CASE REPORT

Intradiaphragmatic hybrid lesion: surgical decision-making and value of minimal invasive surgery

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SUMMARY

Hybrid lesions (HLs) have elements of congenital pulmonary airway malformation and extrapulmonary sequestration (EPS) and belong to the congenital lung lesions. EPS usually arises in the thorax or the abdomen but rarely in the diaphragm. The preoperative diagnostic work-up based on chest radiograph, ultrasound (US) and CT often shows imprecise results. Therefore, the exact localisation of the lesion can only be ascertained intraoperatively. Here we present a patient, with an intradiaphragmatic HL, and demonstrate the difficulties of surgical decision making regarding the localisation of the lesion and discuss the value of minimal invasive surgery.

BACKGROUND

Congenital pulmonary airway malformation (CPAM) formerly named congenital cystic adenomatoid malformation, pulmonary sequestration (PS) and congenital lobar emphysema are the three most common congenital lung lesions in children.¹ There are two types of PS, the intralobar sequestration and the extralobar sequestration (EPS).³ Most congenital lung lesions are diagnosed during antenatal screening ultrasound (US).⁴ Postnatally, US, Doppler US, plain radiographs, CT or MRI is

used to confirm the diagnosis of congenital lung lesion and for preoperative planning. Since there are many pathological and clinical similarities in the cited malformations, it is probably more correct to consider them as a spectrum of disease.⁵ The so-called hybrid lesion (HL) which has elements of CPAM and EPS emphasises this. Cystic lung lesions can present from the neonatal period to adulthood. The spectrum of complication reaches from asymptomatic to lethal hydrops fetalis or postnatal respiratory distress. There is a certain risk in CPAM for malignant transformation.^{6,7} The recommendation is to resect affected lobes. However, CPAM can spontaneously regress and therefore the treatment of asymptomatic patients is controversial.

The preoperative diagnostic work-up often shows imprecise results regarding the exact localisation of the lesion.^{8,9} Often, only the operation can clarify the entity. By presenting a patient with an intradiaphragmatic HL, we demonstrate the difficulties of intraoperative surgical decision-making regarding the localisation of the lesion and discuss the value of minimal invasive surgery (MIS).

CASE PRESENTATION

We report the case of a 9-month-old girl, term born, with an intradiaphragmatic HL. On antenatal US, a bronchopulmonary malformation or a congenital diaphragmatic hernia (CDH) was suspected. Immediately after birth, the patient showed signs of respiratory distress. Following intubation, she had a plain chest radiography to rule out a CDH. The patient stabilised after a few minutes and was extubated within an hour. She stayed asymptomatic afterwards.

INVESTIGATIONS

Postnatally, a plain chest radiograph (figure 1) and US (figure 2) showed a mass with cysts less than 1 cm in diameter in the left lower lobe which was classified as CPAM type II. CT (figure 3) confirmed this finding with the additional identification of an aberrant arterial supply of the lesion coming from the coeliac axis, which is the key feature of PS. These findings suggested an intrathoracic HL.

DIFFERENTIAL DIAGNOSIS

The initial diagnosis in antenatal US was an eventration of the left diaphragm or a CDH. In the postnatal US however, a mass with the typical echogenic structure for a CPAM II was seen. The aberrant arterial supply coming from the abdominal

Figure 1 Postnatal plain trunk X-ray with mild diffuse opacity in the left lower zone of the thorax.

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Figure 2 Postnatal sonography with typical echogenic structure for a congenital pulmonary airway malformation type II.

aorta was typical for an EPS. Having diagnosed an intrathoracic mass with an echogenic structure in US of a CPAM type II and an aberrant arterial supply, we expected an intrathoracic HL. However, some EPS may be found below the diaphragm.

TREATMENT

Despite of the child being asymptomatic, we recommended elective surgery due to the risk of bronchopulmonary infections and malignant transformations later in life. At the age of 5 months, the girl underwent thoracoscopy showing an eventration of the diaphragm, with a slightly irregular surface, but no mass effect was notable underneath. The lung appeared normal; the expected HL could not be identified. Assuming the HL was intra-abdominal, we performed a thorascopic plication of the eventration of the diaphragm. In order to shorten the duration of anaesthesia and to avoid the morbidity of a two-cavity approach, laparoscopy was postponed. An MRI at the age of 9 months showed no changes of the mass itself, and again the radiologist suspected an intrathoracic location. During laparoscopy, we could identify the HL intradiaphragmatically (figure 4) and removed it.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful. Histology confirmed an HL with a CPAM type II lesion in the sequestration.

Figure 3 CT coronary slides of the contrast study. The lesion and its aberrant vessel coming from the coeliac axis are shown in the left diaphragmatic area.

Figure 4 Intradiaphragmatic hybrid lesion during laparoscopy.

DISCUSSION

Although CT remains the criterion standard imaging examination in the diagnostic work-up of EPS and for planning surgery,¹⁰ the exact localisation of such lesions may be discovered only during surgical exploration. Thoracotomy is still the traditional approach but thoracoscopy is possible. The thoracoscopic approach is safe and accurate by means of obtaining certainty of the anatomical situation and treating the malformation. Complications and adverse effects like scoliosis, adhesions, postoperative pain and respiratory impairment are reduced to a minimum with the MIS approach.¹¹ A possible disadvantage from the pathological perspective is that the specimen may be more fragmented, the architecture may be distorted, making the specific diagnosis after pathological criteria more difficult.

Since the patient was asymptomatic, the operation probably could have been postponed after the first year of life but this would have exposed the patient to a higher risk of superinfections and the development of a symptomatic lesion, complicating further treatment and causing morbidity.

In our literature research, we could identify 10 reported cases of either intradiaphragmatic EPS or HL.^{12–17} In five cases, a two-cavity surgery for resection of the mass was performed.^{13–15} In four of these cases, an open surgical approach was chosen primarily or the minimal invasive approach had to be converted to open surgery.^{12 13 16 17} Oliveri *et al*¹⁶ described that they incised the diaphragm on suspicion and resected the intradiaphragmatic lesion. However, we cannot regard this approach as safe.

Like in our case, an aberrant arterial supply of a systemic vessel, the key anatomical landmark, was identified preoperatively in three of the reported cases.^{13 15 16}

If the chance for a two-cavity approach is high, MIS is useful for definitive diagnosis and treatment. The option of a two-step surgical procedure should be kept in mind for

Learning points

- ▶ There is a preoperative uncertainty according to localisation and 'labelling' of congenital lung lesions.
- ▶ Identifying the arterial blood supply of the lung lesion can be crucial.
- ▶ A minimal invasive surgical approach has high value for definitive diagnosis and treatment.
- ▶ Careful evaluation and decision-making is crucial. Operating time, risks of two-cavity surgery, the option of a two-timed approach, conversion to open procedure and complications by fatigue of the surgeon should be considered.

intraoperative decision-making, especially when a certain amount of time with the child being under anaesthesia has already elapsed and the patient has been asymptomatic until surgery. Despite the thoroscopic plication of the diaphragmatic eventration, the laparoscopic resection of the intradiaphragmatic HL was not complicated by adhesions, but it should be kept in mind while deciding on the strategy that a second operation could be more demanding than the first one.

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