



A thoracic duct cyst in 10-year-old boy: The youngest case report and review of the literature



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ARTICLE INFO

Article history:

Received 26 December 2015

Received in revised form

11 January 2016

Accepted 16 January 2016

Key words:

Thoracic duct cyst

Mediastinal cyst

Thoracic surgery

Video-assisted thoracoscopic surgery

ABSTRACT

We report the youngest case with a thoracic duct cyst located just above the diaphragm that was treated with thoracoscopic resection. The cyst was observed with serial ultrasonographic examinations during his prenatal period. No interval evidence of a deleterious effect to the fetus by this cyst was observed and born by normal vaginal delivery at term. Although the boy remained clinically asymptomatic at the age of ten, surgical excision for definitive diagnosis of the cyst was undertaken. The mediastinal cyst contained milky-white chylous fluid. Pathologically, the wall was similar in structure to the thoracic duct. The mediastinal thoracic duct cyst is an extremely rare cystic lesion. The etiology may be related to a congenital or degenerative weakness in the wall of the thoracic duct. They are generally asymptomatic but may sometimes cause pressure effects on adjacent structures. Surgical resection is the treatment of choice. The thoracoscopic resection is especially advisable for the patient with a small, asymptomatic cyst. Chylothorax is the most common postoperative complication. To avoid chylothorax after the resection of a mediastinal thoracic duct cyst, identification of the communication of the cyst with thoracic duct and ligation of its inferior pedicle without fail is necessary.

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The thoracic duct is the largest lymphatic vessel of the lymphatic system. The vessel originates from the cisterna chyli in the abdomen, traverses the diaphragm at the aortic aperture and penetrates through the posterior and superior mediastinum. Thoracic duct cysts of the mediastinum are extremely rare. The etiology is related to a congenital weakness and degenerative change in the wall of the thoracic duct, which result in aneurysm-like dilatation and subsequent cyst formation [1].

Herein, we report the first case of a child with a thoracic duct cyst located just above the diaphragm that was treated thoracoscopically.

1. Case report

The patient was 10-year-old boy who had been observed in our outpatient clinic with a history of congenital cystic mass at the supra-diaphragmatic mediastinum. The cyst was observed with serial ultrasonographic examinations during his prenatal period. No

interval evidence of a deleterious effect to the fetus by this cyst was observed and he was born by normal vaginal delivery at term.

Postnatal investigations included a computed tomography (CT) scan of the chest at 3 months of age, confirming the ultrasonographic findings of a posterior mediastinal cyst. It demonstrated a 4 cm, non-enhancing, homogeneous, hypodense cystic lesion in the posterior mediastinum that covered the anterior vertebral body and displacing the esophagus and descending aorta anteriorly (Fig. 1A). There was no enhancement with intravenous contrast. The differential diagnosis was that of a cystic lymphangioma. Preoperative CT images indicated that the size of the posterior mediastinal cyst was unchanged but showed no displacing the esophagus and descending aorta (Fig. 1B and C).

Laboratory findings and a chest X-ray showed no abnormalities. Gastro-endoscopy showed no mucosal abnormality to indicate a fistula or communicating cyst with a submucosal mass in the lower thoracic esophagus. Endoscopic ultrasonography showed a normal esophageal wall and a cystic mass next to the esophagus (Fig. 2).

Although the boy remained clinically asymptomatic, surgical excision for definitive diagnosis of the cyst was undertaken. Thoracoscopic resection of the mass was performed under general anesthesia in the left lateral decubitus position under single lung

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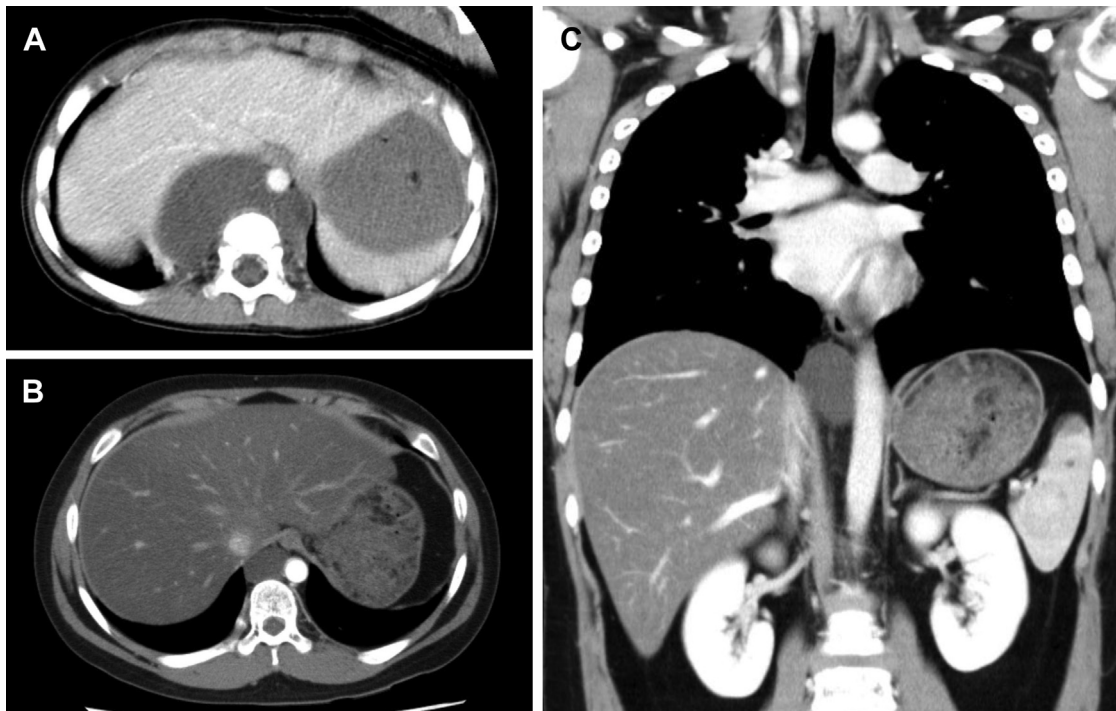


Fig. 1. (A) Computed tomography (CT) scan at 3 months of age shows a posterior mediastinal, non-enhancing, homogeneous, hypodense cyst covered the anterior vertebral body and it displaces the esophagus and descending aorta anteriorly. (B) Preoperative CT images showing the mediastinal mass next to the descending aorta. Axial view. (C) Coronal view.

ventilation on the left side. The lesion was a sessile cyst-like mass fixed to the posterior mediastinum. During dissection, the cyst wall was opened and a milky-white chylous fluid was observed. We dissected the cyst from the surrounding structures and the cystic mass was successfully removed. The thoracic duct was clipped and ligated using a pre-tie knot on the caudal side of the cyst to ensure closure (Fig. 3). The patient was sent to the general ward without any problems or hemodynamic instability. Postoperatively, the patient was well without any complications such as hemothorax or

chylothorax. The chest tube was removed on the 4th postoperative day with the confirmations that there are no symptoms and no chylous fluid after adequate oral intake and he was discharged two days later.

Microscopically, the inner surface of the cyst was sloughed off because of denaturation, and no endothelia had positive for D2-40; the wall was composed of connective tissue, smooth muscle cells, and elastic fibers. Histologically, the cyst wall was similar in structure to the thoracic duct.

On follow-up examination 5 months postoperatively, he was noted to be in excellent health with a normal chest roentgenogram and no limitations in his activities.

2. Discussion

Carbone [2] described the first mediastinal thoracic duct cyst, which was discovered during an autopsy examination. Cysts of the thoracic duct are uncommon disease described rarely in medical literature since the first case treated surgically by Emerson in 1950 [3]. In Mattila's report published in 1999 [4], the number of cases was 30 (19 mediastinal, 10 cervical, and 1 abdominal). As a result of the additional review up to today, we found additional 33 cases with mediastinal thoracic duct cysts reported previously (Table 1). The ages of the patients previously reported ranged from 17 to 86 years. To our knowledge, this is the youngest case to ever have been described.

Etiology of the thoracic duct cyst is uncertain and related to developmental defect in the wall of the thoracic duct or degenerative and lymphangiomatous changes from infection and inflammation [19] and they may arise at any level. With smaller mediastinal cysts, patients may be asymptomatic. Since the mediastinum contains a large amount of loose areolar tissue, enlargement of any single structure by a benign process will usually lead to displacement of other structures without significant compression [20]. Thoracic duct cyst may become apparent with manifestations

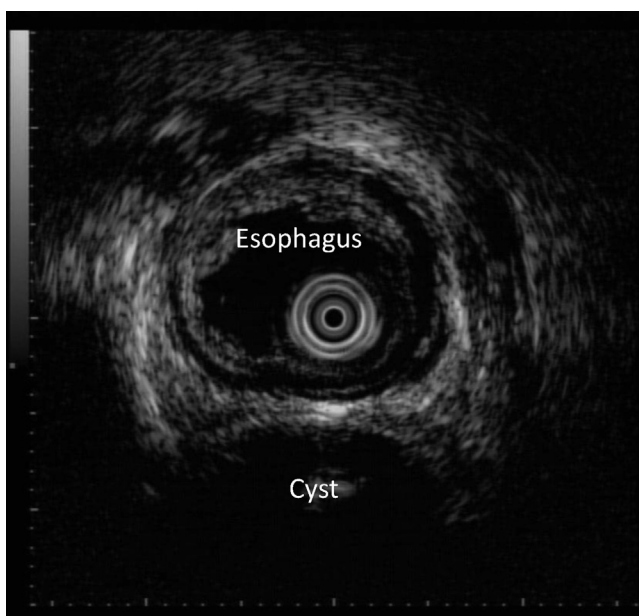


Fig. 2. Endoscopic ultrasonography showed a normal esophageal wall and a cystic mass next to the esophagus.

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