Congenital mediastinal cysts: imaging findings*

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Abstract

Congenital mediastinal cysts are uncommon benign lesions generally caused by an abnormal embryological development of the foregut or coelomic cavity. They are expansive lesions, frequently asymptomatic that may manifest as a result from compression of adjacent structures. Bronchogenic, pericardial, enteric, thymic, esophageal duplication cysts and lymphangiomas are the main entities in this group of lesions. Congenital mediastinal cysts morphology is typical and imaging methods allow a correct diagnosis in a great number of cases. Surgical treatment is indicated only in cases of symptomatic lesions, considering that these lesions present no potential for malignant degeneration. The present study was aimed at commenting and illustrating the most relevant imaging findings of these lesions based on a retrospective review of ten cases collected in the records of the involved institutions.

Keywords: Congenital mediastinal cysts; Cystic neoplasms; Cysts.

INTRODUCTION

Congenital mediastinal cysts are benign lesions caused by abnormal embryological development of the anterior intestine and/or coelomic cavity[1]. These lesions represent 10% of the mediastinal masses and include, among their main entities, bronchogenic, pericardial, enteric, thymic, esophageal duplication cysts and lymphangiomas, generally asymptomatic although sometimes may present as a medical emergency, particularly because of adjacent organs compression.

Imaging methods, especially computed tomography (CT) capable of evaluating the density of such structures, play a critical role in the diagnostic elucidation and therapy planning if necessary. Other parameters such as localization and change in the lesions aspect according to the different patient positionings (for example, decubitus and orthostatic position) also can be relevant in the differential diagnosis, especially in the interpretation of radiographic images of the chest.

Therefore, radiologist’s familiarity with this theme is crucial, considering that most of times, their reports are the ending point of the diagnostic investigation.

Ten cases were selected from the records of the institution where the present study was developed in the period between 1999 and 2006, and six of these cases were confirmed by surgical and histopathological findings. The diagnoses of the other four cases were based on imaging findings and on the progression along the follow-up.

Among the cases included in this study, three pericardial cysts, three bronchogenic cysts, one esophageal duplication cyst, one enteric cyst, one thymic cyst, and one lymphangioma were diagnosed. No prevalence of these lesions in male or female individuals was observed, and the mean patients’ age at the moment of the diagnosis was 40 years, ranging from 5 to 70 years.

DISCUSSION

At CT, congenital mediastinal cysts are characterized as expansile, internally hypodense and homogeneous lesions with well-defined thin walls. Although these lesions are generally Noncontrast uptaking (Figure 1), capsular enhancement may be...
observed in a minority of cases and also in
the evaluation by magnetic resonance im-
aging (MRI)\(^2\).

The initial evaluation by chest radiog-
raphy is a useful diagnostic tool, demon-
strating a radiodense, expansile lesion with
variable localization whose aspect can
change according to the different patient
presentations (for example, dorsal decubi-
tus and orthostatic position). Because of
the absence of ionizing radiation, ultrasono-
graphy (US) is more appropriate for the
pediatric group, generally demonstrating
an anechoic, homogeneous, well-defined
cystic lesion confined either to a single or
multiple mediastinal compartments.

Clinical manifestations, when present,
result principally from extrinsic compres-
sion of adjacent structures such as the tra-
cheobronchial tree (dyspnea, stridor) and
esophagus (dysphagia). Other possible present-
tations result from rupture, infection
(see Figure 5), and hemorrhage\(^3,4\). In the
case of hemorrhage, CT demonstrates an
increase in size and density of the cyst,
complicating the differential diagnosis
with solid lesions\(^2\). So, it is essential that
the mass density is measured before and
after intravenous contrast injection, since
a density of > 10 Hounsfield units is con-
sidered as significant, indicating the pres-
ence of a solid lesion.

MRI can also be useful in this differen-
tiation of cystic lesions because of the high
signal intensity observed on T2-weighted
sequences (see Figure 7). Among the men-
tioned methods, CT stands out by the high
sensitivity and relative availability.

The proper determination of the specific
nature of the different types of congenital
mediastinal cystic masses is not always
feasible, and in this case these lesions are
otherwise classified as foregut cysts.

In the present paper, the main types of
congenital mediastinal cystic lesions are
separately described.

**Pericardial cysts**

Pericardial cysts results from anomalies
in the formation of coelomic cavities. These
lesions are invariably attached to the
pericardial leaflet, although communica-
tion with the pericardial cavity is observed
in only few cases\(^5\). Classically, pericardial
cysts arise adjacent to the right anterior
cardiophrenic angle, but less frequently can
also be seen at other sites such as the su-
perior pericardial recess\(^5\) (Figure 2). Occa-
sionally, the cysts may demonstrate rect-
tilinear contour and atypical shapes (for
example, triangular), probably because of
parietal adhesion. Among the pericardial
cysts described in the present study two
were found in the middle mediastinum and
one in the anterior mediastinum. Figure 3
is an example of the classical CT imaging
finding of this entity, demonstrating an
expansile, hypodense lesion adjacent to the
right anterior cardiophrenic angle. The blurring of the cardiac silhouette on the postero-anterior chest radiograph indicates the anterior positioning of this lesion (silhouette signal).

**Bronchogenic cysts**

Bronchogenic cysts originate from abnormal foregut protrusion in the period between the 26th and 40th days of pregnancy\(^1\). From the histopathological point of view, these lesions are lined with a ciliated columnar epithelium with a cartilaginous component and, less frequently, bronchial glands. Most frequently, these lesions are found in the middle mediastinum, near the tracheal carina, and less frequently in any part of the mediastinum and chest such as lungs, pleura and diaphragm\(^3,6\). Parietal calcifications can occasionally be observed (Figure 4).

The present casuistic is in contrast to the classical description of this entity, demonstrating one case with intrapulmonary localization (Figure 5) and the other two in the upper mediastinal region (right paratracheal and retrotracheal — respectively Figures 5 and 6).

**Esophageal duplication cyst**

Esophageal duplication cysts are classified into three groups according to their histopathological characteristics\(^1\); in the first group, a lesion adjacent to the esophageal wall is observed; the second group comprises the rarest presentation character-
ized by an extrinsic lesion communicating with the esophageal body; and the third group includes intramural, extramucosal lesions. These lesions are most frequently found in the posterior mediastinum, near the thoracoabdominal transition (Figure 7). Occasionally, gastric mucosa may be observed within these lesions, resulting in peptic complications such as ulcer and perforation\(^7,8\).

**Enteric cysts**

Enteric cysts are uncommon lesions, typically localized in the posterior mediastinum without a communication with the esophagus. The association of these lesions with vertebral abnormalities, particularly hemivertebra, is well documented\(^1\). Presence of gastric mucosa with the possible complications may also be observed. Figure 8 represents the classical description of these lesions, with posterior mediastinal localization and association with hemivertebra.

**Thymic cysts**

Thymic cysts represent 3% of anterior mediastinal lesions and can be congenital or acquired. The first group are similar to the other congenital mediastinal cysts, with regular contour and homogeneous and hypodense contents. Acquired lesions are frequently associated with inflammatory/infectious processes (HIV, lupus and Sjögren’s disease, among others) and neoplastic processes (such as non-Hodgkin lymphoma and, less frequently, thymoma, thymic carcinoma and mediastinal seminoma) presenting like heterogeneous, multilocular formations with intermingled septations. The differentiation between congenital and acquired lesions is important, considering that acquired lesions require histopathological study to rule out the association with neoplasms\(^9\). In these cases, the utilization of intravenous contrast agent is essential for demonstrating intralesional septations in acquired thymic cysts otherwise indistinguishable on non-contrast enhanced studies\(^10\). The present study demonstrates an anterior mediastinal well-defined, homogeneous, hypodense lesion compatible with congenital thymic cyst in an asymptomatic elder patient (Figure 9).

**Lymphangiomas**

Lymphangiomas are congenital benign lesions characterized by focal proliferation of lymphatic tissue with a multicystic pattern. These lesions are histologically classified according to the depth and size of lymphatic vessels involved, as follows:
Simple (capillary), cavernous and cystic (hygroma), this later being the most frequently found \(^1\). Axillas and neck are most commonly affected. Primary thoracic lesions correspond to only 1% of cases \(^2\). With a predominantly cystic composition, these lesions may present with septa and, less frequently, with a solid component. Calcifications are rarely observed. Generally, because of their soft consistency, these lesions tend to develop without compressing adjacent structures. The lymphangioma described in the present study (Figure 10) is characterized by a bulky, cystic, homogeneous lesion in the posterior mediastinum, involving aorta and esophagus, and extending to the pulmonary fields. Despite the size of this lesion, there is no sign of a significant luminal reduction of the descending aorta. Imaging findings compatible with cystic hygroma. One-year follow-up has demonstrated a significant involution of this lesion.
num, involving the descending aorta and the esophagus, with no sign of significant compression of these structures. One-year follow-up demonstrated a marked involution of this lesion.

**CONCLUSION**

Familiarity with the localization and with imaging findings of congenital mediastinal cysts is essential, considering that, most of times, the definite diagnosis is based on the interpretation of imaging studies to avoid unnecessary invasive procedures.

**REFERENCES**