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Unanticipated mystery unravelled

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Abstract
Embryologically the ventral bud develops into primitive foregut and tracheobronchial tree. Any malformation in this would cause a cystic lesion along the tracheobronchial tree or the foregut. The incidence of this occurring is about 1 case per 68,000 populations. Here we present a case of sublingual swelling in a 22 year young female, with no other complaints. Her history was impeccable. All routine investigations were normal. MRI head and neck suggested a ranula. She was planned and taken up for excision biopsy. The excised specimen was sent to histopathological examination which revelled the swelling to be a bronchogenic cyst. Bronchogenic cysts are an embryological anomaly and are found anywhere along the foregut. They can present in both adults and children. 7 to 15 % of all foregut cysts are bronchogenic cysts says literature. These cysts are generally asymptomatic but have a 10% chance of turning malignant.

Keywords: tracheobronchial tree, histopathological

Introduction
Bronchogenic cyst is a congenital malformation of the ventral portion of the primitive foregut, causing a bronchopulmonary alteration. This usually occurs between the fifth and sixteenth week of pregnancy, when the primitive fore gut divide into dorsal oesophagus and ventral tracheobronchial tree. These cysts may be single or multiple and are usually located in close relation to the lung parenchyma or tracheobronchial tree. These may be symptomatic or asymptomatic depending on their proximity to the airway. Symptoms may range from frequent airway infections, stridor, wheezing, dysphonia and difficulty breathing. Lesions can also occur in the neck, pericardium, diaphragm, paravertebral and sub pleural regions. These are usually asymptomatic and are diagnosed in young adults. The bronchogenic cysts have ciliate columnar epithelium like the bronchioles and may contain cartilage, muscles and mucous glands [1–4].

Case
Here we present a 22 year old female, who came with complaints of sublingual swelling for the past 20 years. She had no other complaints and no other significant history. All routine blood investigations were done and found to be within normal limits. MRI head and neck was done which suggested features of a ranula. [Figure 1] She was thus taken up for surgery, excision biopsy of the swelling under general anaesthesia. [figure 2] Intra-operative and post-operative periods were uneventful. She was discharged on POD 2. Histopathology reports revealed the excised specimen to be a bronchogenic cyst as it contained ciliated columnar epithelium. [figure 3] On further follow up of the patient, she had no complaints and no other swelling on screening.

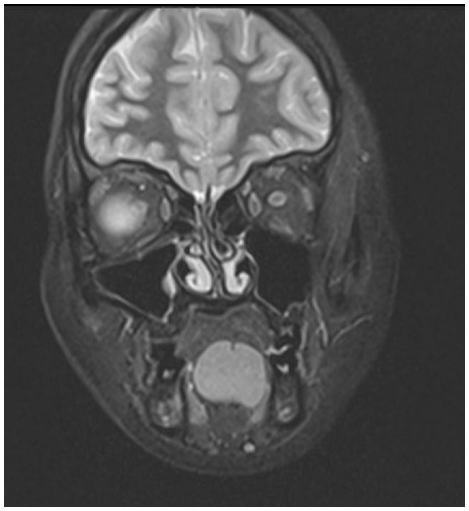

Fig 1: MRI head and neck shows features suggestive of ranula.

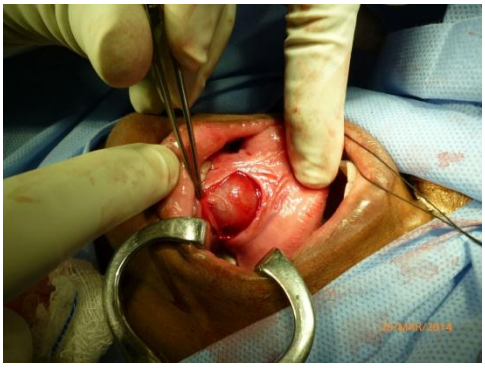

Fig 2: Intra operative picture of the swelling after incision

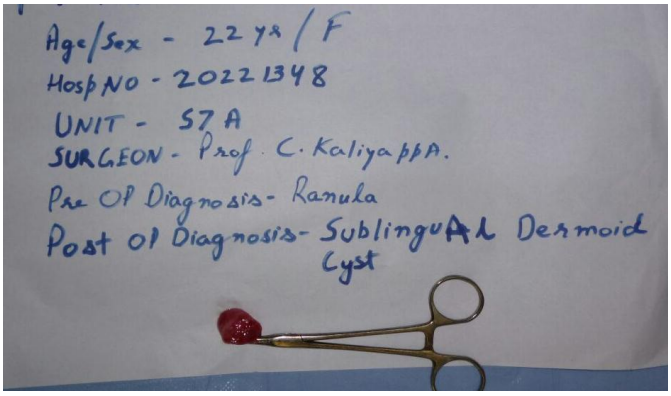

Fig 3: Gross appearance of the excised specimen.

Discussion

Embryologically the ventral bud develops into primitive foregut and tracheobronchial tree. Any malformation in this would cause a cystic lesion along the tracheobronchial tree or the foregut. Most frequently unilocular, they contain clear fluid or, less commonly, hemorrhagic secretions or air. They are lined by columnar ciliated epithelium, and their walls often contain cartilage and bronchial mucous glands. According to the structure of the cyst wall, foregut cysts can be divided into four types, these are bronchogenic cysts, esophageal duplication cysts, enteric duplication cysts and mixed cysts.

In 1911 Blackader first described bronchogenic cyst. The first surgical resection was performed in 1948 by Maier. The bronchogenic cyst is a remnant of the primitive foregut, secondary to trachea-bronchial malformations. Bronchogenic cyst was first described as a congenital pulmonary malformation by Bush in 2001 and that it represented from 14% to 22% of congenital pulmonary malformations and 10% of mediastinal malformations [5].

In addition, bronchogenic cysts can occur in the lungs, esophagus, stomach wall, paravertebral gutter, skin and subcutaneous area. The cysts can also move to atypical sites including neck, myocardium, pericardium, thymus, lung ligament, diaphragmatic and retroperitoneal region and abdomen.

Although some bronchogenic cysts are asymptomatic and are incidental findings upon radiography, most cysts are symptomatic—and complications are more common in symptomatic patients [6].

Clinical manifestations of bronchogenic cyst are associated with the cyst location, size and the oppression on its surrounding organs. If the cyst is small and does not oppress its surrounding organs as described in this patient, there may be no symptoms and can only be observed during physical examination [7]. When the cysts are large, there may be symptoms of cough, chest tightness, chest pain, difficulty in breathing or swallowing difficulties [with cough being the most common symptoms] due to its oppression on trachea, bronchus, pulmonary or esophageal [8]. A small number of patients have precordial murmur due to the oppression on pulmonary artery by the bulge of the cyst under the hilar pulmonary. Patients at younger age are more likely to have the compression symptoms because the trachea and bronchial tree are relatively soft and vulnerable to oppression. Some patients may have acute compression and fever symptoms because of the sudden increase of the cyst after bleeding or secondary infection within the cyst. A few patients have special manifestations, such as pleural effusion, even under the condition of intact cyst without infection or rupture. If the cyst penetrates into other adjacent organs, cavities, lung, trachea, esophagus and pleural cavity, symptoms of hemoptysis, hemorrhage or empyema can occur [2].

CT scans and radiographs are valuable diagnostic studies. Bronchogenic cysts appear as spherical or oval masses with smooth outlines and are usually unilocular and noncalcified [6]. Computed tomography is valuable in demonstrating the size and shape of the cyst and in determining its position in relation to other structures. The fluid in these cysts has an average CT density of 0 Hounsfield units [6].

Identification of the lesions and preoperative localization, and diagnosis rely mainly on imaging examinations. Chest X-ray examination is not helpful for the location of the lesion and diagnosis of the disease [1, 2, 5]. X-ray diagnosis is merely speculative. For those bronchogenic cysts located in the front of the bulge or behind the trachea, esophageal barium meal examination can observe local exogenous pressure trace in esophagus, but it is not easy to distinguish from esophageal cysts. CT scan is very helpful to determine the location of bronchogenic cyst and its relationship with the trachea and bronchus, as well as the compression of the cysts on the adjacent organs. CT scan is also helpful for the diagnosis of cyst bleeding and infection. Calcification may occur in approximately 10% of bronchogenic cysts. CT is very accurate for the diagnosis of cyst wall calcification. The CT value of the cysts can vary from a typical water density [0–20 Hu] to high density [80–90 Hu] according to the material composition of cyst fluid, different content of protein, calcium, and the presence of infection [3, 4, 9, 10].

Compared to CT, MRI is more sensitive for the diagnosis of bronchogenic cyst and more accurate for the lesion localization. MRI can more clearly show the different tissue components of the cyst. According to the literature, the preoperative CT diagnostic accuracy rate was 69.2%, while MRI diagnostic accuracy rate was 100% [7, 10].

Bronchogenic cyst also needs to be differentiated from lipomas, fibroma, neurogenic tumors, epithelioid hemangioendothelioma, and encapsulated fluid, since all these have similar imaging presentations [6].

Bronchogenic cysts are pathologically characterized by the ciliated columnar epithelial cells covering the inner surface of the cyst. The inner cyst wall may contain cartilage and smooth muscle component. Presence of typical respiratory epithelium covering the wall is a necessary condition for pathological diagnosis. In contrast, cartilage or muscle components within the cyst are not essential for diagnosis [11]. In the case reported here, the inner surface of cysts was covered with typical respiratory epithelium, however, cartilage or smooth muscle was not observed within the cyst wall. We speculated that this was possibly due to the deterioration of cartilage and smooth muscle during the development of cysts [6].

Surgery is the preferred treatment for adults with symptomatic foregut cysts. In contrast, it is still controversial if surgery should be applied for those asymptomatic cysts. Some studies propose dynamic examination for those patients. However, multiple studies have shown that foregut cysts [including bronchogenic cyst and esophageal cyst] in old patients may become malignant. Furthermore, with the

growth of bronchogenic cyst, most patients may have complications, which results in complicated disease conditions and surgical difficulties. Therefore, in principle, surgical resection should be applied as early as possible for the patients even without clinical symptoms [6, 7, 9, 11].

If the cysts are large and affect resection, puncture decompression can be applied. For those patients with difficulties in complete removal of the cyst, e.g., close adhesion between the mediastinal cyst wall and membranous trachea, some cyst wall can be left and treated with ablation using electrocautery [6, 7, 10].

Alternatively, the mucosal membrane can be stripped from the residual cyst wall followed by burning using iodine or anhydrous alcohol in order to prevent the cyst recurrence from the residual cyst wall or mucosal membrane [6].

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