroma of the mediastinum in non-von Recklinghausen's disease.

CASE REPORT

A 33-year-old woman with a non-specific medical history was admitted to our hospital due to sudden-onset dyspnea and right-sided chest pain. Physical examination revealed tachypnea (respiratory rate: 44/min) and decreased breath sounds in the right lung base. No other abnormalities were noted. Laboratory tests showed a low hemoglobin level of 8.6 g/dL. Coagulation tests were normal. A chest radiograph revealed a large amount of right-sided pleural effusion (Figure 1). Thoracic computed tomography (CT) demonstrated a right-sided hemothorax and an hyperdense mass of the posterior mediastinum which was measured 146×126×114 mm in size with an intense contrast enhancement. A collapse of the right lung and a mediastinal shift to the left were noted (Figure 2).

A written informed consent was obtained from the patient and a transfusion of three units of blood was done, followed by a thoracotomy. During operation, an abundant hemothorax was noted with a bleeding

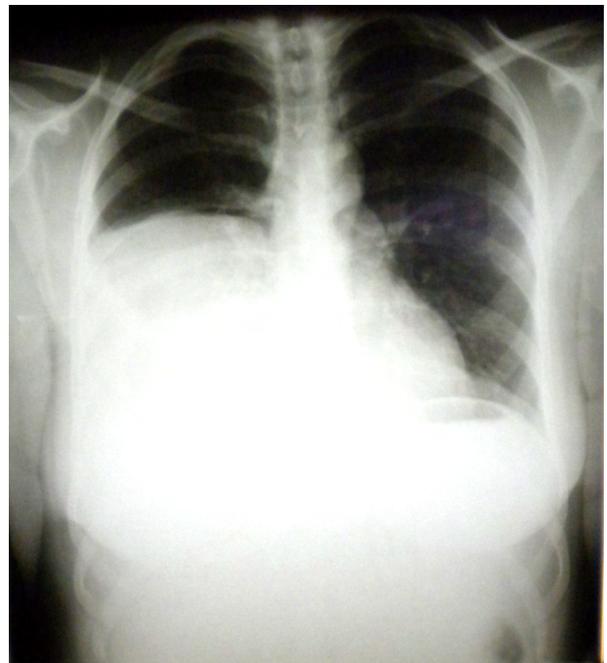


Figure 1. A chest radiograph showing large amount of right-sided pleural effusion.

Corresponding author: Slim Charfi, MD. Department of Pathology, CHU Habib Bourguiba, 3029 Sfax, Tunisia.

Tel: 00216 98 972 756 e-mail: charfislim@gmail.com

Citation:

Charfi S, Hentati A, Abid W, Frikha I. Spontaneous massive hemothorax related to a neurofibroma: A case report. *Cardiovasc Surg Int* 2018;5(1):9-11.

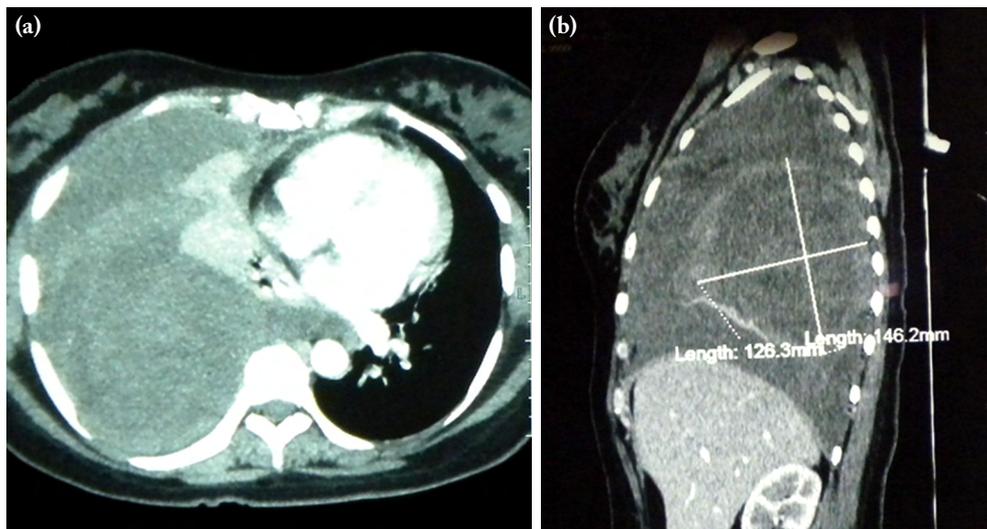


Figure 2. Thoracic computed tomography showing (a) massive hemothorax and (b) mass of the posterior mediastinum.

mass of the posterior mediastinum which extended through an intervertebral foramen. Therefore, a *monobloc* and complete tumor resection with removal of the pleural hematoma was performed. Pathological examination showed a bilobate and encapsulated mass measuring 14×11×11 cm in size. The cut surface was white-to-yellowish with myxoid and hemorrhagic areas. Histological examination revealed a low cellular spindle cells proliferation with no cellular pleomorphism and mitosis. The stroma was fibrous with myxoid areas. Immunohistochemical study showed that tumor cells were positive for PS100 and vimentin and negative for smooth actin muscle, desmin, and CD34. The diagnosis of a neurofibroma was considered. A careful examination of the patient did not suggest any signs of von Recklinghausen's disease. In addition, there was no café-au-lait spots, hyperpigmented macules, and axillary or inguinal freckles. Also, there was no history of first-degree relative with neurofibromatosis. The patient was discharged on Day 5. The follow-up was unremarkable.

DISCUSSION

Spontaneous massive hemothorax secondary to a neurofibroma is a rare and often lethal complication.^[1] Spontaneous massive hemothorax usually occurs secondary to pulmonary infarction, arteriovenous fistula, ruptured aneurysm, and tumors. Numerous benign and malignant tumors have been blamed for the development of spontaneous

massive hemothorax including angiosarcoma, chondrosarcoma, fibrosarcoma, peripheral neuroectodermal tumor, teratoma, and neural tumors.^[2-6] Neural tumors includes neurofibromas, schwannomas, malignant peripheral nerve sheath tumors, and ganglioneuromas.^[6,7] In the literature, all published cases of spontaneous massive hemothorax secondary to a neurofibroma have been shown to be related to von Recklinghausen's disease.^[1] In this context, this is the first case of spontaneous massive hemothorax secondary to a neurofibroma in non-von Recklinghausen's disease.

In conclusion, neurofibroma should be considered among neural tumors causing hemothorax even for patients without von Recklinghausen's disease.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Pulivarthi S, Simmons B, Shearen J, Gurram MK. Spontaneous hemothorax associated with neurofibromatosis type I: A review of the literature. *J Neurosci Rural Pract* 2014;5:269-71.
2. Varsano S, Edelstein E, Gendel B, Smorzik J. Bilateral and unilateral spontaneous massive hemothorax as a presenting manifestation of rare tumors. *Respiration* 2003;70:214-8.

3. Hadithi M, van Boxem TJ, Giaccone G, Postmus PE. Exercise-induced spontaneous hemothorax insinuates trauma; yet unmasks a latent disorder. *Neth J Med* 2001;59:292-4.
4. Shiota S, Nakaya Y, Sakamoto K, Iwase A, Aoki S, Matsuoka R, et al. Spontaneous hemothorax secondary to immature teratoma of the mediastinum. *Intern Med* 1999;38:726-8.
5. Larrieu AJ, Hashimoto SA, Allen P. Spontaneous massive haemothorax in von Recklinghausen's disease. *Thorax* 1982;37:151-2.
6. Vaziri M, Mehrzama M. Massive spontaneous hemothorax associated with Von Recklinghausen's disease. *Ann Thorac Surg* 2006;82:1500-1.
7. Fuyuno G, Kobayashi R, Iga R, Horio H, Nomori H, Koderu K, et al. A case of Von Recklinghausen's disease associated with a hemothorax due to a rapidly growing malignant schwannoma. *Nihon Kyobu Shikkan Gakkai Zasshi* 1995;33:682-5.