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Case Report

Successful Sleeve Lobectomy of Inflammatory Myofibroblastic Tumor in a 4-year-old Child

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Primary pulmonary tumors in small children have remained a challenge for pediatric surgeons. Pneumonectomy and radical lobectomy are limitedly indicated due to surgical difficulties and sequelae. Here, we present our experience with a 4-year-old patient who suffered from an inflammatory myofibroblastic tumor. A left lower sleeve lobectomy was performed, and the patient recovered significantly after surgery. At the last follow-up, the child was growing well without any sequel, which supports our hypothesis that in small children, sleeve resection is the preferred treatment for tumors on the main stem bronchus and presents an alternative to an otherwise unavoidable pneumonectomy.

Keywords: inflammatory myofibroblastic tumor, sleeve lobectomy, pediatric surgery

Introduction

Primary pulmonary tumors in children are exceedingly rare. The majority of them seen in clinical practice are malignant (approximately 75%), making benign lesions even more rare.[1)] Inflammatory myofibroblastic tumor (IMT), which is composed of fascicles of bland myofibroblastic cells with an inflammatory infiltrate, accounts for less than 10% of the published cases of pediatric primary pulmonary tumors, and has been reclassified by WHO as a low-grade mesenchymal malignancy.[2)] Although rare, recurrence, invasion of adjacent organs and death are reported.[3)] In order to get a good prognosis, early diagnosis and complete resection are essential.

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Case Report

A 4-year-old boy was referred to our department because of left lung complete atelectasis. Prior to admission, he had a history of recurrent fever and cough for 3 months. After being diagnosed as having left-sided pneumonia, he had been treated with antibiotics which caused little improvement.

On admission, the patient presented with aggravated symptoms and dyspnea. His body temperature was over 39°C and his arterial oxygen saturation (SaO_2) was under 90% in room air. Physical examination was normal except for a shrunken left thorax with absent breathing sound. His white blood cell count was 12000/mm^3 with 73.8% neutrophils. A computed tomography (CT) scan showed complete atelectasis on the left lung and an endobronchial mass on the left main stem bronchus (**Fig. 1A**). The mass, which totally obstructed the left main stem bronchus, was located near the left lower lobar bronchus (**Fig. 1B**). On the third day after admission, bronchoscopic removal of the tumor was tried. Unfortunately, we did not succeed because the neoplasm completely obstructed the lumen of the left main stem bronchus with no space for maneuver of the bronchoscopy. The next day, after achievement of general anesthesia with a 4.5-mm single-lumen endobronchial tube, a left posterolateral

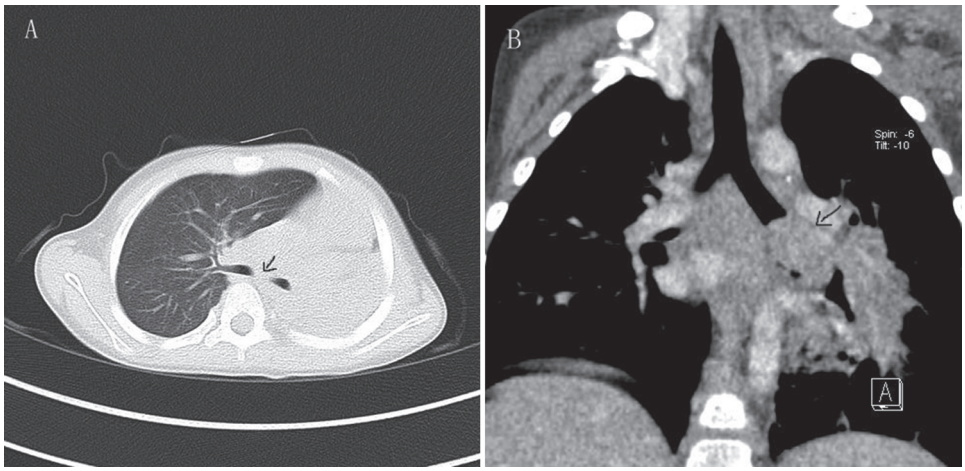

Fig. 1 (**A**) Chest computed tomography showing a totally collapsed left lung, and (**B**) coronal reconstruction chest computed tomography demonstrating a mass within the left main bronchus (arrow).

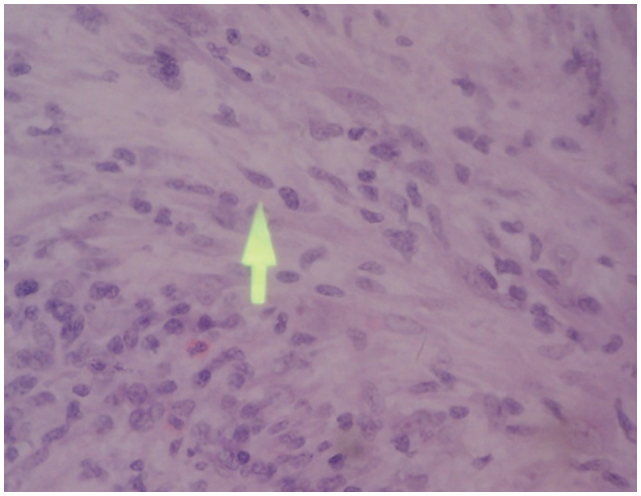

Fig. 2 Microscopic photograph of the specimen showing features of low-grade malignancy IMT (arrow).

thoracotomy was performed through the fifth intercostal space. Operative findings showed a round tumor with a diameter of 3 cm, which originated from the left lower lobe orifice and protruded into the left main stem bronchus. The lumen of the left upper bronchus was patent, leading us to perform a left lower sleeve lobectomy with surgical margins free of tumor involvement. The remaining left main bronchus was in continuity with the left upper bronchus, followed by the end-to-end anastomosis and was checked for air leaks. Histologically, the tumor was a low-grade malignant IMF (**Fig. 2**), and no metastasis was found in any of the sampled lymph nodes.

After the operation, right single-lung ventilation had been maintained for 2 days with FiO_2 at 0.4, PEEP at 4 cm H_2O, PIP at lower than 20 cm H_2O, SIMV at 15 breaths/minute. A chest CT scan performed after successful weaning showed the remaining left lobe was adequately inflated, although its volume was smaller when compared with the normal right lobe (**Fig 3A and 3B**). A follow-up by 3 month postoperatively indicated there was only slight narrowing of the bronchoplasty site and no atelectasis. At the last visit, 16 months after the resection, the patient was growing well and was free of recurrence.

Discussion

Although primary pulmonary tumors in children are exceedingly rare, approximately 75% are malignant. After having proved to carry a relatively good prognosis,[1] the condition still requires urgent diagnosis and treatment due to metastatic disease and irreversible lung parenchyma damage caused by recurrent pneumonia. Over the last several decades, bronchoscopic removal of primary pulmonary tumor has been a more attractive way, along with simpler and less invasive lesions.[2] For bronchogenic malignancy, endoscopic excision does not guarantee radical resection and the tumor may recur locally or disseminate to the distant site, thus surgical resection is thought to be a more reliable choice for malignant lesions.

However, pediatric surgeons are often reluctant to perform radical resection for endobronchial tumors in small children, particularly those on the main stem bronchus, which have an indication for pneumonectomy. The reasons are obvious: there are great difficulties during the operation and devastating sequelae after the operation.

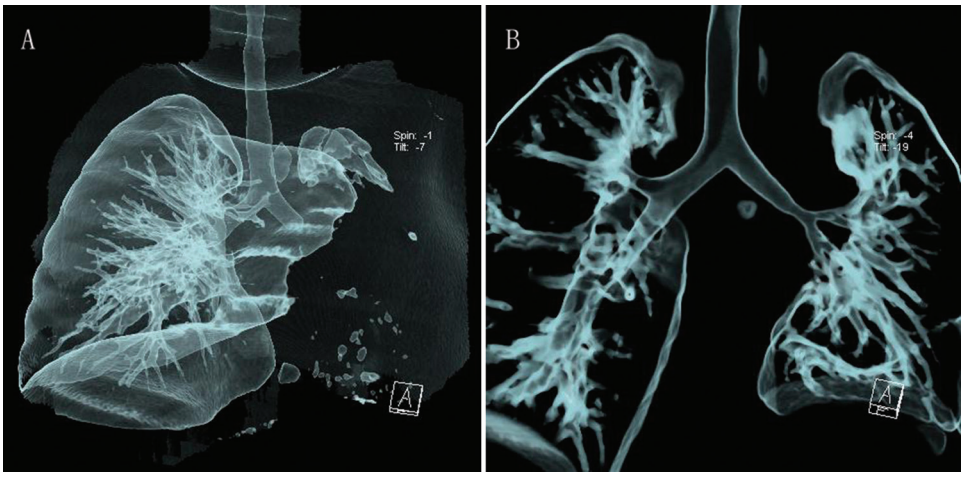

Fig. 3 (**A**) Preoperative three-dimensional airway reformation showing a totally obstructed left lung, and (**B**) postoperative three-dimensional airway reformation demonstrating an aerated left lung with a smaller size, compared with a normal right lung.

Compared with surgical difficulties, pediatric surgeons are much more concerned about the sequelae because of their influence on patients' quality of life. Major complications in children involve a reduction in pulmonary function, failing to thrive, chest deformities, spinal deformities (mostly scoliosis) and life-threatening right postpneumonectomy syndrome, all of which severely lower patients' quality of life.[4)]

Luckily, it has been proved by several institutional studies that if the nodal status is limited to N2, radical sleeve lobectomy can almost utterly avoid the sequelae, with survival results at least equal to those of pneumonectomy.[5)] However, until now, sleeve lobectomy is mainly operated in adults and adolescents. Pediatric surgeons still have limited experience in performing sleeve lobectomy in small children. In the literature, less than 15 cases in small children, who received radical sleeve lobectomy, have been reported, and the patient in our case was the youngest child. Herein, we report our successful experience, combined with another case reported by Yangki, et al.[2)] We draw the conclusion that with experienced surgeons, it is completely feasible to apply sleeve lobectomy in small children. Regardless of difficulties during the operation, sleeve lobectomy can certainly guarantee both a low mortality and low complication rates. Meanwhile, an enormous endobronchial tumor on the main stem bronchus is not an indication for pneumonectomy if there is no metastasis in the atelectatic lobe.

Here, we also want to emphasize the importance of early diagnosis. As the tumor was diagnosed as a low-grade IMT, with prompt recognition, the patient in our case could probably avoid the aggressive excision by endoscopic resection or laser ablation. Unfortunately, the patient was misdiagnosed as having pneumonia because the clinical and radiographic presentations were nonspecific and mimicked presentations of pneumonia. Clinically, cough is the most common symptom in small children, followed by fever and no pulmonary symptoms. Yu, et al. found hemoptysis is the least common complaint in the pediatric arm, as compared to the adult arm,[6)] making the diagnosis in small children even more difficult. With radiographic presentations also mimicking presentations of common diagnoses, including collapse and consolidation, primary pulmonary tumors in small children are easily misdiagnosed as pneumonia and bronchial asthma. Often, the possibility of a primary pulmonary tumor is considered only when radiographic abnormalities or symptoms persist or fail to respond to therapy, at this time, most patients are at an advanced stage, like in this case. Therefore, we appeal to pediatric physicians for accepting the suggestion given by Curtis, et al.: any child who presents with persistent unexplained consolidation that fails to resolve within 2 weeks of therapy should be referred for further examination, particularly bronchoscopy.[7)]

Conclusion

Pediatric primary pulmonary tumor is a condition requiring urgent diagnosis, and with experienced surgeons, sleeve lobectomy is the preferred treatment in small children for lesions on the main stem bronchus

and presents an alternative to the otherwise unavoidable pneumonectomy.

Disclosure Statement

The authors have declared no conflict of interest.

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