bodies. 9. Biconvex upper and lower endplates, and platyspondyly (spinal bone between the chest and the abdomen). 10. Enlarged "oar-shaped" ribs with periosteal reaction. **E,** Upper limb. 11. Metaphyseal fraying and periosteal cloaking. **F,** Lower limb. 12. Hypomineralization, classic metaphyseal fraying subperiosteal bony destruction and periosteal reaction and new bone formation (periosteal cloaking), most prominent at the metaphyses of the femur. **G,** Lower limb. 13. Hypomineralization, classic metaphyseal fraying subperiosteal bony destruction and periosteal reaction, and new bone formation (periosteal cloaking), most prominent at the metaphyses of the femur. 14. Anterior convex bowing deformity of the distal tibia.

Cyst-Like: An Incongruent Chest Radiograph

A 10-month-old boy was admitted with cough and fever despite a course of oral antibiotics. Oxygen saturation was normal in air. There were reduced breath sounds posteriorly on the left side of the chest, with normal percussion. Bowel sounds were not heard in the chest.

Chest radiography (**Figure 1**) showed an unusual multilocular cystic lesion, suggestive of bowel loops, on the left side of the chest, with lung tissue seen inferiorly and minimal mediastinal deviation to the right. This suggested a left-sided diaphragmatic hernia. Chest computed tomography (**Figure 2**) revealed a left sided cystic parenchymal lung lesion.

Congenital diaphragmatic hernia is being increasingly diagnosed antenatally. Postnatally, it can present with respiratory distress, a scaphoid abdomen, bowel sounds in the chest and abnormal chest radiograph. It occasionally presents in childhood with respiratory or abdominal symptoms.[1] The differential diagnosis includes congenital cystic adenomatoid malformation, bronchogenic cyst, pulmonary sequestration, or necrotizing staphylococcal pneumonia. Congenital thoracic malformations are usually smaller with later presentation or, if extensive, evident from birth or antenatally. Children with staphylococcal pneumonia would be very ill.

Open lobectomy and histology (**Figures 3** and **4**) revealed the rare primary neoplasm pleuropulmonary blastoma (PPB), type II. The 3 histological subtypes of PPB correlate with age of presentation and long-term outcome.[2] Type I has multiloculated cysts with thin septa. Types II and III are very aggressive, with cystic and solid components in type II and only solid components in type III, in which survival is the lowest.[3] In 25% of cases PPB is associated with a germline loss-of-function (non-sense) mutation in the *DICER1* gene on chromosome 14q32, for which this patient tested positive. A second tumor-specific missense mutation in the RNase IIIb domain may also be present, for which this patient was not tested. Patients with the *DICER1* mutation and their families should have surveillance for PPB, thyroid nodules and tumors, germ cell tumors in females, Wilms tumor, and retinoblastoma.[4,5]

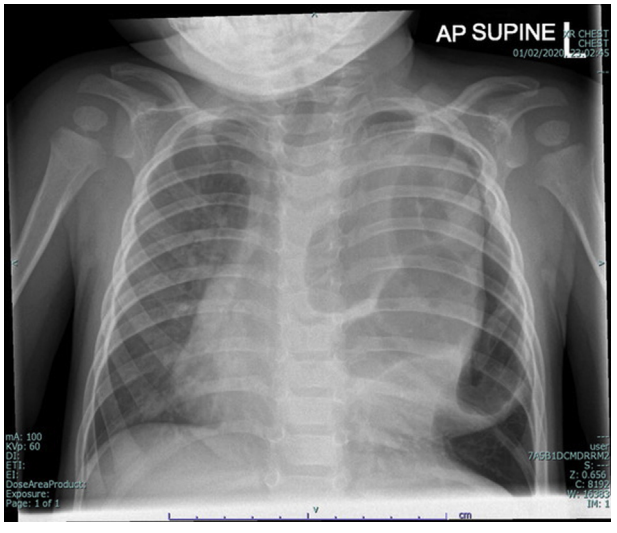

Figure 1. Chest radiograph showing the unusual cystic lesion.

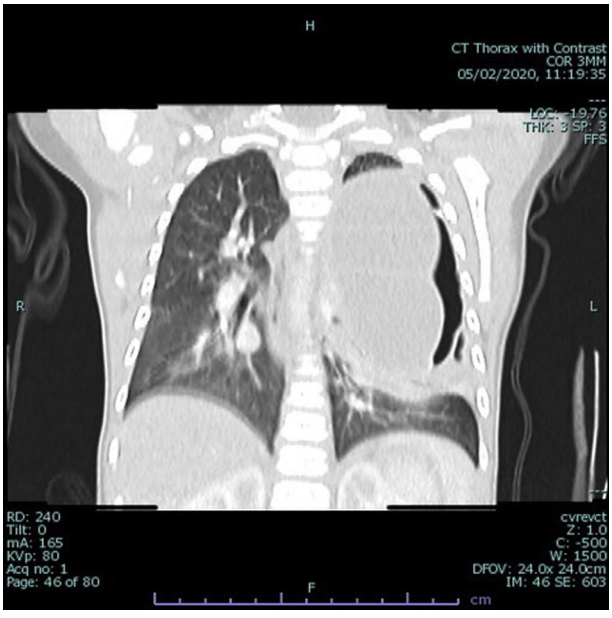

Figure 2. Computed tomography scan of the chest showing a large multiloculated fluid-filled mass within the left hemithorax causing mediastinal deviation to the right.

J Pediatr 2021;229:304-5.

https://doi.org/10.1016/j.jpeds.2020.09.036

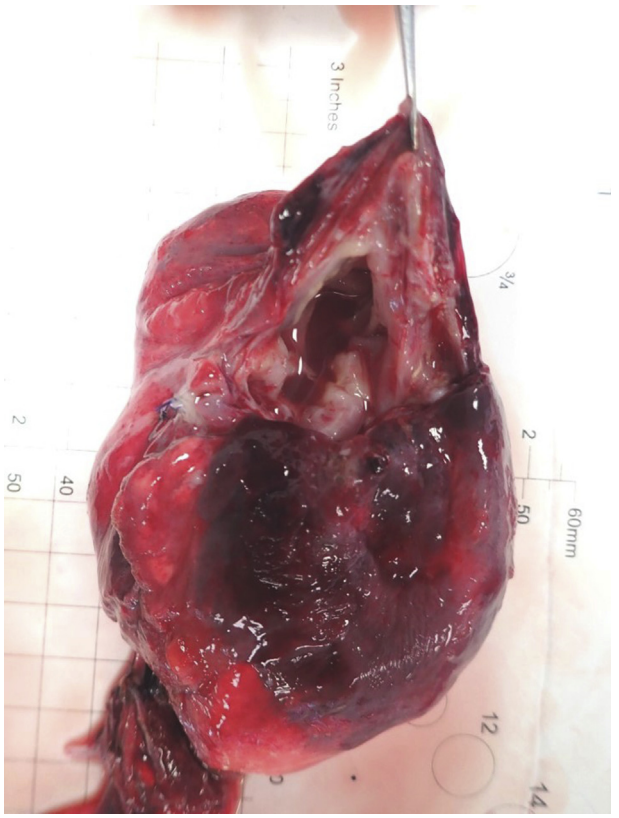

Figure 3. Pathology specimen removed during thoracotomy.

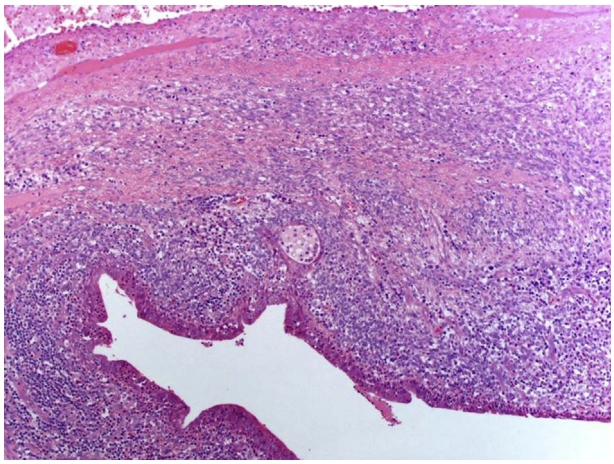

Figure 4. Histology slide showing a cyst with benign respiratory epithelium and underlying malignant small, round blastema-like cells and elongated sarcomatoid cells in the thick septum.

Data Statement

Data sharing statement available at www.jpeds.com.

Andrew Hallett, MRCPCH
Department of Pediatrics
Royal Glamorgan Hospital
Llantrisant, Wales

Lena Uzunova, MRCPCH
Department of Pediatric Oncology
University Hospital of Wales
Cardiff, Wales

Lena Thia, MRCPCH
Department of Respiratory Pediatrics
University Hospital of Wales
Cardiff, Wales

References

1. Zaleska-Dorobisz U, Bagłaj M, Sokołowska B, Ładogórska J, Moroń K. Late-presenting diaphragmatic hernia: clinical and diagnostic aspects. Med Sci Monit 2007;13(Suppl 1):137-46.
2. Messinger YH, Stewart DR, Priest JR, Williams GM, Harris AK, Schultz KA, et al. Pleuropulmonary blastoma: a report on 350 central pathology-confirmed pleuropulmonary blastoma cases by the International Pleuropulmonary Blastoma Registry. Cancer 2015;121:276-85.
3. Knight S, Knight T, Khan A, Murphy AJ. Current management of pleuropulmonary blastoma: a surgical perspective. Children (Basel) 2019;6:86.
4. Schultz KA, Williams GM, Kamihara J, Stewart DR, Harris AK, Bauer AJ, et al. DICER1 and associated conditions: identification of at-risk individuals and recommended surveillance strategies. Clin Cancer Res 2018;24:2251-61.
5. The International Pleuropulmonary Blastoma/DICER1 Registry. https://www.ppbregistry.org. Accessed June 15, 2020.