Complete resection of a huge hypervascular inflammatory myofibroblastic tumor in right hemithorax after embolization

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Case report

Background: **Inflammatory myofibroblastic tumor (IMT) is a rare and mostly benign tumor that has the possibility of malignant change.**

Methods: **Radiological findings revealed a huge mass that filled most of the right hemithorax of a 17-month-old female infant. Tumor extirpation was stopped due to massive bleeding and limited exposure of the tumor. Embolization was conducted to obstruct the arteries feeding the mass. Complete resection was performed.**

Results: **Histopathologic examination led to the diagnosis of IMT. Postoperative recovery was uneventful.**

Conclusions: **Hypervascularity of IMT should be considered. Preoperative embolization can be effective to reduce intraoperative blood loss and facilitate the surgical procedure.**

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Key words: **embolization; granuloma; mediastinal neoplasms; plasma cell**

Introduction

Inflammatory myofibroblastic tumors (IMTs) are rare, with an incidence of only 0.04-1% of primary lung neoplasms.[1,2] Pathological features are proliferative myofibroblastic spindle cells along with inflammatory cells that consist of plasma cells, lymphocytes and eosinophils. Although IMTs generally have a benign clinical course, aggressive local invasion and malignant changes have also been reported.[3,4]

We report a rare case of hypervascularity of a large mediastinal IMT that almost filled the right hemithorax. Because the small volume of the circulation of the infant resulted in significant hemodynamic instability, complete resection was performed after preoperative embolization.

Case report

A 17-month-old female infant who had no significant medical history was admitted with persistent fever, cough, sputum and weight loss of 1 kg over 3 months. Chest radiography showed that most of the right hemithorax was occupied with a huge mass (Fig. 1A). Chest computed tomography (CT) revealed a mass sized 10.2 cm × 7.5 cm × 6.7 cm with subtotal atelectasis of the right lung and mediastinal shifting to the left side, resulting in compression of both right and left atriums (Fig. 1B). No definite chest wall or bronchial invasion was found. Complete blood count demonstrated anemia (hemoglobin 5.5 g/dL), thrombocytosis (platelets 724 000/mm^3) and leukocytosis (white blood count 16 100/mm^3). Elevated erythrocyte sedimentation rate (ESR) and C-reactive protein were also found. Tumor markers such as ferritin and neuron-specific enolase (NSE) were normal.

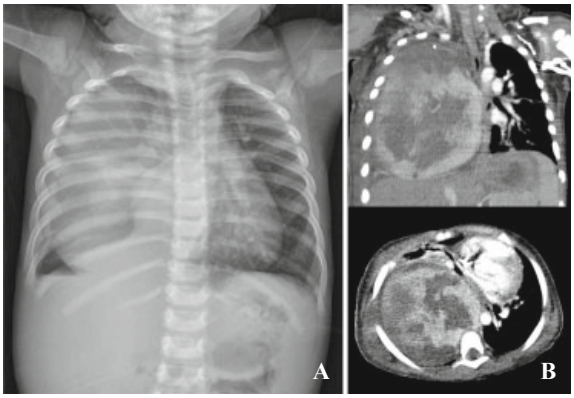

Fig. 1. A: Chest radiograph shows most of the right hemithorax occupied by a huge mass; **B**: Chest computed tomography reveals a 10.2 cm × 7.5 cm × 6.7 cm-sized mass with multifocal central necrosis, subtotal atelectasis of the right lung and a mediastinal shift to the left.

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We decided on surgical removal because the large tumor was compressing the heart. To prevent airway collapse and cardiovascular compression by the tumor if muscle tone was relaxed by a muscle relaxant, general anesthesia was performed while maintaining spontaneous ventilation without using a muscle relaxant. Despite 500 ml blood transfusion and volume replacement, more than 700 ml excess bleeding from hypervascular tumor wall caused hemodynamic instability during surgery. The difficulty of maintaining hemodynamic stability due to the excessive bleeding and limited exposure of the tumor made us stop the surgery after a biopsy specimen was obtained. Embolization was then planned to obstruct the arteries feeding the mass. Angiography revealed hypervascularity of the IMT (Fig. 2) and, under general anesthesia, we performed embolization of 3 intercostal arteries, the intercostobronchial artery and a branch of the lateral thoracic artery, using polyvinyl alcohol particles (150-250 μm Contour®, Boston Scientific/Target Vascular, Cork, Ireland). Two days after embolization, the mass was extirpated by right lateral thoracotomy after suturing the intercostal arteries and azygous vein before draining into the superior vena cava. After the embolization we were able to perform complete resection with decreased blood loss. The mass originated from the posterior mediastinum and vertebra, and after it had been excised the compressed right lung was totally expanded.

Histopathologic examination revealed myofibroblastic spindle cells with infiltration of lymphocytes and plasma cells, and led to a final diagnosis of IMT. Immunohistochemical staining was positive for vimentin, smooth muscle actin (SMA), and desmin, and staining for anaplastic lymphoma kinase (ALK) and S100 was negative.

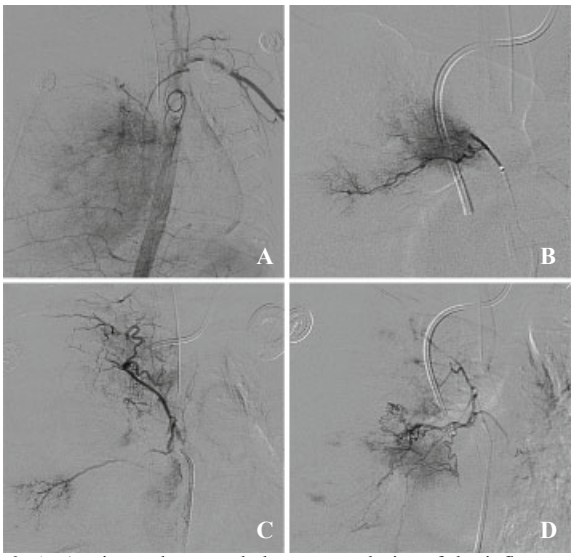

Fig. 2. A: Angiography reveals hypervascularity of the inflammatory myofibroblastic tumor; **B**: Intercostal artery supplies the tumor lesion; **C**: Intercostobronchial artery supplies the tumor lesion; **D**: Intercostal arteriography reveals hypervascularity of the tumor.

Postoperative recovery was uneventful, and laboratory results returned to normal within 3 days. Medical therapy with ibuprofen 25 mg/kg/day for 1 month was started postoperatively. No evidence of recurrence was found in follow-up CT and laboratory tests 6 months after the surgery.

Discussion

The most common site of IMT is the lung, but it can occur at many other locations such as mesentery, mediastinum, bladder, liver, spleen, head and neck and even heart.[3,5] Its definite etiology is not known, and it can be accompanied with nonspecific symptoms such as cough, dyspnea, hemoptysis, fever, and impaired growth depending on the location of the tumor. Laboratory abnormalities including hypochromic microcytic anemia, increased immunoglobulins, elevated ESR, thrombocytosis, and leukocytosis are resolved after tumor excision and this feature can be used to diagnose recurrence.[6]

In the present case we first suspected neuroblastoma, which is common in children, because of the larger size of the tumor and the necrotic lesions on CT. But the normal levels of ferritin and NSE, and the well-circumscribed tumor were inconsistent with neuroblastoma. Pleuropulmonary blastoma, carcinoid tumor, and immature teratoma were also considered as differential diagnosis. As chemotherapy and radiotherapy are not effective for these diseases, we decided that surgical excision should be the first line treatment.

Because the clinical and radiological features of IMT are variable and nonspecific, it is hard to differentiate it from other malignancies. Even with needle biopsy and frozen sections it is often difficult to rule out a neoplasm with surrounding inflammation, so an accurate diagnosis should be based on permanent histological examination.[1] A local recurrence rate of 8-37% has been reported and most such cases are associated with incomplete resection.[5,6] All these facts support the view that complete resection, which is necessary to confirm IMT, is also the most effective treatment.[5]

As the hypervascularity of IMT is not widely recognized and because of the possibility of renal injury by the contrast medium, enhanced CT was not performed before the first operation. However, as in other malignancies, hypervascularity should always be considered before surgery, and an effort should be made to decrease the risk of intraoperative bleeding by preoperative management such as embolization. When embolization is conducted, surgery should be performed within 2 days to prevent rapid revascularization, which tends to cause greater intraoperative bleeding.[7]

Although complete surgical resection is the treatment of choice, excision may be impossible due to invasion of vital thoracic and abdominal structures, or the patient's general condition. In such cases, chemotherapy, radiation therapy, glucocorticoids and nonsteroidal anti-inflammatory drugs (NSAIDs) can be alternative treatments.[5,6,8,9] NSAIDs inhibit angiogenesis by blocking vascular endothelial growth factor and cyclooxygenase-2, which are present in IMTs, and they have been reported to cause complete tumor regression after incomplete resection.[8,10] In addition, ALK expression, detected in half of IMTs, is associated with localized disease and better prognosis.[9] The fact that our patient was negative for ALK emphasizes the need for careful follow-up. Because there was a possibility of a poor prognosis, NSAID treatment was brought forward by 1 month postoperatively without any resulting complications.

In conclusion, we describe a huge IMT that shifted the mediastinum and compressed both atriums despite its benign clinical course. IMTs are not known to be hypervascular, but hypervascularity should always be considered, as for other tumors. To reduce intraoperative blood loss, which facilitating the surgery, preoperative embolization can be performed as one aspect of effective management.

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Contributors: Kim KN was in charge of the interpretation of the patient's data. Kim DW was responsible for the assessment of references and writing of the article.

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