Adult presentation of congenital cystic adenomatoid malformation: Successful surgical management

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At thoracotomy, we found multiple, thick-walled, gelatinous cystic lesions occupying the left hemithorax. Surprisingly, these appeared to be originating within the fissure and had not replaced the lobe itself. Fortunately, this was appreciated before any vessels were divided. At the conclusion of dissection, both lobes reinflated and appeared macroscopically normal. Despite a small air leak, she had a good recovery. During an outpatient visit 3 weeks later, she reported a considerable improvement in her symptoms. Histopathology confirmed type II CCAM.

CCAM was first described in 1949. It is a developmental hamartomatous abnormality of the lung with adenomatoid cyst proliferation.1

It is believed to result from aberrant embryogenesis at or before the seventh gestational week, producing adenomatoid proliferation of the terminal bronchioles and cystic distortion of the lung architecture. Blood supply is through the pulmonary circulation.

The cause of CCAM is unknown, but the condition nearly always presents prenatally (incidentally picked up on prenatal ultrasonography) or within the first year of life, manifesting as respiratory distress in neonates or recurrent infections in older children.2

Adult presentation of CCAM is very rare, with few cases documented in the literature.3-5 Mildly affected individuals can remain asymptomatic, with the disease even being diagnosed postmortem in elderly patients who have died from an unrelated illness.

The Stocker classification subdivides CCAM according to the size of the cysts and other histologic criteria:

- Type 1: one or more cysts (2-10 cm diameter) with muci
- nogenic differentiation. This is the most common form.
- Type 2: Smaller cysts (classically 0.5-2 cm) lined by cuboid or columnar epithelium.
- Type 3: Microscopic adenomatoid cysts. These macroscopically might appear to be a solid mass.

The prognosis of CCAM is very variable. Overall mortality is quoted at 25% to 30% but might approach 100% in severely affected individuals without treatment. There is a spectrum of severity through to the asymptomatic adult. Type 3 CCAM carries the worst prognosis but fortunately is the least common.4

Treatment is primarily surgical, and although some specialist centers are now undertaking fetal surgery (predominantly indicated for those with hydrops fetalis), most documented postnatal intervention has centered around lung resection (in the form of lobectomy/pneumonectomy) to remove the lesion and relieve com-
pression of the residual lung. This initially appeared to be merited on the basis of radiologic imaging.

The advantage of surgical intervention is that it also prevents the development of secondary complications of CCAM, namely hemorrhage and recurrent infection, and the development of malignancy. Rhabdomyosarcomas, pulmonary blastomas, bronchioloalveolar carcinomas, and squamous cell carcinomas have all been described.

CCAM (especially type 2 CCAM, as in this case) is associated with other congenital anomalies predominantly affecting the renal, gastrointestinal, and cardiac systems. It is important that such patients be routinely screened to anticipate any further problems before they manifest clinically.

This case highlights a rare presentation of CCAM in an adult. It is also testament to the fact that unusual conditions should be managed on an individual basis because premeditated lobectomy (the treatment of choice for previously documented cases of CCAM and merited on imaging) would have resulted in an unnecessary lung resection in this patient.

References

Figure E1. Coronal computed tomogram showing extensive replacement of left lung with cystic lesions.

Figure E2. Sagittal chest computed tomogram demonstrating some residual upper and lower lobe lung parenchyma.