Case Report

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Cutaneous Bronchogenic Cyst: An Unusual Cause of Lump: A Diagnostic Challenge

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ABSTRACT

Bronchogenic cysts are rare developmental anomalies, seen primarily in the pediatric population. Cutaneous manifestations of bronchogenic cysts are still rarer. This lesion poses diagnostic challenge for the clinicians and in almost all, histopathological examination gives the precise diagnosis. We report a case of presternal cystic lesion in 11 year old boy, clinically diagnosed as dermoid cyst, however the lesion was suggestive of cutaneous bronchogenic cyst on histopathology.

Keywords: *Bronchogenic Cyst, Cutaneous, Presternal*

Introduction

Bronchogenic cysts are rare benign congenital developmental anomalies of the ventralforegut.[1] They are endodermal cysts and when they are lined by the respiratory epithelium predominantly, it is termed as a bronchogenic cyst.[2]Bronchogenic cysts have a reported prevalence of 1 in 42,000 to 1 in 68,000. [3] They are usually intrapulmonary and most common extrapulmonary location is the mediastinum,particularly posterior to the carina.[2] In a large series composed of 2,163 cases of mediastinal lesions, 3.3% were foundto be bronchogenic cysts. [3]Unusual sites of presentation include skin, subcutaneous tissue,pericardium and retroperitoneum.[2,4]Under cutaneous location, most common site is the suprasternal notch, followed by presternal area, neck and scapula.[5]

Case report

An 11 year old male child presented to the outpatient department with a swelling over manubrium sterni since birth. The swelling was initially smaller in size and in the past few months has progressed to its present size. On examination, a cystic, fluctuant, nontender, 2x 2 cm mass was noted in the upper presternal region. The overlying skin showed no signs of inflammation or presence of punctum. There were no other associated respiratory complaints. The child was scheduled for an elective surgery. Surgical exploration revealed a cystic mass which wascompletely removed and was sent for histopathological examination. Grossly a single irregular, grey brown toyellow fatty tissue bit was identified which on cut section showed a tiny cyst surrounded by yellow fatty areas.Microscopy revealed a cyst lined by ciliated columnar respiratory epithelium, overlying oedematous fibrocollagenous stroma with occasional mucinous glands, scattered scant lymphocytic infiltrate, muscle fibres (Massonstrichrome positive) and congested blood vessels overlying fibroadipose stroma (Fig 1,2,3,4). A diagnosis of Cutaneous bronchogenic cyst was given.

Discussion

Cutaneous bronchogenic cysts are rare cystic lesions that occur due to congenital developmental aberrations.Ever since its first description in literature in the year 1945, not many cases of cutaneous bronchogenic cysts have been reported.[4]This is possibly due to its rarity and at times unusual presentation. The diagnosis of cutaneous bronchogenic cyst is usually a histological surprise and preoperative diagnosis of this entity is seldom made.

Majority of these lesions have been reported in pediatric population, however, few cases involving the adults have also been mentioned in literature.[3] Sanli et al. [3] reportedbronchogenic cyst in the cervical area in a 48year old woman. Shreya et al. [6] identified a case of cystic cervical mass proved to be bronchogenic cyst on histopathology in an elderly female with history of thyroiditis. Moz et al.[7] also reported bronchogenic cyst in a 39 year old male. In pediatric population, most cases present shortly after birth, however until 14 years of age cases have been reported. [8] Bronchogenic cysts are four times more common among the male population than females. [9]

Bronchogenic cysts can be intrathoracic or extrathoracic in location. Most common extra thoracic location being the mediastinum. Nearly 50% of these cysts are located in the posterior mediastinum, 14% in the superior mediastinum and around 35% are seen in the pericardial area.[1]

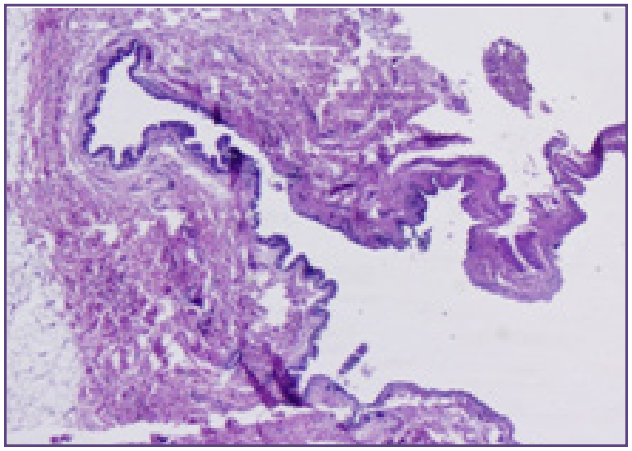

Fig. 1: Cyst overlying fibrocollagenous and fibroadipose stroma H&E X20.

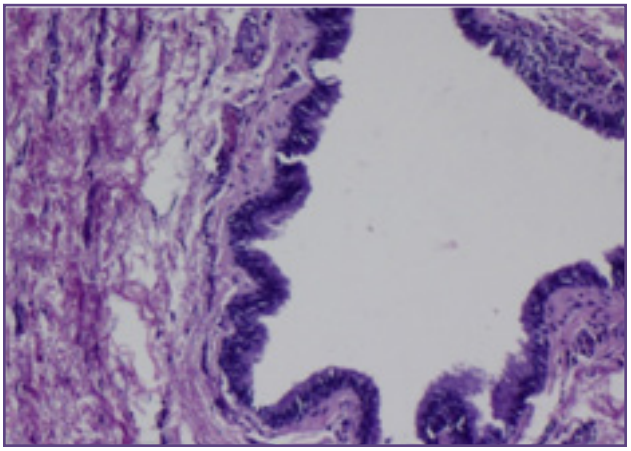

Fig. 2- Respiratory epithelium overlying oedematous fibrocollagenous stroma with mucin glands H&E X100.

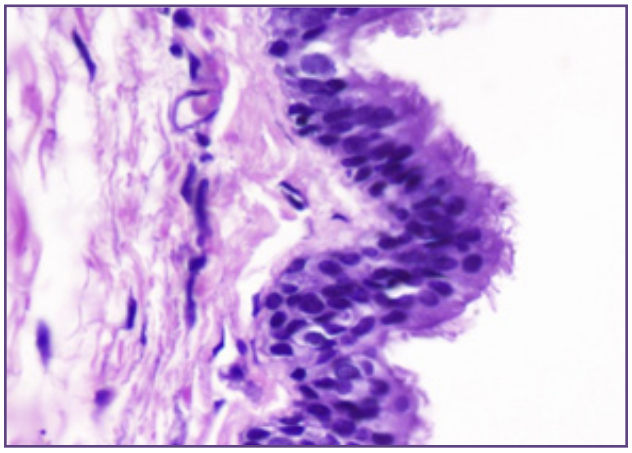

Fig. 3: Cyst lined by pseudostratified ciliated columnar respiratory epithelium H&E X400.

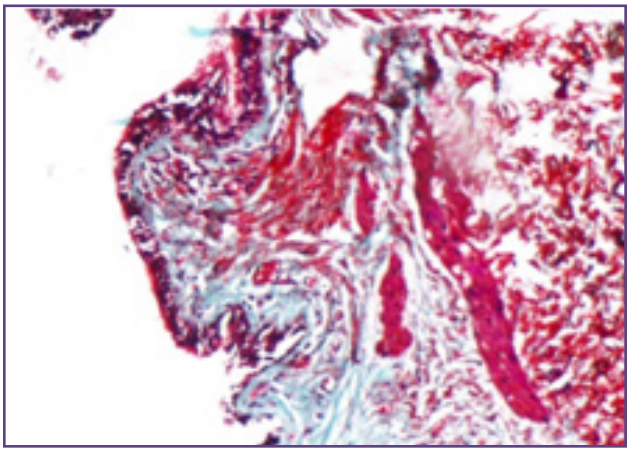

Fig. 4: Smooth muscle fibres in red Masson trichrome X100.

The pathogenesis for the development of bronchogenic cyst lies in embryogenesis. During the beginning of the 5th week of gestation, laryngo-tracheal groove separates the primitive foregut into dorsal and ventral structures.[2,4,5] The dorsal anlage forms the oesophagus, whilethe ventral anlage develops into tracheobronchial tree.Bronchogenic cysts form due to abnormal development occuring in the distal tracheobronchial tree.[8] However, the origin of the cutaneous bronchogenic cysts can also be explained by another hypothesis which states that a part of the anterior portion of the developing lung gets pinched off during the process of fusion of the right and left bars of sternum precursors, leading topresternal localization of the bronchogenic cysts.[4]As far as extrathoracic locations at unusual sites like neck, chin,shoulder are concerned, pathogenesis can be explained by the migration of these sequestered structures in the developing embryo or in situ development of metaplastic respiratory epithelium from pre-existing cutaneous tissue and primary anomalous differentiation in the developing skin.[2]

These lesions usually present as an asymptomatic midline subcutaneous cystic and nontender nodule. [8]If the cysts are large in size and localized in the cervical area, then symptoms like dyspnoea, respiratory distress, cough and dysphagia may be seen. The cases of secondary infection may result in the formation of sinus tract causing external drainage of purulent material or an abscess formation.[1] However, our case did not present with any respiratory symptoms or any associated secondary infection. Due to the lack of any characteristic clinical features it is clinically challenging to distinguish bronchogenic cysts from other cysts occurring in the same region.

Bronchogenic cysts are characteristically lined by pseudostratified ciliated columnar respiratory epithelium with interspersed goblet cells. Smooth muscle fibers are associated in 70%, mucus gland in 53% and cartilage in upto 7% cases.[1,4,5] Our case also had similar clinical presentation and microscopy to warrant the diagnosis of bronchogenic cyst.

A differential diagnosis for bronchogenic cyst includes branchial cleft cyst, thyroglossal duct cyst, mature cystic teratoma, cutaneous ciliated cyst, dermoid cyst, epidermal inclusion cyst, infundibular cyst and trichilemmal cyst. Characteristically, branchial cysts are located either along the sternocleidomastoid muscle, or mandibular region or in the preauricular region. They are predominantly lined by stratified squamous epithelium or rarely by pseudostratified ciliated columnar respiratory epithelium with surrounding lymphoid follicles and lymphocytic infiltrate.Thyroglossal duct cysts are noted as midline cystic neck nodules and characterised by the presence of thyroid follicles and lymphocytic infiltrate.Trichilemmal cysts are derivative of outer root sheath of hair follicle, can occur at any site and are lined by stratified squamous epithelium withcharacteristic absence of granular layer andabrupt keratinization. Cutaneous ciliated cysts usually occur in females, seen in the lower extremities and lined by ciliated columnar epithelium with papillary projections resembling fallopian tubes.Dermoid cysts can be differentiated as they possess epidermal appendages and epidermal inclusion cyst have stratified squamous layer as the liningepithelium.[1,2,4,5]

Bronchogenic cysts are benign lesions, however few cases with malignant transformation have been reported in literature. Tanita et al. [10] have reported development of malignant melanoma from a cutaneous bronchogenic cyst over scapular region. Calzada et al. [11] mentioned about a case of poorly differentiated adenocarcinoma arising from a bronchogenic cyst and Jakapovic et al. [12] reported transformation of bronchogenic cyst into large cell carcinoma.

Treatment of choice for bronchogenic cyst is complete surgical excision. Due to the possibility of development of malignancy in these lesions, histopathological examination of the entire cyst is considered very important. [1] Incomplete excision may lead to recurrence, hence follow up is advocated.Hasegawa et al .[13]reported a case where recurrence of bronchogenic cyst in a 42 year old man occurred nearly 15 years after initial resection and the probable cause stated was incomplete resection.

Conclusion

Bronchogenic cysts are uncommon congenital malformations. They may occur at unusual locations and thus should be included in the differential diagnosis of cystic and nodular skin lesions on the upper chest, neck and upper back. A high index of clinical suspicion is required to diagnose this lesion preoperatively. Histopathological examination is mandatory and plays an integral role for establishing precise diagnosis.

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