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Case report

Clinico-pathological features of the intradiaphragmatic bronchogenic cysts: report of a case and review of the literature

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Key words

Bronchogenic cysts • Intradiaphragmatic localization

Summary

Bronchogenic cysts represent congenital malformations deriving from an abnormal development of the primitive foregut during embryogenesis. These lesions are rarely found and they are most frequently localized in the mediastinum, or in lung parenchyma. Intramuscular localization is extremely rare, especially within the diaphragm. We report a case of a 54 year old man showing a large lobulated cystic lesion in the left hemidiaphragm. Complete surgery was performed and histological diagnosis of intradiaphragmatic bronchogenic cyst was made during surgery and confirmed after a total section analysis. Moreover we reviewed the other cases published in the English literature, including clinical, surgical and pathological data.

Introduction

Congenital mediastinum cysts are malformations derived from a developmental anomaly of the foregut during embryogenesis, showing bronchogenic, esophageal, or gastrointestinal epithelium. Bronchogenic cysts are more frequent in children, but they have been reported also in adults, especially localized in the posterior zone of the mediastinum, along the tracheobronchial tree or within the pulmonary parenchyma. Occasionally they have been found in the skin, subcutaneous tissue, within the esophageal wall, or in the retroperitoneum [1 2]. The intradiaphragmatic location is a very exceptional finding. It should be noted that only few cases have been reported in the last two decades [3-5].
In our report, we expose the case of an intradiaphragmatic bronchogenic cyst in 54-year-old man, who presented chest pain and dyspnea.

Case report

A 54 year old man was referred to our hospital with persistent chest pain on the left side and dyspnea. The patient was never smoker, he had undergone to a previous knee ligament surgery, but no other clinical data of interest were reported. General blood tests displayed normal results. A chest X-ray was performed and presence of a cystic lesion was revealed in the left crus of the diaphragm (Fig. 1). The CT scan confirmed the presence of the lesion. The tumor was localized and removed, after selective intubation, by a left lateral thoracotomy on the 7th intercostal space.

Pathological findings

The resected cystic mass was sent to the Pathology Department. The mass size was 7 x 4 x 2 cm. It appeared well encapsulated with an associated small part of diaphragmatic muscle, and it contained seromucinous liquid.
Histologically, the cystic lesion was composed by a fibrovascular connective wall, containing seromucosus glands (Fig. 2E), and thin layers of smooth muscle and islands of cartilaginous tissue (Fig. 2D). The inner cyst surface was lined by ciliated pseudostratified or tall cylindrical epithelium (Fig. 2A, B, C). Dysplastic changes or evidence of malignancy were not found. We did not observe bone, skin, neural or other teratomatous struc-

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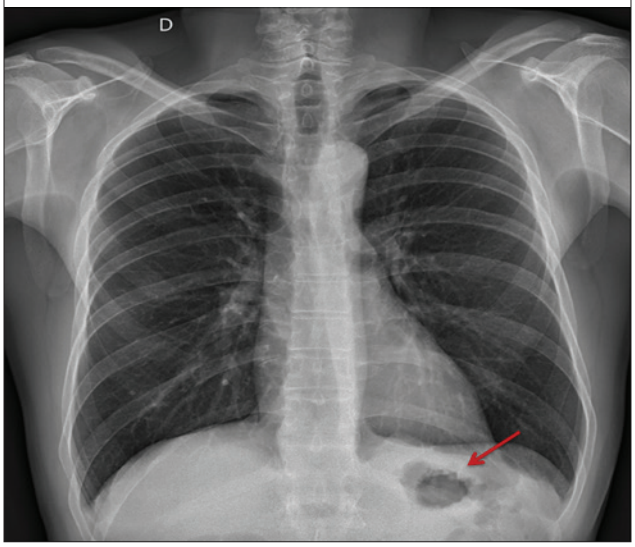
Fig. 1. Radiological image of the intradiaphragmatic cyst. Preoperative chest X-ray showing a cystic mass within the left crus of the diaphragm (red arrow).



tures or tissues. The lesion was finally diagnosed as intradiaphragmatic bronchogenic cyst.

Discussion

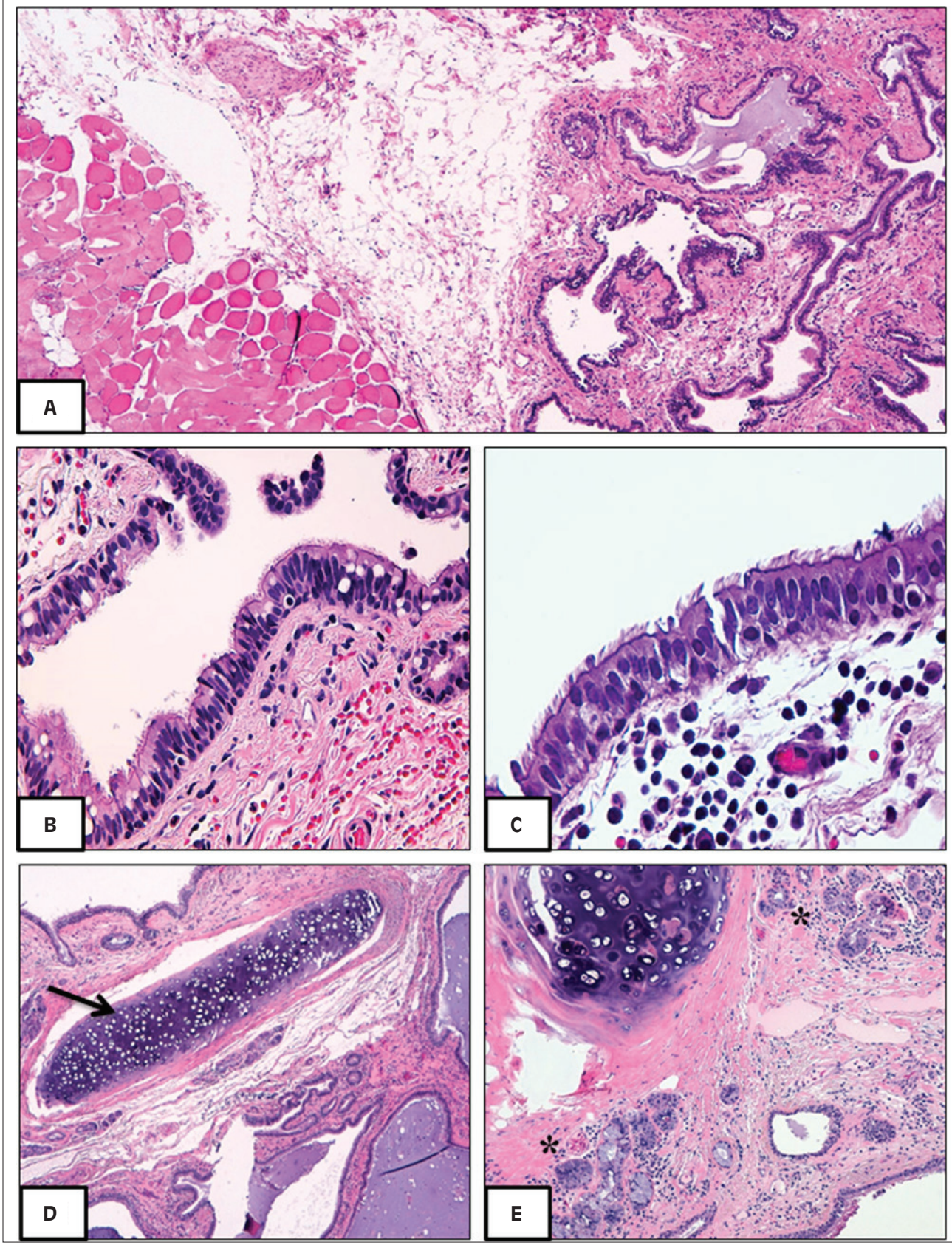
Bronchogenic cysts are rare congenital lesions resulting from abnormal budding of the primitive tracheobronchial tube, morphologically composed by bronchogenic, esophageal, or gastrointestinal epithelium [1 2]. Although apparently derived from alterations during embryogenesis, the pathogenesis of these lesions remains unclear. Basing on a case observed in the Pathology Department of our hospital, we realized an exhaustive review of the English literature. We found that 26 cases of intradiaphragmatic bronchogenic cyst were published from 1955 to 2017 and we analyzed clinical, radiological, surgical and morphological data. Table I summarizes the reported cases in English literature of intradiaphragmatic bronchogenic cysts.

Tab. I. Review of the cases of intradiaphragmatic bronchogenic cyst in English literature.

First author	Year	Journal (ref.)	Age (year)	Sex	Size (cm in diameter)	Side of diaphragm	Prevalent symptom	Surgical Approach
Kesseler et al.	1955	J Thorac Surg [14]	41	F	N/a	N/a	N/a	Thoracotomy
Buddington et al.	1957	NEJM [15]	62	M	6.5	Left	Asymptomatic	Thoracoabdominal
Felder et al.	1959	Am J Surg [16]	36	F	N/a	Right	Abdominal pain	Thoracotomy
Aaron et al.	1965	J Thorac Cardiovasc Surg [17]	21	F	N/a	N/a	N/a	Thoracotomy
Gourlay et al.	1966	Can J Surg [18]	41	M	N/a	N/a	N/a	Thoracotomy
Greenberg et al.	1974	JAMA [19]	26	M	20	Right	Abdominal pain	Laparotomy
Leone et al.	1985	RAYS [20]	N/a	N/a	N/a	N/a	N/a	N/a
Fischbach et al.	1994	Pediatr Radiol [21]	12	M	3	Right	Asymptomatic	N/a
Dagenais et al.	1995	Ann Thorac Surg [22]	51	F	4	Right	Abdominal pain	Thoracoabdominal
Rozenblit et al.	1998	Clin Radiol [8]	64	F	6	Left	Abdominal pain	Laparotomy
Hoang et al.	1999	Clin Exp Pathol [23]	51	M	5.5	Left	Abdominal pain	Laparotomy
Desrumaux et al.	2001	JBR-BTR [24]	50	M	N/a	N/a	N/a	N/a
Liou et al.	2001	J Formos Med Ass [25]	34	M	N/a	Left	Asymptomatic	Thoracotomy
Anile et al.	2006	Eur J Cardiothorac Surg [3]	38	F	N/a -	Left	Hiccup	Thoracotomy
Chang et al.	2006	J Laparoendosc Adv Surg Tech A [13]	74	F	1	Right	Asymptomatic	Thoracoscopy
Chang et al.	2006	J Laparoendosc Adv Surg Tech A [13]	54	F	5	Left	Cough	Thoracoscopy
Zügel et al.	2008	JSLS [12]	43	F	8	Right	Hiccup	Laparoscopy
Elemen et al.	2008	Pediatr Surg Int [11]	1,6	F	4	Right	Fever	Laparotomy
Kim et al.	2011	Korean J Thorac Cardiovasc Surg [29]	56	F	8	Right	Chest pain	Thoracotomy
Jiang et al.	2013	Ann Thorac Surg [4]	38	F	5	Left	Asymptomatic	Thoracotomy
Subramanian et al.	2013	WMJ [5]	13	M	4.8	Left	Back pain	Thoracoscopy
Herek et al.	2015	Ind J Surg [26]	42	M	N/a	N/a	Back pain	N/a
Mubang et al.	2015	J Cardiothorac Surg [27]	41	M	5.9	Left	Back pain	Thoracotomy
Chumakova et al.	2016	BMJ Case Rep [28]	34	M	11.8	Left	Chest pain	Laparotomy
Gao et al.	2017	Tumori [7]	22	M	12.7	Left	Chest pain	Laparotomy

M: Male; F: Female. n/a: data not available.

Fig. 2. Histological images of the cystic lesion.
The histological examination showed an intramuscular cystic formation (A) lined by ciliated pseudostratified or tall cylindrical epithelium with goblet cells (B, C). The cystic wall was formed by fibrovascular connective tissue, containing thin layers of smooth muscle, islands of cartilaginous tissue (D, arrow) and seromucosus glands (E, asterisks). Hematoxylin-Eosin stain; A: x20 magnification, B: x40 magnification, C: x100 magnification, D: x20 magnification, E: x40 magnification.



It has been described that these lesions are more common in female patients. We found that there is not a big difference between sexes: the patients were 12 females, 12 males, one unknown, with ages ranging from 19 months to 74 years old, with a mean age of 39.4 years old. Symptoms are frequently nonspecific and depending on the size and localization of the cysts. In 20% of cases the lesion is asymptomatic and an incidental finding during routine chest radiography. When present, symptoms varied from persistent hiccup or cough to back or chest pain or including manifestations due to compression or irritation of adjacent structures such as dyspnea, cough, dysphagia or recurrent infections [6 7].

Bronchogenic cysts are more frequently localized in the mediastinum, along the tracheobronchial tree or within the pulmonary parenchyma. However they have been occasionally found in the neck, skin, subcutaneous tissue, within the esophageal wall, pericardium, thymus or in the retroperitoneum [1 2]. In the computed tomography (CT) images, bronchogenic cysts appear as round, low-density masses, with an average size of 1.5-10 cm. However, obtaining radiologic diagnosis results is difficult because the density of the cyst could be heterogeneous on CT, depending on the substance contained in the cyst [8]. Therefore, the definitive diagnosis needs pathologic confirmation.

On gross examination, the tumor size ranged from 1 to 20 cm, six (24%) were multilocular, while three (12%) were unilocular, with the rest of cases not described. Grossly the cysts show a fibrotic wall containing or not glands, cartilage and smooth muscle. Histologically the inner surface is lined by respiratory epithelium, with or without squamous metaplasia [5 6]. Malignant transformation of bronchogenic cysts has been reported, but it is extremely rare [9 10]. None of the reviewed cases showed dysplastic changes or malignancy; however two cases denoted the presence of squamous metaplasia. The differential diagnosis of cystic lesions of the diaphragm include; hydatid, mesothelial or gastrointestinal duplication cysts, peripheral lung neoplasms or sequestration, diaphragmatic or neurogenic tumors, or esophageal diverticulum [11].

The management of the patients with intradiaphragmatic bronchogenic cysts requires a complete surgical excision. The removal of a mediastinum mass is possible with minimal invasive surgery, such as video-assisted thoracoscopy or laparoscopy [12 13]. Postoperative morbidity, mortality and complications are extremely rare. They can include infections, diaphragmatic paralysis or bronchopleural fistulas. All reviewed patients showed a very good follow-up.

Conclusions

Basing on a diagnosis of a case of intradiaphragmatic bronchogenic cyst in a 54 year old man observed in our hospital, we realized an exhaustive review of the English literature analyzing all clinical, radiological, surgical and morphological data. These rare lesions show a slight prevalence in females then in males, in all age groups and generally asymptomatic. Due to the non-specificity of the symptoms (when present) and the heterogeneity of the radiological images, the definitive diagnosis requires a complete surgical excision and the pathologic confirmation of respiratory epithelium accompanied by glands, cartilage and smooth muscle. These lesions show a very good prognosis and they should be considered in the differential diagnosis of uncommon lesions of the diaphragm.

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