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# Thoracoscopic resection of a lung keratocyst associated with pulmonary sequestration in a neonate

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ABSTRACT

*Introduction:* Keratocyst is a cystic lesion that has distinctive histological features composed of an epithelial lining with surface keratinization that is corrugated and predominantly parakeratotic. Although rare, they present in human pathology mainly as odontogenic keratocysts and have occasional association with Gorlin-Goltz syndrome.

*Case report:* We present a neonate with an antenatal suspicion of congenital pulmonary airway malformation. While stable and imaging confirmation studies were accomplished, thoracoscopic exploration and excision of a bronchogenic-like lung cyst attached to an extralobar pulmonary sequestration superomedial to the left upper lobe were performed successfully at day 44 of life. Histopathological study has revealed keratocystic features of the lung cyst attached to a pulmonary sequestration tissue.

*Discussion and conclusion:* Up to our knowledge and review of the literature, this is the first case and occurrence of Lung Keratocyst. Long-term follow up is needed to assess the outcome of the thoracoscopic management of such lesion, and its hypothetic relation with odontogenic keratocysts or any associated syndromes.



## 1. Introduction

The term “Lung Keratocyst” is not existing in literature. Keratocysts in literature are almost always of odontogenic origin and called “Keratocystic Odontogenic Tumors” that affect the mandible in most cases. We report an exceptional case of a lung cyst of histopathological keratocystic features associated with an extralobar pulmonary sequestration that underwent a thoracoscopic resection.

## 2. Case report

A 37 days old male baby, who was referred to our care from a peripheral hospital with an antenatal diagnosis of congenital pulmonary airway malformation and a postnatal suspicion of a bronchogenic cyst according to postnatal chest radiographs and computed tomography images. The child was term and had a birth weight of 3700 g with stable hemodynamic and cardiorespiratory status after birth. He had good general health conditions with no any respiratory symptom. Chest radiograph has shown a well demarcated round radio-opaque structure occupying the left upper hemithorax with slight shifting of mediastinum toward the right side. Upper gastrointestinal contrast study has ruled out any communication with the gastrointestinal tract (Fig. 1). Computed tomography scan with contrast has shown a non-enhanced round cystic lesion of fluid density occupying the left paramediastinal region in corresponding to the apico-dorsal segment of the left upper lobe, 23 × 24 × 24 mm in size, in close contact with the descending thoracic aorta and pushing a what suspected to be the apical segment arterial branch anteriorly (Fig. 2). Our primary diagnosis was a bronchogenic cyst and therefore thoracoscopic resection was planned.

At day 44 of life, thoracoscopy was performed and we found a

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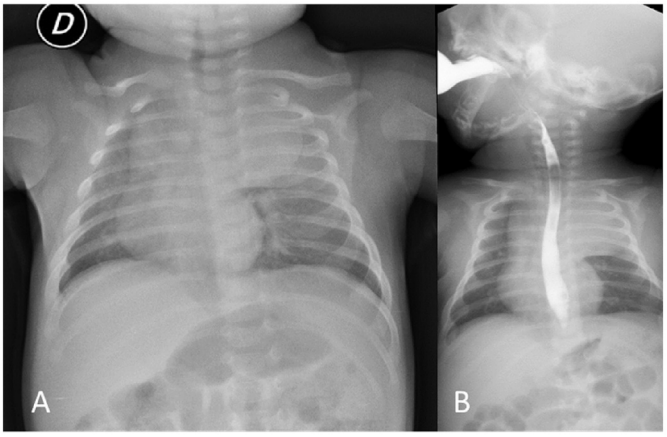


**Fig. 1.** A) Chest radiograph showing the round radio-opaque lesion that occupies the left upper hemithorax with slight shifting of mediastinum toward the contralateral side. B) An upper GI contrast study showing normal esophageal contour with no evidence of communication with the cystic lesion.

bronchogenic-like cyst that was firmly attached to an extralobar pulmonary sequestration and to the adjacent normal lung tissue as well. It was difficult to dissect and isolate the cyst from the surrounding structures, that is why it was evacuated and opened where whitish creamy fluid came out of the cyst compatible with keratinaceous material. The part that was attached to the normal lung tissue was removed using Ligature™. The feeding vessel of the sequestration tissue was doubly ligated using Endoloops® and transected. The sequestration tissue with the attached remaining part of the lung cyst were removed and extracted out together (Fig. 3). Chest drain was left in place and operative time was 100 min. The patient had a smooth recovery postoperatively and he was discharged home at the tenth postoperative day. Follow up for 6 months does not show any complication.

Histological examination of the specimen has reported a pulmonary sequestration that is characterized by unaerated pulmonary parenchyma, serpiginous vessels of systemic type, and subpleural arterioles. The attaching cyst looks of bronchogenic origin due to the presence of sero-mucinous glands, cartilage and layers of smooth muscle cells, but surprisingly, it shows typical histologic characteristics of a keratocyst as well. These characteristics include the presence of few layers of squamous epithelium with incomplete keratinization, the corrugated and irregular luminal epithelial layer, and the complete absence of mucous-secreting goblet cells all through the wall of the lung cyst (Figs. 4 and 5).

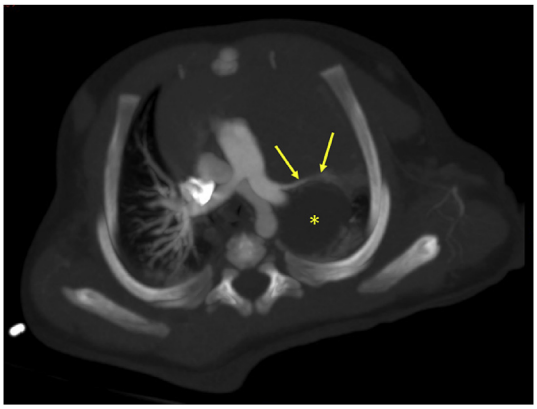


**Fig. 2.** Thoracic CT scan showing the cystic lesion at the left upper hemithorax (asterisk). The cyst is pushing a systemic vessel anteriorly (arrows) which was found intraoperatively to be the feeding vessel of an extralobar pulmonary sequestration.

## 3. Discussion

We describe the first case of an unusual occurrence of a lung cyst which has histological features of both a bronchogenic cyst and a keratocyst, with an unusual association with upper extralobar pulmonary sequestration. The lung cyst and sequestration were resected by thoracoscopy.

When mentioning a keratocyst, it has no occurrence in literature rather being an odontogenic one. Odontogenic keratocysts (OKC) are cystic lesions of the bone that are lined with keratinized epithelium and thought to originate from the dental lamina [1]. It is known for its high recurrence rate, aggressive behavior, and its occasional association with the nevoid basal cell carcinoma syndrome (NBCCS) or the Gorlin-Goltz syndrome [2]. Three-fourths of OKCs present in the mandible [3]. The term OKC was first coined by Philipsen in 1956 [4], until in recent years when the WHO has recommended that the term “keratocystic odontogenic tumor” (KCOT) replace the term “odontogenic keratocyst”, as it better reflects the neoplastic nature of the lesion [5]. Diagnosis is usually made with histopathology studies, where the main histologic features are composed of a thin lining epithelium of a parakeratinized surface which is typically corrugated, rippled or wrinkled, a remarkable uniformity of thickness of the epithelium, usually ranging from 6 to 10 cells thick, and a prominent palisaded, polarized basal layer of cells often described as having a ‘picket fence’ or ‘tombstone’ appearance [6].

Gorlin-Goltz syndrome is a rare inherited multisystem disorder, where affected patients have multiple developmental anomalies and early development of multiple basal cell carcinomas, odontogenic keratocysts, especially in the mandible, palmar and plantar pitting, ectopic intracranial calcification, and family history of NBCCS [7,8]. It has never been reported an association between any of the OKC or the Gorlin-Goltz syndrome with a lung sequestration or any other bronchopulmonary malformation.

On the other hand, bronchogenic cysts (BC) are congenital anomalies arising as an abnormal ventral budding of the tracheobronchial tree and so have an epithelium which is similar to bronchi [9]. Histologically, they are lined by ciliated pseudostratified respiratory epithelium or metaplastic squamous cells and have focal areas of hyaline cartilage, smooth muscle, and bronchial glands within their walls [10]. Furthermore, by definition and the histological components, our cyst is neither an enteric duplication cyst, an epidermal cyst, nor a dermal cyst.

An association of pulmonary sequestration (PS) and BC is unusual. We have reviewed the MEDLINE® database searching for the reported cases of PS that are associated with other pathologies, and specifically with BCs (Table 1). There is a wide range of associated anomalies and disorders but with few reported cases of each. There are 12 reported cases that describe an association between PS and BC. They include 7 cases of extralobar PS type [11–17], and 5 cases of intralobar type [18–22]. In addition, there are five other reported cases of association between PS and enteric duplication cysts.

Leaning on the aforementioned results, we suppose our case report to be the first occurrence of a “Lung Keratocyst”, of a bronchogenic origin, in association with an unusual left upper lobe extralobar PS. Hereby, we assume to introduce the new term of “Bronchogenic Keratocyst”, where the tissue of origin is not the usual odontogenic one. Could there be an association between bronchogenic and odontogenic keratocysts? Would bronchogenic keratocysts present aggressiveness and infiltrative behavior