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An Unusual Cause of a Tension Pneumothorax

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Abstract

We present a case of spontaneous tension pneumothorax in a 17 year-old man treated with an intercostal chest drain. Subsequent radiology post resolution of pneumothorax showed a large bulla culminating in bullectomy. Histological specimens were consistent with mucinous type 1 Congenital Pulmonary Airway Malformation (CPAM) and subsequently he managed in keeping with having a primary lung adenocarcinoma. He underwent completion lobectomy without complication and he is currently under follow up in a respiratory outpatient clinic.

Keywords: Congenital Pulmonary Airway Malformation; Pneumothorax; Pathology; Adenocarcinoma

Introduction

A 17-year-old man with no past medical history, who had never smoked, presented with a right-sided tension pneumothorax. An intercostal chest drain was inserted without complication with rapid clinical improvement and re-inflation of the lung.

Chest x-ray 2 days post drain insertion showed an area of lucency at the right lung base and CT (computed tomography) showed a 14 x 13 x 9.5cm right lower lobe bulla (figure 1). The intercostal drain was removed once the air leak had settled and the patient was referred to the neighboring thoracic surgical unit for bullectomy. Alpha-1 antitrypsin level was normal.

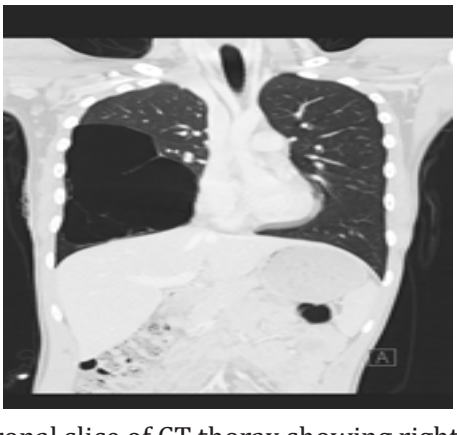

Figure 1: Coronal slice of CT thorax showing right basal bulla.

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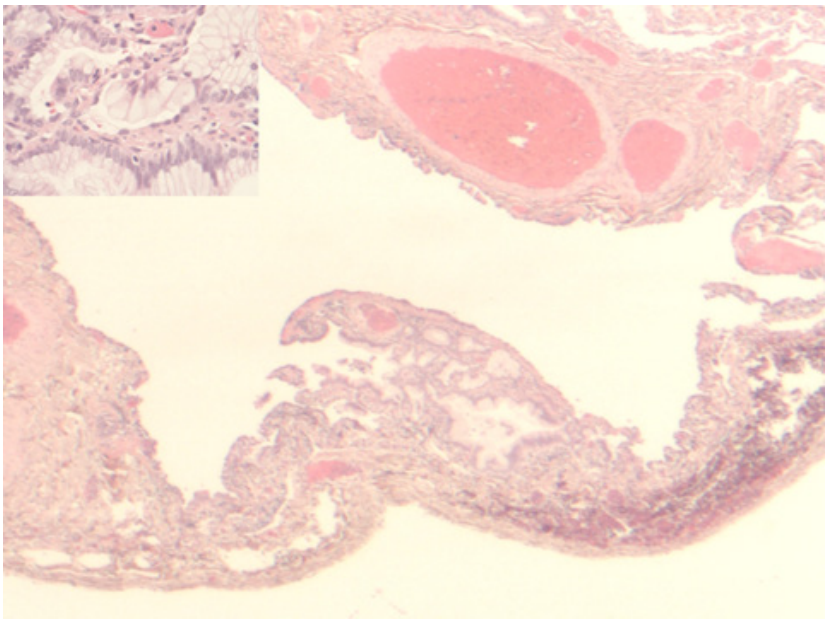

Figure 2: Cystic lesion at low magnification (HE x20) mainly lined by cuboidal or cylindrical epithelium. Focal areas of bland mucinous epithelium layering with tufting are also present (inset. HE x400).

The patient underwent bullectomy and talc pleurodesis without complication. Histological specimens showed evidence of type 1 congenital pulmonary airway malformation with patchy monomorphic populations of columnar mucinous cells lining the wall of the cyst with a lipidic pattern extending focally into the surrounding alveolar wall (Figure 2).

As thought to be a carcinoma-in-situ, a staging CT was performed which did not show any distant metastasis. He subsequently underwent completion lobectomy without complication and was well on subsequent out-patient review.

Discussion

Congenital pulmonary airway malformation (CPAM) is a rare developmental anomaly in the lower respiratory tract of the newborn. Incidence is thought to be in the range of 1:8300 to 1:35000 of live births. Affected individuals are usually asymptomatic until later in life through some neonates will present with respiratory distress.[1] CPAM is usually identified on neonatal ultrasound scans although they occasionally present in adulthood with pneumothorax, hemoptysis or recurrent chest infections.

There are five classifications of CPAM: type 0-4. Type 0 (1-3%) is the rarest form [2]. The cysts are usually widespread and small measuring a maximal diameter of 0.5cm, originating from tracheal or bronchial tissue. Affected neonates tend to die at birth.

Type 1 is commonest, constituting 50-65% of all CPAMs and comprise either single or multiloculated cystic lesions ranging from 2 to 10cms in diameter lined with pseudostratified columnar epithelium[3]. Mucinous producing cells are seen in about 33% of the patients and are associated with lung malignancies. The exact incidence of malignancy is poorly delineated, although a single center review of 16 patients by MacSweeney et al quoted an incidence of adenocarcinoma in situ of 31%.[4] No recurrence has ever been seen post resection. Other lung malignancies seen in CPAM include pleuropulmonary blastomas, rhabdomyosarcomas, and adenocarcinomas.

Type 2 CPAMs have an incidence of 15-20% and consist of multiple cysts that blend into solid tissue and can resemble terminal bronchioles. They are lined by ciliated cuboidal or columnar epithelium and are sometimes mistaken for pulmonary sequestration with their own blood supply, though hybrid forms may exist.[2] Type 2 CPAM is not associated with malignancy and typical sizes range from 0.5 to 2cm.

Type 3 CPAMs have an incidence of 5-10% and are often large cystic lesions which can affect an entire or multiple lobes. They consist of an adenomatoid proliferation of distal airways and/or are acinar in origin. They can be entirely solid or a combination of cystic and solid lesions. They are not associated with malignancy and affected neonates usually present with respiratory distress or death during the neonatal period.[2]

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Type 4 CPAM compromise 5-10% of all CPAMs. The cysts consist of flattened, non-ciliated, alveolar lining cells with no mucus or skeletal cells. They have a maximal diameter of 7cm and may present at birth or in childhood with pneumothorax. Type 4 CPAMs have a strong association with malignancy, especially pleuropulmonary blastomas.

We would like to highlight that the diagnosis of type 1 CPAM, in this case, was incidental. Due to the large bulla seen on CT, and the high risk of recurrent pneumothorax inherent to this, bullectomy was performed with the postoperative histological findings favoring type 1 CPAM. Although current evidence is sparse, existing literature supports an association between adenocarcinoma in situ and type 1 CPAM with mucinous cells, as in this case[5]. Surgical resection remains the definitive treatment for this rare condition.

Conclusion

In conclusion we would like to present a case of unusual case of pneumothorax with underlying CPAM, rarely seen in adulthood.

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