Mediastinal Cystic Lesions; Experience of 77 Patients

Mediastinal Kistik Lezyonlar; 77 Hastalık Tecrübe

Mediastinal Kistik Lezyonlar / Mediastinal Cystic Lesions

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Özet

Amaç: Mediastenin kistik lezyonları nadirdir. Çoğu konjenital lezyonlar olup primer mediasten kitlelerinin % 20 ila % 30' unu oluştururlar. Primer mediastinal kistik lezyonları (PMKL) klinik, radyolojik ve patolojik özelliklerinin yanı sıra cerrahi yönetimin erken ve uzun dönem sonuçları açısından retrospektif olarak araştırdık. Gereç ve Yöntem: Ocak 1998 -Temmuz 2008 arasında kliniğimizde PMKL olan 77 hasta-47 kadın ve 30 erkek, 4-81yaşları arasında- tedavi edildi. Tüm hastalar yaş, cinsiyet, semptomlar, kist tipleri ve uygulanan ameliyat çeşidi açısından incelendi. Bulgular: Çalışmaya otuz erkek hasta (% 40), yaşları 4-81 yıl, ortalama yaş 39.2 yıl ile kırk yedi kadın hasta (%60), yaşları 16-65 yıl, yaş ortalaması 35.8 yıl alındı. Daha önceden benign kist tanısı konan hastaların patoloji slaytları tekrar incelendi ve yeni tanıları ile yeniden sınıflandırıldı. 31 bronkojenik kist (44 %; 19 kadın, 12 erkek), Onsekiz coelomic perikardiyal kist (% 24; 7 kadın, 11 erkek), beş mediastinal kist hidatik (% 6; 4 kadın, 1 erkek), beş enterogenous kist (6 %; 3 kadın, 2 erkek), sekiz timik kistler (% 10; 7 kadın, 1 erkek), iki kistik lenfanjiomatozis (% 4; 1 kadın, 1 erkek), beş teratojenik kist (% 6; 4 kadın, 1 erkek) ve üç plevral kist (2 kadın, 1 erkek) vardı. Başlıca semptomlar ağrı, ateş, nefes darlığı ve öksürükdü. Yirmi üç hasta (% 30) ise asemptomatikdi. Tüm hastalara cerrahi uygulandı. Hastanede kalma süresi ortalama 8 gündü. Hiçbir hastamızda ölüm olmadı. Tartışma: PMKL'nu olan hastaların çoğu kadındı. PMKL'lerin çoğu foregut lezyonlardır. Farklı lokalizasyon ve histolojik tiplere rağmen mediastinal kistlerin klinikleri benzerdi. Tüm PMKL olgularında cerrahi en iyi tedavi şansı sağlar.

Anahtar Kelimeler

Kist; Mediasten; Tedavi

Abstract

Aim: Cystic lesions of the mediastinum are rare. Most of them are congenital lesions and account for 20% to 30 % of all primary masses of the mediastinum. A retrospective study of primary mediastinal cystic lesions (PMCL) was conducted to review their clinical, radiological, and pathological features, as well as the early and long-term results of surgical management. Material and Method: From January 1998 through July 2008, 77 patients—47 females and 30 males, aged 4–81 years—with PMCL were treated in our department. All of the patients were analysed acording to the age, gender, symptoms, types of cysts and type of surgery. Results: There were thirty male patients (40 %), aged 4–81 years with a mean age of 39.2, and forty-seven female (60 %) patients, aged 16–65 with a mean age of 35.8. Some of the patients' pathology slides who were diagnosed as benign cysts before are re-examined then reclassified with the new diagnosis. There were thirty-one bronchogenic cysts (44 %; 19 female, 12 males), eighteen pericardial coelomic cysts (24 %; 7 female, 11 male), five mediastinal hydatid cysts (6 %; 4 female, 1 male), five enterogenous cysts (6 %; 3 female, 2 male), eight thymic cysts (10 %; 7 female, 1 male), two cystic lymphangiomatosis (4 %; 1 female, 1 male), five teratogenous cysts (6 %; 4 female, 1 male), and three pleural cysts (2 female, 1 male). The main symptoms were pain, fever, dyspnea, and coughing. Twenty-three patients (30 %) were asymptomatic. All of the patients underwent surgery. Mean stay in the hospital was 8 days. We did not have any deaths. Discussion: Most of the patients with PMCL were female. Most of the PMCL were foregut lesions. Despite varied location and histology, the clinical presentation of mediastinal cysts was similar. Surgery provides the best chance for cure in all cases of PMCL.

Keywords

Cyst; Mediastinum; Treatment

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Introduction

Cysts of the mediastinum are benign masses and important diagnostic group, representing 15 to 18 % of all primary mediastinal tumors [1;2].

Despite the frequent discovery of these cysts in infants and children, most of these cysts are not identified until the third or fourth decade of life. PMCL can be classified as foregut originated lesions (pulmonary sequestration, congenital cystic adenomatoid malformation, congenital lobar emphysema, bronchogenic, and esophageal duplication cysts), with mesothelial cell lesions (pleural, pericardial, and thoracic ductus cysts) and acquired infected lesions (teratogenic, thymic, and hydatid cyst) [2].

Our study examines the spectrum of mediastinal cysts with an emphasis on their clinical and pathological features.

Materials and methods

We operated on 77 patients who were admitted, investigated, and treated for PMCL, and were were clinically and radiologically identified at our hospital from January 1998 through July 2008. Our diagnostic work-up in all cases consisted of a detailed history, physical examination, and posteroanterior and lateral chest radiographs and thoracic computed tomography (CT) scan. Finally, 77 patients were included in this study and are analyzed.

Results

Forty-seven (60 %) of the patients were female, ranging in age from 16 to 65 years (mean age of 35.8 years) and thirty (40 %) of the patients were male, ranging in age from 4 to 81 years (mean age of 39.2 years). Of the forty-seven female patients, 19 had bronchogenic cysts (26 %), 7 had pericardial coelomic cysts (9 %), 4 had mediastinal hydatid cysts (5 %), 3 had enterogenous cysts (4 %), 7 had thymic cysts (9 %), 1 had a cystic lymphangiomatous (2 %), 4 had teratogenous cysts (2 %), and 2 had pleural cysts (1 %). Of the thirty male patients, 12 had bronchogenic cysts (17 %), 11 had pericardial coelomic cysts (16 %), 1 had a mediastinal hydatid cyst (1 %), 2 had enterogenous cysts (2 %), 1 had a thymic cyst (1 %), 1 had cystic lymphangiomatous (1 %), 1 had a teratogenous cyst (1 %), and 1 had a pleural cyst (1 %) (Table 1).

The diagnostic work-up consisted of a detailed history, physical examination, posteroanterior and lateral chest radiographs, thoracic CT scans, and thoracic ultrasonography in all the patients. In some patients, we performed thoracic magnetic resonance imaging.

The main symptoms were pain, dyspnea, and coughing. Some of the patients who had complicated lesions complained of fever, and some patients who had an enterogenous cyst complained of dyspnea. Twenty-three of the patients (30 %) were asymptomatic (accidental radiological findings in recruitment centers). Sixteen of the patients with bronchogenic cysts (50 %) had chest pain. Only two of the patients with pericardial coelomic cysts had hemopytsis (Table 2).

The surgical approach used to be mostly through a posterolateral thorocotomy and a median-sternotomy, especially to the thoracic inlet located cysts. Forty-five patients had a right thorocotomy, nine patients had a median-sternotomy, and twenty-three patients had a left thorocotomy (Table 2). Mean stay in the hospital was 8 days. We did not have any deaths.

Discussion

Mediastinal cysts are uncommon lesions in children and adults. The classification of the cysts is usually based on their etiology [2]. Histologically, they may be classified into congenital cysts,

Table 1. Types and number of the cysts per years and sex.

Type of cysts	<15 years	15-50 years	>50 years	Female	Male	Total
Bronchogenic cyst	1	17	13	19	12	31
Coelomic cyst	1	13	4	7	11	18
Hydatic cyst	-	3	2	4	1	5
Enterogenous cyst	2	3	-	3	2	5
Thymic cyst	3	4	1	7	1	8
Cystic lympangioma	-	1	1	1	1	2
Teratogeneous cyst	-	3	2	4	1	5
Pleural cyst	-	2	1	2	1	3
Total	8	45	24	47	30	77

Table 2. Symptoms of mediastinal cysts and operation form

	Bronchogenic cyst	Coelomic cyst	Cystic lympangioma	Teratogeneous cyst	Thymic cyst	Enterogenous cyst	Hydatic cyst	Pleural cyst
Asymptomatic	9	4	1	2	4	1	2	-
Cought	8	12	-	2	1	-	1	1
Chest pain	16	6	1	1	2	1	2	2
Dyspnea	4	6	-	1	1	1	-	-
Hemoptisis	-	2	-	-	-	-	-	-
Chronical fatigue	6	2	-	-	-	-	-	-
Dysphagia	-	2	-	-	-	1	-	-
Countinous crying, cyanosis	-	1	-	-	-	-	-	-
Palpitation	-	-	-	-	-	1	-	-
Right thoracotomy	20	15	1	2	-	3	2	2
Left Thoracotomy	11	3	1	3	-	2	2	1
Median sternotomy	-	-	-	-	8	-	1	-

such as foregut cysts, mesothelial cysts, and lymphatic cysts; acquired cysts, such as cystic teratomas, thymic cysts, and dermoid cysts; and an infectious group, such as hydatid cysts [3;4]. Thus, the cysts may be found either in the superior, anterior, middle, or posterior mediastinum.

Foregut cysts are categorized on the basis of their anomalous embryonic origin into bronchogenic, esophageal, gastric, congenital parenchymal, and neurenteric cysts. Such lesions are malformations produced during the stage of differentiation and embryological development of the primitive intestine [4;5]. In some publications, foregut cysts are reported as forming 50 % of all cases of PMCL. Bronchogenic cysts are seen more often in the foregut cysts group. Despite the variety of foregut cysts, the symptomatology is nearly identical in all cases and the main symptom is chest pain. Other symptoms like coughing, dyspnea, and dysphagia are all considered manifestations of compression or irritation of the major airways and esophagus by the cysts [3-7]. The bronchogenic foregut cyst results from sequestration of cells from the laryngotracheal groove during early embryologic development. It accounts 50–60 % of all mediastinal cysts, is usually found in adults, and is more common in men than in women [4]. In our series there were 31 bronchogenic cysts with a female predilection of 26 % to 17 %, which is a reverse of the literature (Figure 2-D). In accordance with the

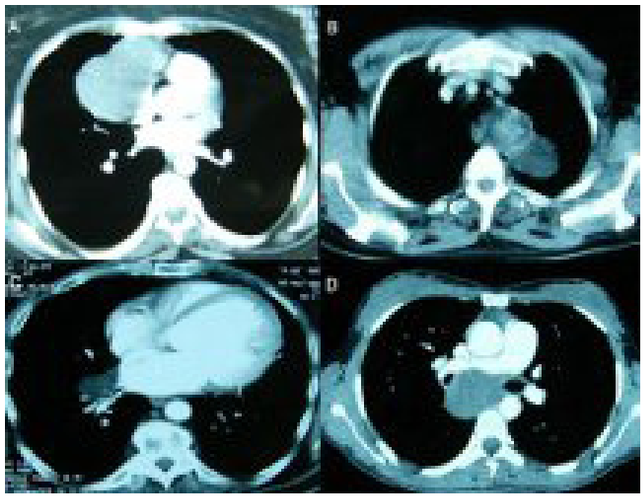


Figure 2. Cystic thymoma is showed on computed thorax tomograpy (A), Cystic teratoma is showed on computed thorax tomograpy (B), Pleural cyst is showed computed on thorax tomograpy (C), The broncogenic cyst is showed on computed thorax tomograpy (D).

literature, seventeen of our patients were in the 15–50 years age group and thirteen were older than 50 years; only one patient was younger than fifteen. All of our patients underwent surgical resection with a posterolateral thorocotomy.

Enteric cysts are rare congenital malformations consisting of a smooth muscle wall and gastrointestinal-like mucosal lining that develops from the primitive foregut. Enteric cysts of the pericardium are rare. Typically, such cysts are located at the right cardiophrenic angle and are adjacent, but not attached to the heart [8;9]. Up to 20 % of mediastinal foregut cysts lack specific histological features that would permit further classification, possibly because of prior hemorrhage of infection [10]. They are especially common in childhood [5;11]. In our series, two patients were younger than fifteen and the other three were 15–50 years. More of our patients were female than male. The main symptom was dyspnea in these patients. Three patients were asymptomatic and they were diagnosed during routine controls (Figure 3, Figure 4).

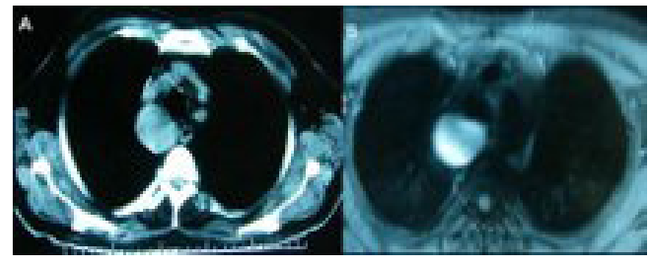


Figure 3. Enteric cyst is showed on computed thorax tomograpy (A), Enteric cyst is showed on magnetic resonance imaging (B).

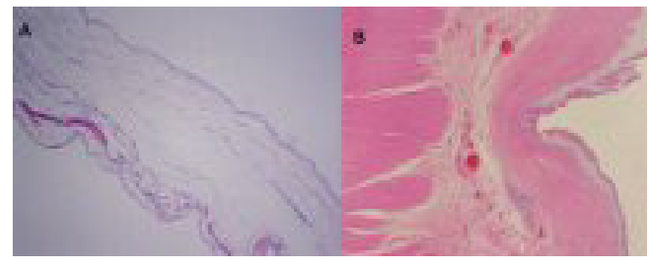


Figure 4. The pathological specimens are showed (A,B,C,D).
A: pericardial cyst, B: enteric cyst, C: thymic cyst, D: cystic teratoma

Among mediastinal cysts, pericardial cysts are also common lesions with frequencies ranging from 18–20 % [6]. In embryonic life, failure of one of the pericardial lacunes to merge results in subsequent cyst formation [3;12]. Pericardial cysts are usually formed by the parietal recess persisting during development as an aberrant recess fusion; therefore, the pericardial diverticulum is regarded as an incomplete form of a pericardial cyst in terms of its embryogenesis. They are unilocular with clear fluid and flat mesothelial cells set on a loose fibrous wall. They are usually asymptomatic and discovered on routine radiological examination in the fourth to fifth decades of life [4;6;13]. They can alter the cardiovascular hemodynamics and/or the pulmonary expansion, producing signs and symptoms that can mimic tricuspid stenosis, pulmonary stenosis, or constrictive pericarditis. The compression of mediastinal structures typically causes symptoms of dyspnea, thoracic pain, and coughing due to the unusually large size of the cyst [14]. The most usual radiographic appearance of pericardial cysts is a well-defined, smooth-walled round mass located along the right border of heart. Echocardiography is useful to establish a diagnosis, however, computed tomography and nuclear magnetic resonance are often needed to differentiate the pericardial cyst from the solid mediastinal mass [15;16]. In our series, there is concordance with the literature in that thirteen of our patients were in the 15–50 years age group and six of these patients were asymptomatic. There was a male predilection in our series. The cysts were generally located near the heart (Figure 1, Figure 4). A pleural (mesothelial) cyst is an uncommon developmental anomaly of the parietal pleura described by Clough and Beirne in 1955. It usually arises from the parietal pericardium, typically in the right cardiophrenic angle as described by Drash and Hyer in 1950. Less frequently, it is found on the surface of the

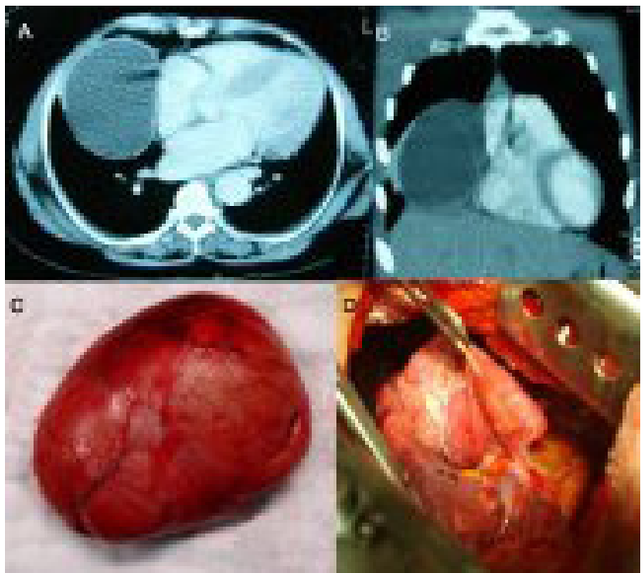


Figure 1. Pericardial cyst is showed computed thorax tomograpy (A,B), Macroskopic specimen of pericardial cyst (C), Intraoperative appearance of pericardial cyst (D).

diaphragm as described by Freedman and Simon in 1936. Jamplis et al. described a rare pleural cyst arising from the parietal pleural, covering of the upper mediastinum and paravertebral areas, and presenting radiographically as a mediastinal mass. Only two cases have been reported in which it arises from the posterior mediastinum (Figure 2-C).

Thymic cyst can be seen as congenital, neoplastic, or degenerative. Thymic cysts are classified as unilocular or multilocular [17]. Unilocular cysts are considered to be congenital, originating from the remnants of the thymopharyngeal duct and, hence, may more often be located in the neck [3;4]. In all of our cases, the thymic cysts were in the anterior mediastinal location. Thymic cysts are thin-walled, tense cysts filed with brown fluid. These cysts are generally asymptomatic, and symptoms develop due to enlargement of the cysts secondary to fluid accumulation. They can cause respiratory or cardiovascular compromise, such as coughing, dyspnea, dysphagia, congestive heart failure, or chest pain from irritation of the pleurae [17]. Chest radiography, computed tomography, and magnetic resonance imaging can be used to establish the diagnosis of a cystic chest mass and determine its origin and extension. Tumor markers such as sialylated Lewis X-i, carbohydrate antigen 19–9, tissue polypeptide antigen, HCG, carcinoembryonic antigen, AFP, and echinococcal antibodies should be obtained [17]. Four of our eight patients were asymptomatic and there was a female predilection of seven to one (Figure 2-A, Figure 4).

Hydatid disease is a clinical entity endemic to many sheep and cattle raising areas and is still a health problem in the world. hydatid cysts is the most common seen form of pulmonary cysts in the intrathoracic location. It is rarely seen in chest wall, mediastinal, pericardial, myocardial, fissural, and pleural space location. The extrapulmonary location of it in the thorax is rare. About 36 % of primary mediastinal cysts have been described in the thymic area [18]. The main symptom is dyspnea because of compression. There were 5 hydatid cysts in our series, and four of them were women. Two of them were asymptomatic.

Benign cystic teratomas of the mediastinum are rare lesions, accounting for approximately 8 % of all tumors of this region [6]. Patients are usually young adults and about 36–62 % of patients are found to be asymptomatic [6;18]. In our series, there were five patients-four female and one male; there was a female predilection. The tumors had a large cystic centre and 50 % of the lesions contained a ball of hair; ectodermal derivatives predominated (Figure 2-B, Figure4).

Lymphangiomas are composed of lymphatic sacs lined with endothelial cells. They are classified as simple lymphangiomas, cavernous lymphangiomas, or cystic lymphangiomas. Lymphangiomas are generally located in the anterior mediastinum, represent less than 1 % of cases, and may manifest as asymptomatic lesions in middle aged patients [19]. In our series, there were two patients, one of them was female and one was male, and both patients were older than fifty.

A meningocele is generally localized in the posterior mediastinum and it is often not alone. Although the etiology of intrathoracic meningocele is unknown, it is frequently associated with von Recklinghausen disease [2]. It may be defined as a saccular protrusion of the spinal meninges through an intervertebral foramen. It is formed after protrusion of the spinal menings as a sack from the intervertebral area. It causes sensitive deficits, atypical reflex, and headaches. We did not see a meningocele. It can be explained because our hospital is branch hospital.

Surgical intervention is the preferred line of management for all mediastinal cysts, regardless of size and clinical presentation, so as to prevent the development of various complications. Foregut cysts can be treated by aspirating the cyst's contents. Aspiration of these cysts with mediastinoscopy is reported in the literature. Therefore, we can ask which is better, aspiration or surgery? Some authors defend aspiration and claim that these lesions are benign and do not cause death in most patients, but we offer open surgery because we cannot estimate how these cysts will behave in the future. They can cause infection or mediastinal shift, and it is more difficult to treat these lesions after complications occur. These lesions must be treated with VATS, thorocotomy, and median sternotomy. In our series, all of our patients went under surgery and the cysts were removed completely. During their follow-up appointments, no problems were reported.

In conclusion, PMCL are benign lesions. Their morbidity is generally well but surgical technique is important. The options for treatment of foregut cysts or thymic cysts include mediastinoscopic aspiration in symptomatic patients, but these methods should be used only as temporary procedures in selected cases. We do not have clinical experience for mediastinoscopic aspiration. We want to share our last ten years' of cases with the literature.

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