Dyspnea as a Clinical Manifestation in Primary Retroperitoneal Teratoma

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The retroperitoneum is an unusual location for a teratoma in adults. We report a case of retroperitoneal teratoma with clinical manifestation of dyspnea and chest discomfort. Chest radiograph showed elevated left hemidiaphragm. Ultrasonography (US) showed a hypoechoic tumor in the left subphrenic area. Computed tomography (CT) revealed a hypodense tumor in the left subphrenic area. Endoscopic ultrasound-guided fine-needle aspiration of the tumor suggested a diagnosis of liposarcoma. Magnetic resonance imaging (MRI) showed a tumor in the left subphrenic area that was isointense to the spleen on T1-weighted images and hyperintense on T2-weighted images. No typical imaging characters of teratoma such as fat-fluid level, calcification, hair, or Rokitansky bodies were depicted on US, CT, or MRI in our case. The thoracic symptoms completely disappeared after surgical removal of the tumor.

Key words: Retroperitoneal neoplasm; Teratoma; Magnetic resonance imaging

CASE REPORT

A 31-year-old woman was referred to our cardiovascular outpatient clinic because of dyspnea and chest discomfort. She had visited a local hospital several days previously, and valvular heart disease was diagnosed. She had a history of syphilis that was treated. On physical examination, no significant abnormal findings were found. A posteroanterior chest radiograph showed elevation of the left hemidiaphragm. Echocardiography showed a thickened mitral valve and normal ejection fraction of the left...
ventricle. An abdominal US showed a hypoechoic tumor in the left upper abdomen that measured 5.6 x 7.3 cm (Fig. 1). CT demonstrated a left subphrenic hypodense tumor measuring 5.8 x 8.2 cm posterior to the stomach and spleen (Fig. 2). MRI using a 1.5 T scanner in various scanning planes showed a tumor measuring 7.8 x 6.0 x 5.2 cm in the left upper retroperitoneum (Figs. 3-4). The tumor was of intermediate signal intensity, and was isointense to the spleen on T1-weighted images (Fig. 3), and hyperintense on true FISP (fast imaging with steady-state procession) images (Fig. 4).

Endoscopic ultrasound-guided fine-needle aspiration of this tumor was performed. The cytological findings of clear cells, cytoplasmic nuclear vesicles, and displaced dense nuclei suggested a liposarcoma. The serum carcinoembryonic antigen (CEA) was 5.7 ng/ml (normally less than 5), and CA-19-9 was 147 ng/dl (normally less than 37).

An exploratory laparotomy disclosed a well-encapsulated tumor in the left upper retroperitoneum without adhesion to adjacent structures. The tumor measured 8 x 6 x 5 cm. Intraoperative ultrasonography revealed the cystic nature of the tumor. The tumor was completely excised without rupture or spillage of the tumor content. On gross examination, it was yellow and corrugated in its inner surface, and no hair or bony fragments were found within this tumor. Microscopically, the wall of the cyst contained mature respiratory epithelium with cystic necrosis. There were neither immature elements nor malignant cells. The final pathologic diagnosis was mature cystic teratoma. The dyspnea and chest discomfort completely disappeared, and the serum CA-19-9 level decreased to 21 ng/dl after surgery. The patient has been followed up for 10 months, and the postoperative course was uneventful.

**DISCUSSION**

Primary retroperitoneal neoplasms comprise a diverse group of tumors. Teratomas constitute 1.2% to 10% of primary retroperitoneal neoplasms in various series [1, 4]. The retroperitoneum is a rare site for mature teratomas. Common locations for teratomas are the ovary, testis, and the anterior mediastinum in decreasing order of frequency [5, 6]. Previous studies showed a predominance of female patients by a ratio of 3.4 to 1, and a bimodal curve in age distribution at the time of diagnosis. Teratomas in around 52% of patients occur in a first peak in the first 6 months of life, and 26% of cases are found in a second peak in the first decade [2]. Fewer than 10%-20% of patients with retroperitoneal teratoma are older than 30 years of age [3].

The size of retroperitoneal teratomas varies, with the largest tumor weighting 36 kg [5]. In
adults, retroperitoneal teratomas are usually symptomatic before being diagnosed. The most common clinical manifestations include abdominal pain, abdominal distension, back pain, and edema of the lower extremities. Other rare symptoms include nausea, vomiting, and obstructive uropathy [3]. There have been no reports detailing symptoms resembling those of our patient which presented as merely thoracic symptoms. The subphrenic tumor in our patient might have produced an extra-mechanical load on the left hemidiaphragm, which presented as an elevated hemidiaphragm on chest radiography.

Such a mass effect will reduce functional residual capacity, close airways in the lung bases, and result in a condition mimicking an obesity-hypoventilation syndrome [7].

Retroperitoneal teratomas are often encapsulated and composed of cystic and solid areas. The cysts are lined by a variety of epithelia, including squamous epithelium, glandular-like cylindrical epithelium, and ciliated cells [8]. The Rokitansky body or dermoid plug is an eccentric cystic or solid tissue protrusion into a cystic teratoma. The plug is detected by US or CT in nearly 50% of cases [2].

Calcification within the tumor or a calcified rim of a cystic wall is found in 60%-83% of retroperitoneal teratomas and in 74% of benign ones. However, calcification also occurs in 25% of malignant teratomas [2, 3].

Fat is identified in 61% of retroperitoneal teratomas and not detected in one-third of cases. The fat is either adipose tissue or sebum, or both [2]. A fat-water interface is present if there is sebum and fluid in the cystic teratoma. A chemical shift artifact can be seen at the fat-water interface on T2-weighted sequences in ovarian teratomas. [9]. Necrosis and hemorrhage are apt to occur in malignant cases [8]. In spite of the aforementioned imaging characters, the only way to distinguish benign teratomas from malignant ones is through a histopathologic examination [8].

CT and US have been used to evaluate the retroperitoneum [1, 4]. With the advent of MRI, it was found to be superior to CT and US in delineating tissue contrast and internal contents of a cystic retroperitoneal teratoma, including keratoid tissues, hair follicles, sebaceous glands, fat, bone, and bone marrow [4]. Due to multiplanar capability, a high signal-to-noise ratio, and good tissue contrast, MRI is superior to other imaging studies in delineating the internal components of tumors as well as the relationship of the mass to adjacent organs [4].

An abnormally high serum CA-19-9 level in mature cystic teratomas originating from different areas was proven in the literature [10-12]. CA-19-9 was immunohistochemically demonstrated in the bronchial glands and bronchial mucosa of teratomas [12]. These reports can explain the high serum CA-19-9 level in our patient with respiratory epithelium in the tumor.

The clinician in this case chose endoscopic ultrasound-guided needle aspiration instead of CT-guided biopsy for preoperative diagnosis.
under concern for pneumothorax. In fact, CT-guided biopsy is a safe and accurate modality for diagnosis of retroperitoneal lesions. The rate of complications after CT-guided biopsy is 1.1%, with 73% of complications being hemorrhage and 27% pneumothorax [13]. CT-guided biopsy should have been performed in this case.

The treatment of choice for a retroperitoneal teratoma is complete surgical excision [8]. The percentage of malignant degeneration of a retroperitoneal teratoma is 10% in children and 26% in adults [6]. The figures are much higher as compared with that of 2% for teratomas arising from the ovary, which is the most frequent site of mature cystic teratomas [9]. Complication rates derived from the mass effect are 100% if left untreated [8].

Our case was not diagnosed as a cystic teratoma initially because of the absence of typical imaging characters. The mass was purely cystic with no solid and fat component or fluid-fluid interface on CT and MR images with different pulse sequences. In addition, no Rokitansky bodies, hair, nor calcification were identified on different imaging modalities. In our case, other retroperitoneal neoplasms or masses, especially a mass with a cystic component should be considered. These include cystic neoplasm of the pancreas, pancreatic pseudocyst, cystic lymphangioma, hemangioma, cystic hamartoma, cystic teratoma, pyogenic abscess, cold abscess, lymphocele, and urinoma. Cystic tumors of the kidney and adrenal gland were not favored in this case [5, 14]. Furthermore, the presence of high serum CA-19-9 level and the impression of malignancy by cytological examination made the accurate preoperative diagnosis of this patient more difficult.

REFERENCES

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以臨床呼吸困難表現的後腹膜腔畸胎瘤

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後腹膜腔是成人畸胎瘤不尋常的發生部位。本篇報告一位31歲女性病患因後腹膜腔畸胎瘤以主觀呼吸困難及胸部不適求診。胸部X光片顯示左橫膈膜上移，超音波圖顯示左橫膈膜下低回音腫瘤，電腦斷層攝影顯示左橫膈膜下低密度腫瘤。內視鏡超音波導引細針抽吸術疑為脂肪肉瘤，磁振造影術顯示左橫膈下腫瘤，它在T1加權影像與脾臟同訊號強度，T2加權影像為高訊號強度。畸胎瘤的影像學特徵，如脂肪、液體-液體界面、鈣化、毛髮及Rokitansky氏體在本病例的超音波圖、電腦斷層攝影及磁振造影術均未呈現。病患的胸腔症狀在外科手術切除腫瘤後完全消失。

關鍵詞：後腹膜腔腫瘤；畸胎瘤；磁振造影術