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Review Article

Accessory hemi diaphragm

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ABSTRACT

Background/purpose: Accessory hemi diaphragm (AHD) is an uncommon condition. Its clinical features, diagnosis and management have not been clearly defined.
Methods: We reviewed the world literature and added a case of our own.
Results: There are 40 proven cases in the literature. The lesion occurs almost exclusively in the right hemithorax. Twenty seven cases presented with cardiovascular and/or respiratory symptoms while 3 had symptoms pertaining to other systems. In the remaining 10 patients the diagnosis was either made accidentally or information pertaining to their presentation is missing. Abnormalities of pulmonary and systemic blood vessels often accompany the abnormality. The diagnosis is usually suspected because of an abnormal chest radiograph and confirmed by a variety of tests.
Conclusions: If the AHD prevents proper aeration of the lung it should be excised. If vascular abnormalities coexist they should be treated along with the AHD. Asymptomatic patients may be observed after confirming the diagnosis.

In 1945 Haeberlin [1] described a rare anomaly in which the hemithorax was divided by a transverse septum. Since then this anomaly has been incorrectly named an accessory diaphragm even though it occupies only one side of the chest. Now that a complete accessory diaphragm has been described by Krzyzniak & Gray in 1986 [2] it is appropriate to correctly name it an accessory hemi diaphragm (AHD). Patients with an AHD may or may not be symptomatic, the diagnosis is difficult and management is unclear. By reviewing previously reported cases and one of our own we have been able to better clarify diagnostic criteria, and define management options.

1. Case report

In 1988 a 2630 gram boy was born at an estimated gestational age of 34 weeks. The APGAR scores were 9 and 9 at 1 and 5 minutes. On day 2 his respiratory rate rose to more than 60 breaths per minute. He had a normal appearing chest radiograph at this time. Within the next 12 hours, the respiratory rate increased to 100 breaths per minute. Abnormalities were observed on the repeat chest radiograph. On the frontal projection the right lung field was hazy, the cardiac border was indistinct and a soft tissue shadow was seen at the apex (Fig. 1). The lateral film demonstrated a well-defined anterior density extending from the apex to the xiphoid process (Fig. 2). On echocardiography mild aortic stenosis and mitral regurgitation were diagnosed. A ventilation-perfusion lung scan (VQ scan) revealed no aeration or blood flow in the right upper lung field. No right upper lobe bronchus was visible on bronchoscopy but the bronchus intermedius appeared normal. Contrast could not be instilled in either the upper or the lower lobe bronchi on bronchography. At thoracotomy a thick fibrous membrane was attached to the 2nd and 3rd vertebral bodies and ribs posterolaterally and to the pericardium and the septum transversum anteriorly. There was an empty space in the hemithorax above the AHD and the bronchus and vessels reached the lung through an opening in the medial aspect of the AHD. Once the septum was excised and the lung expanded it became apparent that the upper lobe was absent and the lower lobe was small and atelectatic as the lower lobe bronchus had been compressed by the inferior edge of the opening in the AHD. The AHD was lined by mesothelium on both surfaces and it contained no muscle or nerve fibers. The patient had an uneventful postoperative course, the tachypnea was relieved and he was discharged home. He remained asymptomatic and he did not require cardiac surgery in the 8 years he was followed by us. The family then moved away from the area.

2. Materials and methods

Table 1 lists 40 confirmed cases of AHD in the world literature including ours (Case #33). The literature search was carried out by using the terms accessory diaphragm and diaphragmatic abnormalities in Medline. Seven of these cases were confirmed at surgery or autopsy but clinical details were not provided [33]. We were unable to

Abbreviations: AD, accessory diaphragm; AHD, accessory hemi diaphragm; MDCT, multidetector computed tomography scan; MRA, magnetic resonance angiogram; VQ scan, ventilation-perfusion lung scan.

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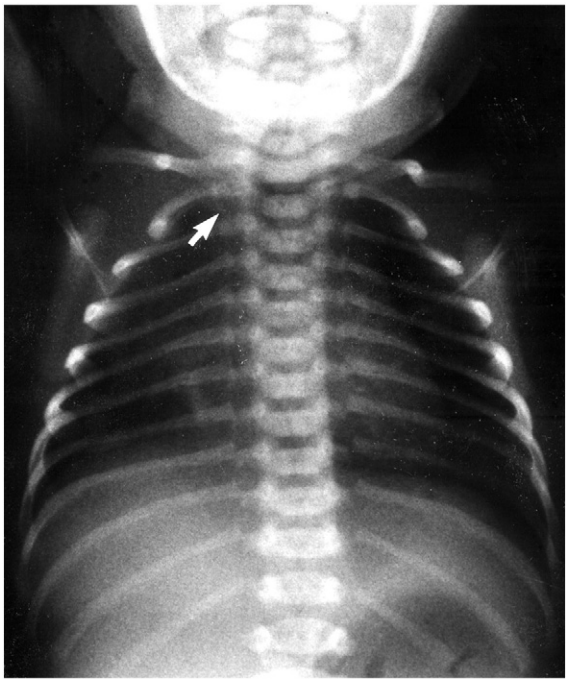

Fig. 1. Frontal chest radiograph demonstrating an indistinct right heart border (silhouette sign) and a right apical density (white arrow).

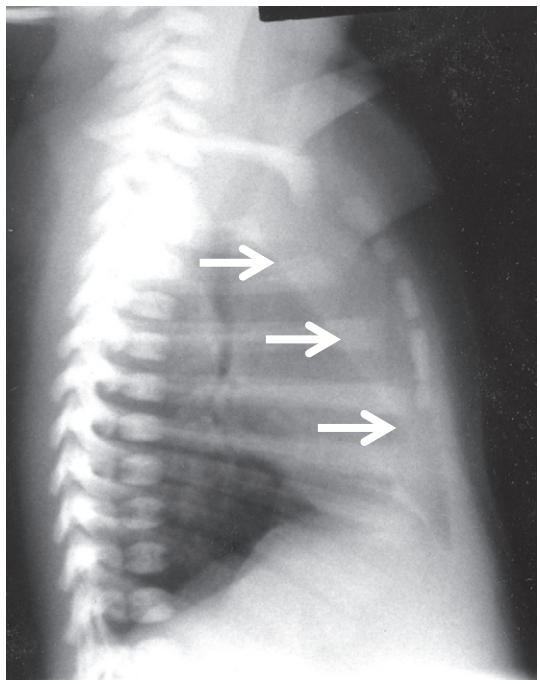

Fig. 2. Lateral chest radiograph demonstrating the anterior dense strip running parallel to the sternum from the apex of the chest to the xiphoid process (white arrows).

confirm the diagnosis in 13 possible cases which have been excluded from this review (Table 2). Wille et al reported the same cases in the English [18] and German literature [19] hence the repeated cases have been excluded. In this retrospective review follow-up is unevenly reported. Our attempts at updating follow-up by phone calls, regular mail and e-mail were only successful in 2 patients (#s 31 and 32).

3. Pathology

In 37 of the 40 confirmed cases the AHD was on the right side, 1 was on the left [12], and the side was not mentioned in 2 cases [33]. The septum attaches anteriorly and medially to the central tendon of the thoracic diaphragm and the pericardium. It extends posteriorly to attach usually to the 5th, 6th or 7th ribs but it may attach higher as it did in our patient. Both surfaces of the AHD have a serosal lining. The composition of the AHD varies considerably. In our patient and others in the literature it was a fibrous septum while on other occasions striated muscle had been identified in the center while the periphery was thin and transparent. In one patient the ipsilateral thoracic diaphragm was elevated and devoid of muscle while the AHD was muscular [5]. Rarely the AHD may be innervated by branches of the phrenic nerve [10].

The amount of lung tissue trapped under the AHD depends upon the site of its posterolateral attachment. In high lesions the whole lung or 2 lobes may be trapped beneath it while in low ones only the lower lobe is under the AHD (Tables 3–5). The arterial, venous and bronchial structures to the trapped lobe/s make their way through a 2–3 cm. opening in the posteromedial aspect of the septum. This orientation has been likened to a “back-pocket” [4].

Varying degrees of pulmonary hypoplasia or agenesis occur. In addition, various anomalies of the trapped lobe have also been described. The empty space created in the hemithorax is filled by extrapleural adipose and areolar tissue which has a distinct appearance on chest radiographs.

Anomalous vascular connections commonly accompany AHD (Tables 1, 3). They are invariably present in low AHDs hence it has been postulated that the abnormal vessels may be anchoring the lobe that is trapped under the AHD [14]. The aberrant vessels may traverse the thoracic diaphragm (Table 1, SNs 14, 23, 24, 26, 28, 31).

4. Embryologic considerations

The development of AHD is unclear. Haeberlin [1] stated that Schwalbe believed that the AHD develops when a laterally displaced azygos vein pulls pleura along with it as it traverses the thoracic cavity. This hypothesis does not explain its development in patients without vascular anomalies nor does it explain the presence of striated muscle in the septum. Some authors blame the origin of an AHD on slow descent of the septum transversum or early development of the lung bud [4,5] while others attribute it to splitting of the septum transversum [25]. These hypotheses do not explain how the AHD would extend posterolaterally to the ribs since the septum transversum only forms the base of the pericardium.

We believe that the anomaly is probably the result of the lung bud growing into the posthepatic mesenchymal plate which normally forms the greatest part of the diaphragm [36]. However, it is unclear whether pulmonary hypoplasia is the primary problem or the result of diminished space in the hemithorax.

5. Clinical features

Symptoms and signs are because of varying combinations of pulmonary hypoplasia or agenesis, compression of lung tissue, and cardiovascular and other anomalies. Thus patients can be placed into three groups:

Group 1 Patients with cardiovascular and/or respiratory symptoms and/or signs = 27 (Table 3).
Group II Patients with symptoms and/or signs other than cardiovascular or respiratory = 3 (Table 4).
Group III Asymptomatic confirmed cases diagnosed incidentally = 3 (Table 5).

5.1. Unclassifiable

In Table 6 we have listed 7 patients with confirmed AHD who cannot be classified since the authors have not shared details of clinical features, diagnostic tests or type of operation or outcome after treatment (Table 1, SNs 34–40).