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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

Mediastinal lymphangioma in an adult with a tracheal bronchus

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SUMMARY

Introduction Lymphangiomas, also known as cystic hygromas or cystic lymphangiomas, are cystic abnormalities of the lymph vessels and they are rare benign tumors. Tracheal bronchus (*Bronchus suis* or "pig bronchus") is a very rare congenital anomaly.
The aim of this work is to present a very rare case of a lymphangioma with tracheal bronchus.

Case outline The article presents the rare case of a 35-year-old otherwise healthy man, who was admitted to our thoracic surgery department with a mediastinal tumor. On performing bronchoscopy a tracheal bronchus was found. A thoracic CT scan revealed a well-circumscribed mass in the superior and anterior mediastinum measuring 37 x 39 x 59 mm. First a Carlens mediastinoscopy, and then a right parasternal Chamberlain mediastinotomy were performed. The final pathological diagnosis of lymphangioma was made. In this case, surgery was not performed because the patient was asymptomatic and the tumor did not grow larger during follow-up.

Conclusion The lymphangioma of the mediastinum in an adult is a rare and benign condition with a good prognosis, but it should be considered in a differential diagnosis of mediastinal tumors. We recommend only a minimally invasive diagnostic approach (parasternal mediastinotomy) when the patient is asymptomatic.

Keywords: mediastinum, lymphangioma; tracheal bronchus; mediastinoscopy; parasternal mediastinotomy

INTRODUCTION

Lymphangiomas, also known as cystic hygromas or cystic lymphangiomas, are cystic abnormalities of the lymph vessels [1, 2]. They are rare benign tumors [1]. Nearly 90% of them occur in children up to the age of two years [3]. They appear mainly in the neck (75%) and axillary regions (20%) [4]. Other locations of lymphangiomas are mesenteric, retroperitoneal, orbital, pancreatic, and mediastinal (1%) [5]. They are predominantly congenital or sometimes acquired due to chronic lymphatic obstruction (e.g. infection, radiation). A malignant transformation of a lymphangioma has not been observed. Approximately 200 cases of lymphangiomas in adults have so far been described in literature [6].

Tracheal bronchus (*Bronchus suis* or "pig bronchus," and "tracheal diverticulum" as a variant) is a very rare congenital anomaly with a frequency of about 0.5% of pediatric bronchoscopy procedures [7].

The aim of this work is to present a very rare case of lymphangioma with tracheal bronchus.

CASE REPORT

A 35-year-old otherwise healthy man (A.S.), a cigarette smoker (15 packs per year) was admitted to our thoracic surgery department with symptoms of chronic fatigue and elevated (subfebrile) body temperature, which had lasted for three weeks. His past medical history was unremarkable, except for pulmonary tuberculosis in his adolescence and arterial hypertension. On admission the patient was in good general health. Physical examination revealed normal findings. Laboratory studies showed normal blood morphology and urinalysis. On performing bronchoscopy, a tracheal bronchus (a congenital anomaly) was found, starting from the right lateral wall of the trachea at a distance of less than 2 cm from the carina. A thoracic computed tomography (CT) scan revealed a well-circumscribed mass located in the superior and anterior mediastinum, measuring 37 × 39 × 59 mm in cross section (Figure 1), and displacing the superior vena cava and the right brachiocephalic vein. On October 30, 2012, a Carlens mediastinoscopy

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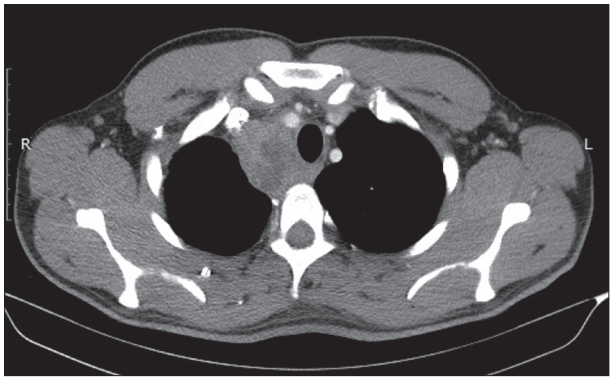

Figure 1. Contrast-enhanced axial CT scan of the thorax showing a well-circumscribed mass located in the superior and anterior mediastinum, measuring 37 × 39 × 59 mm, encasing the superior vena cava and the right brachiocephalic vein

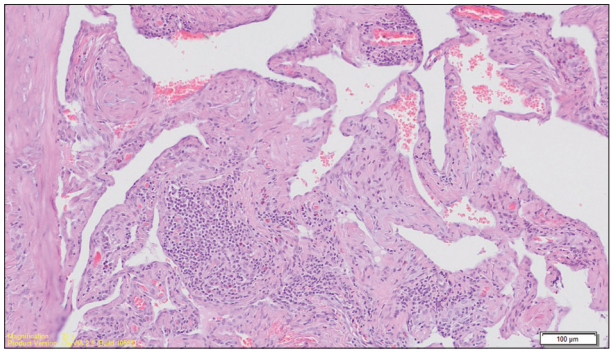

Figure 2. Lymphangioma; there are numerous, irregular vessels composed of endothelial and smooth muscle cells with lymphoid tissue between the vessels; erythrocytes within the lumen of some vessels might be misleading (H&E, ×70)

was performed under general anesthesia. The mediastinal tumor was biopsied with forceps and straw-colored serous fluid of 50 ml in volume was aspirated. The postoperative recovery was uneventful. The pathological examination revealed only necrotic masses, deposits of hemosiderin as well as non-specific fibrotic and/or inflammatory granulation of the tissue. The histopathology report stated that the finding was an organizing hematoma. The acid-fast bacteria culture of the sample was negative. A positron emission tomography scan was performed and confirmed the presence of the soft-tissue tumor in the mediastinum (maximum standardized uptake value of 4.7), which was characteristic of a low-grade proliferation process. Therefore, the decision to perform a Chamberlain parasternal mediastinotomy was taken, and this was carried out on February 14, 2013. During this procedure the right second costal cartilage was removed and the tumor of the anterior mediastinum was sampled. Postoperative recovery was uneventful. The patient was discharged on the first postoperative day. Histopathological examination revealed lymphangioma. H&E stain showed numerous, irregular vessels composed of endothelial and smooth muscle cells with lymphoid tissue between the vessels (Figure 2). Immunohistochemistry showed a positive reaction with the monoclonal antibody D2-40 for podoplanin (as a marker for the presence of a lymphatic endothelium), CD31 and

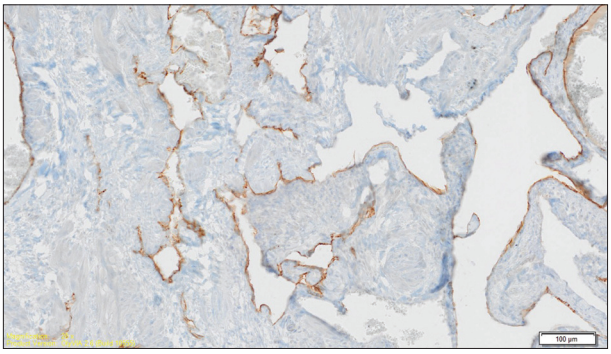

Figure 3. Positive immunohistochemical staining (brown color) with the D2-40 monoclonal antibody recognizing podoplanin, which is the membrane mucoprotein of lymphatic endothelium (×70)

CD34 (indicating the endothelial origin of tissue) (Figure 3). But the reaction to pancytokeratin AE1/AE3 was negative. There were no changes in the patient's condition in follow-up visits.

DISCUSSION

Lymphangiomas are benign tumors of mesodermal origin [8, 9]. They are divided into the following types: cystic (predominant in the thorax), cavernous, or mixed type [1, 10]. Thoracic lymphangiomas remain asymptomatic for many years as they grow slowly, and they are usually discovered incidentally. In this case, the patient was presented with only mild symptoms (chronic fatigue and slightly elevated body temperature), which soon subsided. Only large tumors may cause symptoms by compressing the adjacent anatomical structures, i.e. the tracheo-bronchial tree, the esophagus, or the superior vena cava [9]. Lymphangiomas are found more often in the superior and anterior mediastinum [2, 11, 12]. They account for 0.7–4.5% of all mediastinal masses [11]. The differential diagnosis includes a bronchogenic cyst, a pericardial cyst, a cystic thymoma, a cystic teratoma, a lymphoma, a goiter, a hematoma, or an aneurysm of the brachiocephalic trunk [5, 10, 11, 12]. Thoracic CT scan is helpful in determining the extent of the disease. The CT feature of lymphangioma is a cystic lesion with well-defined margins and without calcifications, which envelopes the thoracic structures [10, 12]. A histological examination with immunocytochemical staining for CD31 confirms the final diagnosis [11]. Some authors recommend complete surgical excision, usually via the right lateral thoracotomy [2, 4, 5, 6, 8, 10]. Others, for example Conte et al. [3], or Gorska et al. [1], advocate a conservative approach (watchful waiting) if the patient is asymptomatic. In this case, the non-invasive option was chosen as upfront surgery is not sufficiently evidence-based in literature. Lymphangioma of the mediastinum in an adult is a rare and benign condition with a good prognosis. It should be considered in a differential diagnosis of mediastinal tumors. We recommend only a minimally invasive diagnostic approach (parasternal mediastinotomy) when the patient is asymptomatic.

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Лимфангиом медијастинума код одраслог болесника удружен са трахеалним бронхом

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САЖЕТАК

Увод Лимфангиоми, познати и као цистични лимфангиоми или хигроми, јесу аномалије лимфних судова класификоване и као бенигни тумори. Трахеални бронх је врло ретка урођена аномалија.

Циљ овог рада је приказ врло ретког случаја лимфангиома са трахеалним бронхом.

Приказ болесника Приказан је мушкарац, 35 година стар, иначе здрав, примљен на одељење грудне хирургије због медијастиналног тумора. На бронхоскопији је нађен трахеални бронх. Компјутеризована томографија груди указала је на масу димензија 37 × 39 × 59 *mm* у предњем горњем медијастинуму. Урађена је прво Карленсова медијастиноскопија, а затим и Чемберленова десна парастернална медиастинотомија, а патохистолошки налаз потврдио је дијагнозу лимфангиома. Хируршко лечење није предузето јер у посматраном периоду медијастинална маса није расла, а болесник је био без симптома.

Закључак Лимфангиоми медијастинума код одраслих су ретки и доброћудни са добром прогнозом, али је неопходна јаснија диференцијална дијагноза од других медијастиналних тумора. Код асимптоматских болесника препоручујемо минимално инвазивни дијагностичи приступ (парастернална медијастинотомија).

Кључне речи: медијастинум, лимфангиом; трахеални бронх; медијастиноскопија; парастернална медијастинотомија