Role and importance of Doppler ultrasound in the diagnosis of pulmonary sequestration: Report of two adolescent cases.

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Abstract
Pulmonary sequestration is a rare pulmonary parenchymal anomaly. The localization of the aberrant artery and its relation with sequestration are of importance in the process of diagnosis in the preoperative period. The feeding artery can be identified by color Doppler ultrasonography. We report two cases of pulmonary sequestration in two adolescents, aged 11 and 13 years old, and this report documents the use of in which color Doppler ultrasound demonstrated the abnormal feeding artery. Color Doppler ultrasonography, a noninvasive and radiation-free imaging modality should be considered in the diagnostic algorithm of pulmonary sequestration.

Keywords: pulmonary sequestration, color Doppler ultrasonography, adolescent

Introduction
Pulmonary sequestration is a rare pulmonary parenchymal anomaly that accounts for 0.15–1.7% of all congenital lung abnormalities and is classified into two subtypes, intralobar and extralobar sequestration. Non-functional lung tissues in pulmonary sequestration do not communicate with the normal tracheobronchial tree and have their own systemic arterial supply [1]. Various radiological modalities are being used in the diagnosis of pulmonary sequestrations. These are: posterior anterior (PA) chest roentgenograms, ultrasonography, computerized tomography (CT), magnetic resonance (MR) imaging and digital subtraction angiography (DSA).

In this brief report we present two cases and we highlight the importance of assessing pulmonary sequestration in the differential diagnosis of patients with a history of recurrent infections since their birth. Additionally, we emphasize the use of sonography and color Doppler ultrasound as potential contributors in the diagnosis of pulmonary sequestrations.

Cases report
Case 1
An 11-year-old girl was brought to the pediatric outpatient clinic with complaints of coughing, orthopnea and transparent fluid flowing back into her mouth, particularly when lying on her back. A detailed history revealed recurrent pneumonia since her birth. Although she had received empirical antibiotics each time, her symptoms did not resolve completely. At admission, her vital signs were as follows: blood pressure 105/55 mmHg; respiratory rate 20 breaths/min; pulse rate 73 times/min; body temperature 36.6°C. Complete blood counts were as follows: white cells count $9.7\times10^3/\mu l$; hemoglobin 10.1 g/dl; platelet count $210\times10^3/\mu l$.

Her PA chest roentgenogram revealed a 10-cm-diameter, well-circumscribed lesion showing air-fluid level and located in the lower zone of the right lung (fig
Due to the absence of fever and leukocytosis and given the fact that the family kept a domestic dog, the pre-diagnosis of abscess was ruled out and rather a hydatid cyst fistulating into the bronchia was considered. A non-contrast thoracic CT examination revealed a mass occupying lesion located within the basal segment of the right lung lower lobe, adjacent to the diaphragm. This lesion was observed to fistulate into the bronchia and showed an air-fluid level (fig 1b). Given these radiological findings, we reported that the lesion might be either a hydatid cyst fistulating into the bronchia, an abscess or a pulmonary sequestration. Oral hydatid cyst therapy was prescribed and immune histochemical assays were requested to confirm the diagnosis. Following 14 days of medical treatment her clinical condition significantly recovered, whereas her PA chest roentgenogram did not improve. The patient was referred to the pediatric surgery outpatient clinic. The grey-scale examination was followed by the color Doppler sonography examination (fig 1c, 1d). A color Doppler ultrasound examination of the lesion, which was close to the diaphragm, demonstrated thick septa with remarkably good blood supply. On tracing the vascular structure supplying the lesion, we discovered that it originated from the subdiaphragmatic area. It has been previously reported that septa within the hydatid cyst are nonvascular and therefore the lesion was concluded to be more in line with pulmonary sequestration. The pre-diagnosis of pulmonary sequestration was validated through postoperative histopathologic examination (fig 2).

Case 2

A 13 year-old male patient applied to our outpatient clinic with complaints of recurrent coughing. His medical history revealed that he experienced cough episodes of long duration, which were resolved after medical therapy but showed frequent relapses. At admission, vital signs were as follows: blood pressure 110/60 mmHg; respiratory rate 19 breaths/min; pulse rate 77 times/min; body temperature 36.4 C. Complete blood counts were as
follows: white cell count $8.4 \times 10^3/\mu l$; hemoglobin 10.8 g/dl; platelet count $197 \times 10^3/\mu l$.

Since the PA chest roentgenogram showed an area with increased homogenous density at the left retrocardiac area, a prediagnosis of pneumonia was made and lung CT was performed (fig 3a). Computed tomography revealed a mass occupying lesion with heterogeneous thick septa at the mediobasal segment of the left lung lower lobe that showed significant vascularization after contrast enhancement (fig 3b). The lesion was prediagnosed as pulmonary sequestration and taking our former experience into account we performed a color Doppler ultrasound examination. Color Doppler ultrasound revealed a heterogenous lesion composed of cystic areas with thick walls and septa, noted to have good blood supply. On tracing the vascular structure supplying the lesion, we found it originated from the thoracic aorta (fig 3c, 3d). The prediagnosis of pulmonary sequestration was validated through postoperative histopathology examination.

Discussions

There are two forms of pulmonary sequestrations, namely intralobar and extralobar sequestrations. Both forms are believed to derive from the same single embryonic origin [2]. Although the embryologic mechanisms are still to be fully elucidated, pulmonary sequestrations are believed to originate from malformations occurring during the branching of the tracheobronchial tree or from the accessory branching of the primitive foregut. Intralobar sequestrations account for 73% of all pulmonary sequestration cases and are usually located in the posterior basal segment of the lower lung lobe [3]. Intralobar sequestrations are usually not associated with other congenital abnormalities. Their venous drainage is through the general portal venous system [2]. Owing to its own pleural sac, the infection risk is low. Extralobar sequestrations may show co-existence with other congenital abnormalities, diaphragmatic hernia being the most frequently observed one at a frequency of 30% [4].

While clinical findings manifest as recurrent pneumonia exacerbations in early childhood, half of the cases can only be diagnosed after 20 years of age, as observed in our cases [5]. Our literature survey revealed an even more striking case diagnosed with pulmonary sequestration at the age of 83 [6]. Given the recurrent and severe nature of pneumonia exacerbations, accurate early diagnosis and effective treatment is imperative.

PA chest roentgenograms and bronchography findings are usually helpful in diagnosing pulmonary sequestration. Depending on their localization, pulmonary sequestrations may usually manifest as cystic structures dem-onstrating fluid-air level as was present in our first case. They rarely appear as mass occupying lesions in a PA chest roentgenogram, as was present in our second case.

A PA chest roentgenogram is usually regarded as the first line diagnostic method for pulmonary sequestration. Prediagnosis is generally pneumonia and management involves the initiation of antimicrobial treatment, which may indeed partially resolve the symptoms in the majority of patients. Since their childhood, both of our cases had recurrent infections, from which they recovered partially following antimicrobial therapy, but showed relapse shortly after. PA chest roentgenograms of both patients were taken at the symptomatic stage and appeared likely to be consistent with pneumatic consolidation. But this approach may lead to the actual underlying disease being overlooked.

Although fundamental radiology and clinical experience describes a limited role for sonography and color Doppler ultrasound imaging in the diagnosis of pulmonary disorders, these modalities may provide useful and important information for the lesions particularly located in close proximity to the diaphragm and chest wall, as in our cases. Moreover, and most importantly, these imaging modalities are noninvasive and free of ionizing radiation.

At the preoperative stage, another important issue for patients with suspected pulmonary sequestration to be taken into account is the localization of the aberrant artery and its association with the sequestrum. The arterial blood supply of a nonfunctioning lung segment is usually from the descending aorta itself, from its branches, or from an aberrant artery originating from the abdominal aorta. Data regarding the number of aberrant arteries and their origin is gradually increasing in the literature [7]. From a surgical perspective, detection of the origin of the aberrant artery that supplies sequestrated tissues is of critical importance, since hemorrhage of these arteries may consequently lead to death [8].

Recent technological advances have led to a wider availability of and increased quality in radiologic imaging. Although DSA and CT angiography provide very useful information in demonstrating an aberrant artery, they are invasive and lead to radiation exposure as well. On the other hand, since most of the patients with pulmonary sequestration are children, performing MR angiography is quite challenging and is limited due to its sensitivity to patient movement. In light of the above-mentioned factors, we suggest that although color Doppler ultrasound is not the gold standard, it should be considered as a valuable imaging modality in the diagnosis of pulmonary sequestration.

In conclusion, we highlight the importance of assessing pulmonary sequestration in the differential diagnosis of patients with complaints of recurrent and refractory
pneumonia exacerbations. Additionally, we recommend color Doppler ultrasound, as an easy to use, cheap, non-invasive and radiation free imaging modality, to be considered after PA chest roentgenograms in the diagnostic algorithm for pulmonary sequestration.

References