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CASE REPORT

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Inflammatory myofibroblastic lung tumor: its birth, its bleeding growth, its difficult diagnosis and its surgical end in a child

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Key Clinical Message

The diagnosis of inflammatory myofibroblastic tumor is based on radiology and histology. The treatment is surgical, and the prognosis is good. For this reason, although this lung disease is rare, when a child show up at hospital with an unknown hemoptysis, this medical condition should not be underestimated.

KEYWORDS

hemoptysis, inflammatory myofibroblastic lung tumor, primary lung neoplasm

An 11-year-old boy arrived at the emergency room with hemoptysis after waking up with dry cough. The child's clinical history revealed two other episodes of blood after coughing; in those cases, an otorhinolaryngologist examination was performed, with negative rhinoscopy. No past acute infectious disease, no impaired hemostasis, no intake of drugs or recent trauma. At the clinical examination in Emergency room, the child appeared pale and painful with respiratory rate 40 acts per minute and hypophonesis at pulmonary apex. Heart rate is 90 beats per minute with normal electrocardiogram (ECG) and QTC values, sinus rhythm, 96% oxygen saturation. Blood tests: GB 4.48 × 10^9/L, VES 32 m/h, Hb 11.6 g/L. At X-ray evidence of epigraphic formation 8 × 5 cm at left lung (Figure 1). The chest CT scan confirmed an 8 cm diameter "massive inhomogeneous solid formation" in proximity to the left pulmonary upper lobe, which improved after contrast medium. Specifically, ground-glass opacity image indicates thickening at the middle and at the level of the lingula and seemed to be infiltrating pulmonary arteries, making the prognosis inoperable (Figure 2).

The child was admitted to the pediatric unit, and a thoracic surgical consultation was requested. The following step was the bronchoscopy, which did not reveal pathological characteristics. Taking into account the stable general condition, the absence of a severe anemia as well as the presence of normal values of the vital parameters, the patient underwent intravenous tranexamic acid therapy and was decided not to perform an urgent and hazardous surgical procedure, choosing waiting and seeing behavior. The day after, a new massive hemoptysis with hematemesis with spontaneous resolution accelerated diagnostic workup: an angio CT scan was performed and revealed that the tumor did not infiltrated pulmonary arteries, but bronchial ones, so that the prognosis suddenly changed. An extemporary biopsy of the lesion showed the presence of inflammatory cells; consequently, in the suspect of an infectious pathogenesis, the child was treated with antibiotics (piperacillin/tazobactam) and antifungal drugs. The definitive histologic diagnosis confirmed the presence of inflammatory cells, and the mass was revealed to be an inflammatory myofibroblastic lung tumor and not just an infection. The child underwent a lobectomy of left pulmonary upper lobe with a good follow-up at X-rays and blood tests.

This article reports the pediatric case of inflammatory myofibroblastic tumor, a rare benign tumor localized in the left lung, composed of spindle cell fibroblast and myofibroblast

proliferation with plasma cells, lymphocytes, and histiocytes.[1] This benign tumor was detected after an episode of moderate hemoptysis.[2] Due to the uncommon onset, it was challenging to establish the accurate diagnosis.

The term "hemoptysis" is referred to the expectoration of blood or the presence of blood in the sputum.[2] The bleeding can arise from both pulmonary arterial circulation and bronchial circulation. Understanding the dimension of the bleeding is essential because it changes the diagnostic workup[3]. Specifically, bleeding can be "scant" (<5 mL), "mild to moderate" (6–240 mL), or "massive" (more than 240 mL).[4] In children, principal causes of hemoptysis are as follows: respiratory infection, aspiration of foreign bodies, and bronchiectasis. However, hemoptysis can be rarely caused by vasculitis and associated syndromes, congenital heart disease and cardiac-related conditions, congenital lung malformations, pulmonary vascular disorders, pulmonary masses, trauma, idiopathic, coagulopathy and thrombosis, toxic inhalation, bone marrow transplant, pulmonary veno-occlusive disease, celiac disease, connective tissue disorders (Ehlers Danlos syndrome and Loeys Dietz syndrome), plasma cell granuloma, and catamenial hemoptysis (females).[5]

The management of the different cases of massive, moderate, and scant hemoptysis varies according to the condition. Massive hemoptysis represents a life-threatening condition that warrants urgent treatment. At first, the patient needs to be stabilized in order to prevent further bleeding. After the stabilization of the patient, the evaluation of the cause of the hemoptysis can be carried out.

Mild hemoptysis and moderate hemoptysis are more frequent and do not represent a medical emergency; in fact, they tend to resolve spontaneously. Their management should be oriented on the basis of an accurate history, focusing the attention on the presence of known underlying disease, physical examination, laboratory tests, such as complete blood count and coagulation study, and chest radiograph. Starting from these four steps, it is often possible to understand if the cause of hemoptysis has good prognosis and does not require further intervention or if it represents a pathologic condition, which requires a special medical attention. In this last case, it is necessary to perform other investigations, such as bronchoscopy, CT of the lung and eventually lung biopsy. Guidelines recommend to suspend chest physiotherapy, to avoid noninvasive ventilation, and to evaluate the possibility of sedation and cough suppression drugs,[4,6] until the cause of the hemoptysis is identified.

Inflammatory myofibroblastic lung tumor belongs to the category of primary neoplasm of the lung[1] and has a prevalence of 0,7%,[7] but, among these types of tumors, of about 25%. Inflammatory myofibroblastic tumor of the lung is made up by spindle cell fibroblast, myofibroblast proliferation with a mixture of plasma cells, lymphocytes, and histiocytes.[1] The major part arises from the lung; only 12% can arise from the bronchi. Pediatric patients are often asymptomatic, and symptoms such as cough, fever, shortness of breath, and hemoptysis are usually rare.[1] For what concerns instrumental diagnosis, the first step is chest radiography: specifically, inflammatory myofibroblastic tumor appears as a solitary pulmonary mass; rarely, atelectasis and pleural effusion are present.[1] Calcifications are common in pediatric patients, too.[1] In 5% of the cases, masses are multiple.[1] The second instrumental step is CT scan: calcifications are more evident and can direct the diagnosis.[1] MRI can be an alternative instrumental examination; in fact, inflammatory myofibroblastic tumors typically have low intermediate T1 signal and high T2 signal.[1] Another second instrumental step is fluorodeoxyglucose positron emission tomography: it can demonstrate inflammatory components.[1]

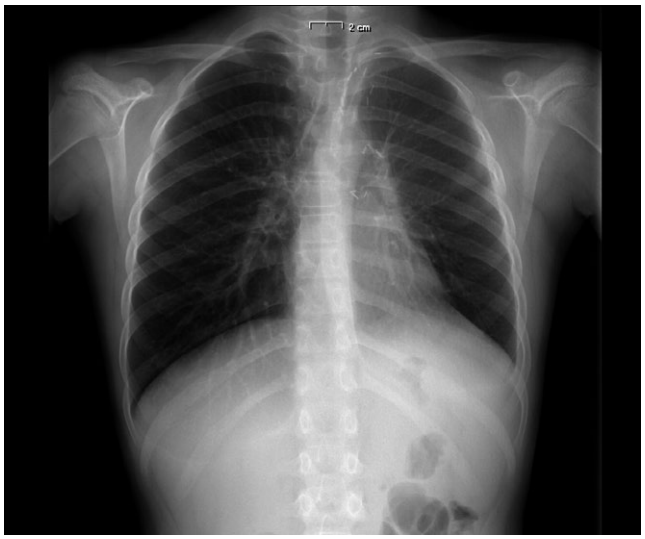

FIGURE 1 (X-ray): Left pulmonary upper lobe occupied by oval expansive formation of 8 × 5 cm

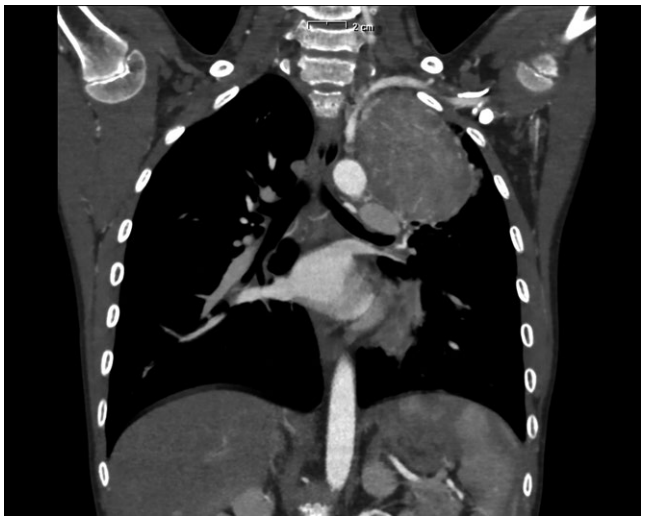

FIGURE 2 (Chest CT Scan): Massive inhomogeneous solid formation (inner diameter 8 cm) in proximity to the left pulmonary upper lobe, which improved after contrast medium

The treatment of inflammatory myofibroblastic tumor is surgical.[1] Surgery can define diagnosis of this type of tumor through the biopsy; specifically, only histological and immunohistochemical studies can confirm the diagnosis.[1]

Chemotherapeutic agents are not useful and do not change the prognosis.[8]

In fact, if the resection of myofibroblastic lung tumor is complete, it is a good signal and there is low risk for recurrence.[7]

AUTHORSHIP

LP: is the main author, he pointed out the idea, revised the literature, and drafted the article. MC and ET: contributed on the idea development and searched the literature and approved the article. GP, AP, and DD: drafted the article, developed the idea, searched the literature, and approved the manuscript in the final form.

CONFLICT OF INTEREST

None declared.

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