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Endobronchial Chondromatous Hamartoma in an Infant

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INTRODUCTION

Hyperinflation or atelectasis of a lung or one of its lobes is generally due to partial or complete bronchial obstruction. Intramural large airway obstructive lesions or cysts are uncommon in the pediatric age group. They are usually discovered in the supraglottic or glottic areas.[1] Primary pulmonary, tracheal, or bronchial tumors are very unusual in children.[2,3] In this report, we describe an infant with an endobronchial chondromatous hamartoma obstructing the right main bronchus. This infant presented with cough, wheezing, and unilateral hyperinflation of the right lung. The patient's symptoms improved after endoscopic resection of the endobronchial lesion.

CASE REPORT

An 8-month-old infant was admitted to the hospital with a history of cough, wheezing, and chest congestion for 4 days. The patient was born prematurely at 28 weeks of gestation and was on mechanical ventilation for 1 month. He was doing well after discharge from the neonatal intensive care unit, except for occasional wheezing. On examination during this admission, he had decreased air entry and wheezing over the right side of the chest only. A chest radiograph showed unilateral hyperinflation of the right lung (Fig. 1). A previous chest radiograph 4 months earlier showed symmetrically expanded lungs. Flexible and rigid bronchoscopies were done, and both showed the presence of a polypoid mass obstructing most of the lumen of the right main bronchus except for a slit-like opening (Fig. 2). The mass was completely removed during the rigid bronchoscopic examination, restoring patency of the right main-stem bronchus. The biopsy consisted of two small fragments of tissue 1 and 2 mm in diameter. Microscopically, the fragments were covered by respiratory epithelium, and the underlying fibrous stroma contained submucosal glands mixed with several tiny disorganized nodules of benign cartilage and large tortuous arteries abnormal for this location (Figs. 3 and 4). No fat, myxoid tissue, smooth muscle, bone, bone marrow, or calcification were found. The lesion was diagnosed as a chondromatous-vascular hamartoma. The postoperative course was uneventful, and the cough and wheezing disappeared. Several chest radiographs were obtained in the 2 years of follow-up, and these showed symmetrical inflation of both lungs with no evidence of unilateral disease.

DISCUSSION

Endobronchial obstructing lesions in children are uncommon. The differential diagnosis includes aspirated foreign bodies, ectopic thyroid tissue, mucosal webs, cysts, and tumors. Intramural cysts of the trachea causing recurrent cyanotic spells were described in three case reports.[4,5,6] The biopsy of the lesion in two cases showed normal respiratory epithelium, scanty capillary vessels, primitive cartilage, and no mucous glands.[4,6] In the third case, the airway obstruction manifested at birth, but no histologic description was provided.[5]

Bronchial and pulmonary tumors in children are rare, and they include papillomas, granulomas, hemangiomas, adenomas, and hamartomas.[2,3,7] In an extensive review of the English-language literature, Hartman and Shochat classified most of these tumors as malignant, although only 8% of them were definitely so.[2] The most common benign primary pulmonary tumors of the lung in children are inflammatory pseudotumors, followed by hamartomas.[2,7] The term ''hamartoma'' was first used by Albrecht in 1904 to describe a malformation comprised of abnormal growth of tissues that are native to a given organ.[8] Benign mesenchymoma or ''adult'' hamartoma is considered the commonest benign tumor in the adult lung.[8,9] It usually occurs in the fifth or sixth decades of life.[8,10–12] These are mainly solitary in location and considered to be very slow-growing lesions.[9] However, they can occasionally be multiple in the lung or associated with other benign hamartomas in other organs.[13,14] Classically, almost 80% of hamartomas are located in the peripheral lung parenchyma, and the rest are central in location.[15,16] However, three recent series of adult pulmonary hamartomas

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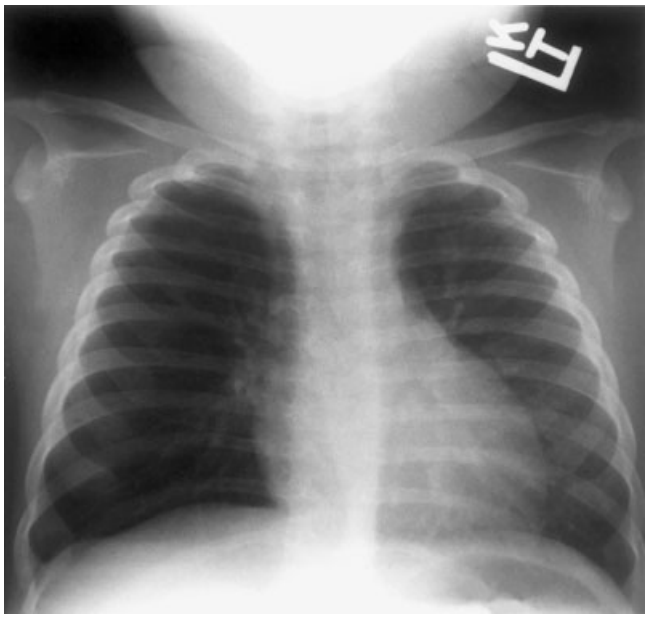

Fig. 1. Postero-anterior chest radiograph, showing hyperinflation of right lung.

found the incidence of bronchial hamartomas to be between 1–12%.[10–12] In his large series, Tomashefski concluded that both the parenchymal and the bronchial hamartomas of the adult lung are similar in nature.[10] These lesions are extremely rare in children. By the early 1980s, only 15 patients were described in the pediatric age group, and only 2 of these patients had bronchial tumors.[2] In the early 1990s, 2 adolescents were reported as having

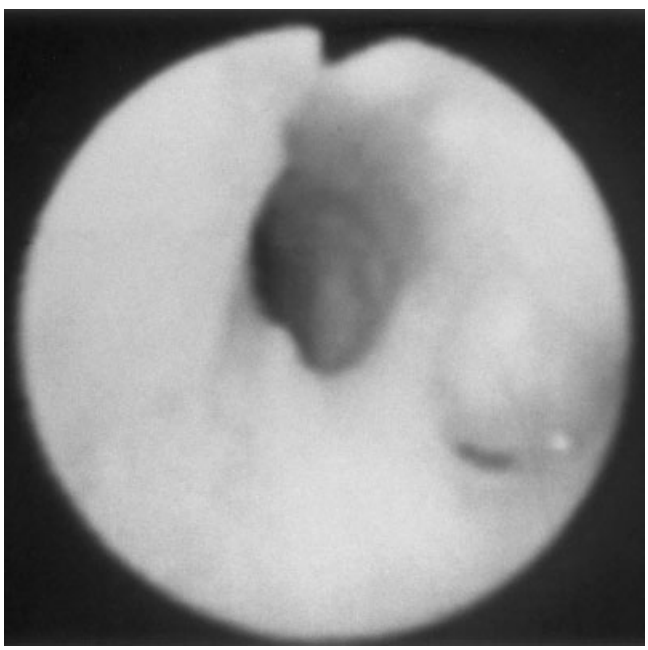

Fig. 2. Bronchoscopic view of lesion obstructing right main bronchus at the carina.

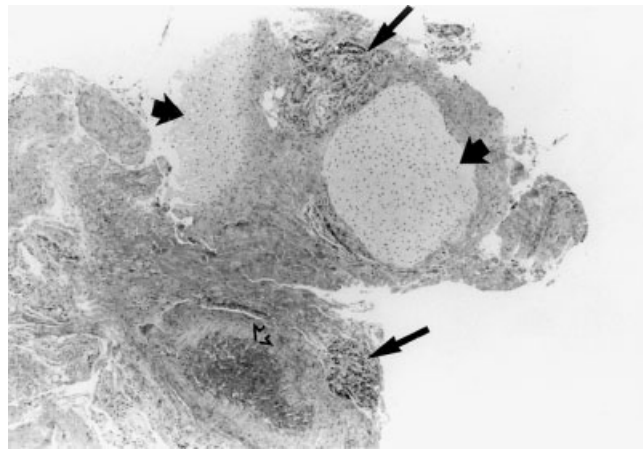

Fig. 3. Cross section of lesion, showing two small nodules of cartilage (thick solid arrows) adjacent to submucosal glands (thin solid arrows) and an abnormal large artery (open arrow) (magnification, ×40).

pulmonary hamartomas among a total of 154 patients in two series, but none of them had a bronchial lesion.[11,12] One additional recent paper in the English-language literature reported the presence of multiple endobronchial chondromatous hamartoma in a 5-year-old child.[17] These numerous lesions caused significant atelectasis and bronchiectatic changes, prompting removal of the whole affected lung.[17]

As stated earlier, hamartomas usually have an abundance of cartilage and glandular tissues.[8,9,18] The lesion in the present child could represent a developmental abnormality of the different tissues native to the airways rather than a "neoplasm." In our differential diagnosis, we considered the possibility of a previous trauma to the airways as an underlying mechanism. Trauma secondary to endotracheal intubation, repeated suctioning, or

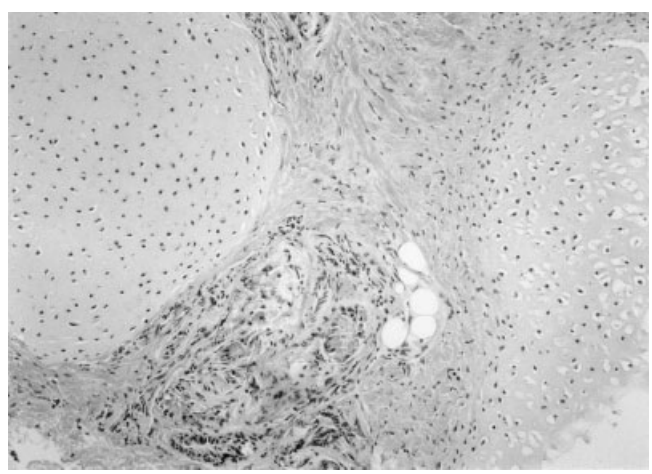

Fig. 4. Larger magnification of cartilage nodules, showing morphology of chondroid component.

mechanical ventilation usually produces fibrous and capillary vascular proliferation, leading to stenosis of the airway. This appeared unlikely in our patient, due to the presence of disorganized cartilage, lack of scarring, and the presence of thick-walled vessels. There were no other lesions in the trachea or the bronchi visualized at endoscopy to raise a suspicion of trauma in this child. Posttraumatic response to injury with chondroid metaplasia was considered. However, the presence of several cartilagenous nodules, the lack of stromal scarring around them, and the abundance of submucosal glands favored the diagnosis of hamartomatous malformation.

We also considered a technical error in that the biopsy might have represented a portion of the bronchial wall. This was also ruled out because of the presence of disorganized cartilage formation, which one would not expect to see in the wall of a normal airway. Also, one would expect postoperative complications if the biopsy injured the bronchial wall. Bronchogenic cysts can contain cartilaginous rings, but these lesions are most frequently seen adjacent to the trachea, carina, or left main bronchus. They are usually attached to the wall of the trachea rather than forming an intraluminal mass, as was noted in our case. Histologically, the lesion in this infant did not appear cystic but was rather solid, with several disorganized nodules of cartilage. Other diagnoses such as simple cysts and mesenchymal cystic hamartomas were also ruled out, since these lesions do not contain cartilage.

Because of their slow growth, hamartomas in adults can be asymptomatic for a prolonged period of time before being discovered, and are typically seen incidentally as coin lesions in chest radiographs.[8] In addition, malignant transformation of pulmonary hamartomas is considered rare. It is suggested that surgical removal of the tumor in older patients should be done only if the tumor is symptomatic or if malignant transformation is suspected.[11] In contrast to adults, both peripheral and centrally located hamartomas can lead to significant symptoms in children, and prompt surgical removal is required. Our patient had decreased air entry and unilateral hyperinflation of the right lung. The lesion must have been a rapidly growing lesion and was fortunately discovered before further atelectatic and bronchiectatic changes had taken place. Almost all of the endobronchial hamartomas were removed via lobectomy or pneumonectomy[2,10–12] except in one adult patient reported by Hansen et al.,[11] in whom the tumor was removed through endoscopy. Fortunately, in our patient the tumor was successfully excised endoscopically, with an immediate improvement in his symptoms.

In conclusion, we described a young infant with an endobronchial chondromatous hamartoma obstructing the lumen of the right main bronchus. While primary pulmonary tumors are rare in children, this is the second case of endobronchial tumor reported by us lately.[3] To our knowledge, this is the youngest reported case of endobronchial hamartoma. We emphasize the need to consider uncommon entities in the differential diagnosis of intraluminal bronchial obstructive lesions in children. Bronchoscopy can be instrumental in visualizing airways and in allowing safe resection of small lesions.

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