Unusual Presentation Of A Congenital Bronchogenic Cyst In A Young Child

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Citation

Abstract
A bronchogenic cyst is a rare developmental aberration that occurs in the paediatric age group. Cutaneous presentations of bronchogenic cysts are rare in all age groups. Previous reports of cutaneous manifestations of bronchogenic cysts have been described as cystic swelling, most frequently with a suprasternal location. We report a unique presentation of a child with an anterior chest wall mass in the sternal region, which was identified as a bronchogenic cyst.

INTRODUCTION
Bronchogenic cysts are rare benign congenital developmental abnormalities of the embryonic foregut. They are usually located in the mediastinum and intrapulmonary regions. The skin is a rare site for bronchogenic cyst. It is difficult to diagnose it clinically and in almost all cases the diagnosis was established by histopathological examination.

CASE REPORT
A 10-year old child presented in outpatient department with painless swelling in the anterior chest wall in presternal area since birth. The swelling was initially of smaller size, but for the last few months it had progresses to the present size. On clinical examination the swelling was present in the midline in upper part of presternal area. It was 5x5 cm soft cystic fluctuant mass, with overlying skin showed no signs of inflammation. USG abdomen showed a cystic mass with no underlying communication. The patient was scheduled for elective surgery through a transverse elliptical incision. Surgical exploration revealed a cystic mass (Fig.1) which was excised and sent for histopathological examination. The histopathology report revealed a bronchogenic cyst (Fig.2). There was no recurrence of a lesion for a 6 month follow-up.

DISCUSSION
Bronchogenic cysts are rare congenital lesions with a
reported prevalence of 1 in 42,000 to 1 in 68,000.1 Bronchogenic cysts occur as a result of a developmental fault during the division and budding of the tracheobronchial tree in the embryonic period. They are located either intrathoracic or extra-thoracic. Fifty percent of cysts are located in the posterior mediastinum, 14% in the superior mediastinum, and 35% in the pericardial area.2 Intra-thoracic cysts may remain connected to the tracheobronchial airway by either a fibrous cord or a patent bronchus-like connection.3 In the absence of an attachment, bronchogenic cysts can migrate and occasionally become displaced anteriorly by fusion of the mesenchymal bars forming the sternum.

Cutaneous bronchogenic cyst is a rare and usually solitary lesion that is four times more common in males than in females. The most common location is the suprasternal notch, followed by the pre-sternal area, the neck, and more rarely the scapular area.4,5 They are usually asymptomatic, but if the cyst is large and present in cervical area, symptoms may occur, including dyspnoea, respiratory distress, cough and dysphagia. Very occasionally, secondary infection may occur which results in sinus tract formation and external drainage of purulent material if the cyst is superficial or with abscess formation if the cyst is deep.

Figure 3
Microphotograph of bronchogenic cyst showing respiratory (pseudo stratified ciliated columnar) epithelium [ thick arrow] with underlying smooth muscle [thin arrow]

There is no pathognomonic sign and symptoms and it is clinically difficult to diagnose and differentiate it from other cysts, which are commonly located in midline particularly thyroglossal duct cysts and branchia cysts. In such cases histological findings are crucial to distinguish this lesion from branchial cyst, thyroglossal duct cyst, cutaneous ciliated cyst, dermoid cyst, infundibular cyst, and trichilemmal cyst.6 Thyroglossal duct cysts present as midline cystic nodules on the anterior neck in children or young adults. The characteristic histological feature is the presence of thyroid follicles composed of cuboidal cells surrounding a homogeneous pink material and a lymphocytic infiltrate. Branchial cysts appear in the preauricular area, mandibular region, or along the sternocleidomastoid muscle. Histologically, these cysts are lined by stratified squamous epithelium or by pseudostratified ciliated columnar epithelium. They are surrounded by a lymphoid infiltrate that includes lymphoid follicles. Trichilemmal cysts are derived from the outer root sheath of the hair follicle. Histologically, they are lined by stratified squamous epithelium and are filled with keratin. The characteristic histological findings seen in bronchogenic cyst are a ciliated pseudo-stratified epithelium respiratory-type lining with the presence of goblet cells. Smooth muscles and cartilage may also be present.6

Surgical excision of the cyst is the treatment of choice followed by histological examination to confirm the diagnosis. Complete excision and follow-up is advocated, as recurrence had been observed after incomplete removal. There are also few reports of cases in which malignancy has arisen from a congenital bronchogenic cyst in adults. These includes mucoepidermoid carcinoma and a case of melanoma, emphasizing the importance of its total surgical excision.7,8

CONCLUSION
In conclusion we highlight the following important facts through this case:
The clinical observation of an asymptomatic mass in presternal or neck region should include the possibility of bronchogenic cyst and its presentation can be mimicked by branchial cleft cyst or thyroglossal duct cyst. Definitive diagnosis is not always possible preoperatively and also there is risk of malignant transformation, we recommend the complete surgical excision of all suspected bronchogenic cyst.

References
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