Unexpected outcome (positive or negative) including adverse drug reactions

An interesting finding in a congenital cystic adenomatoid malformation

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Summary

Here we describe a case of antenatally diagnosed congenital cystic adenomatoid malformation. Due to increasing cyst size and fetal hydrops an antenatal drain was inserted. The lesion was excised surgically soon after birth due to significant respiratory distress. During histological assessment an area of squamous metaplasia was found within the cyst wall. This developed as a result of the drain placement. At 4-year follow-up the child remains well and has experienced no long-term respiratory sequelae.

BACKGROUND

Congenital cystic adenomatoid malformation (CCAM) is the commonest non-hereditary cystic lung lesion detected in utero. It can act as foci of infection and possibly pose a risk of malignancy. Major advancements have been made in the last decade with interventional techniques utilised in fetuses with large CCAM and in the presence of fetal hydrops.

In our case the squamous metaplasia developed as a result of trauma caused by placement of the drain. Metaplastic changes of this nature should not be confused with premalignant change that could lead to unnecessary anxiety to parents and child.

CASE PRESENTATION

Routine 20-week ultrasound scan of a 31 year-old woman showed a large lung cyst measuring 4.9 cm in diameter filling the entire side of the fetal chest, causing the heart to be displaced, with associated compression by the mass. There was evidence of marked ascites as well as oedema of the head and neck; consistent with an appearance of hydrops. No other fetal defects were reported. A repeat ultrasound showed worsening features of hydrops requiring aspiration of the cyst followed by drain insertion. Due to fetal distress the baby was delivered at 36+6 weeks gestational age weighing 2.28 kg. The baby was symptomatic with respiratory distress and required early respiratory support. CT scan confirmed a large CCAM in the left upper lobe of the lung. At surgery on day 4 of life the CT findings were confirmed and the baby underwent a left upper lobectomy. The left lower lobe was normal and expanded easily. Microscopic examination of the resected specimen confirmed a CCAM with a single area of squamous metaplasia. The consultant paediatric pathologist confirmed the presence of chronic irritation within the cyst wall and therefore probable that the squamous metaplasia reflects the presence of the antenatal drain in contact with this area.

OUTCOME AND FOLLOW-UP

Following the surgery, the neonate made an uneventful recovery and on follow-up is well at 4 years of age with no respiratory sequelae.

DISCUSSION

CCAM is a rare developmental lung anomaly. The reported incidence ranges from 1 in 25 000 to 1 in 35 000 pregnancies making it the commonest congenital lung lesion.¹

Indications for surgical excision include respiratory distress and recurrent infections. There have been reports of pleuropulmonary blastoma and bronchioalveolar carcinoma occurring in children and adults with previously diagnosed CCAM.²⁻⁴ A single case of squamous cell carcinoma has also been reported by Usui *et al.*⁵ However, more recent studies have suggested no significant increase in the incidence of malignancy in patients with CCAM compared with unaffected groups.⁶

Microscopically the cyst walls are epithelial lined comprising a variety of cell types, from low cuboidal epithelium to a ciliated pseudostratified columnar epithelium.⁷⁻⁸ Some reports state the presence of mucigenic cells within the cyst walls.⁷ Mucinous metaplasia are thought to be involved in the development of bronchioloalveolar carcinoma.⁹⁻¹¹

There has been significant technological advancement in antenatal scanning of CCAM in the last two decades which has enabled the use of fetal interventional techniques in selected cases. Currently, interventions are applied to fetuses with life-threatening conditions including hydrops fetalis or polyhydramnios. These include amnioreduction, thoracocentesis, steroid administration, CCAM resection and thoracoamniotic shunt placement.¹² There is little in the way of long-term outcome data available as yet as these techniques begin to evolve.

In our case a fetal drain was inserted due to increasing cyst size and worsening fetal hydrops. Histological findings following surgical excision revealed squamous metaplasia within the cyst wall as a consequence of the drain insertion. This could have raised unnecessary alarm bells with

the parents if this had been misinterpreted as premalignant change associated with mucinous metaplasia. There should be increased awareness of the possibility of squamous metaplasia in these patients following drain insertion which has also been noticed by other groups.¹³ The long-term consequence of squamous metaplasia is unknown at this stage and will warrant further investigation in the future.

Learning points

- Increase awareness of the possibility of squamous metaplasia within cyst wall of congenital cystic adenomatoid malformation following in utero drain placement.
- Avoid misinterpretation as premalignant change and prevent unnecessary anxiety caused to patient and parents.
- Need for long-term outcome data on fetal interventional techniques.
- This case demonstrates the successful use of fetal cystic drain insertion in the presence of hydrops fetalis.

Competing interests None.

Patient consent Obtained.

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