Spontaneous massive hemothorax related to a neurofibroma: A case report

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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...
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 14x11x11 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.

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