Intra-abdominal Desmoplastic Small Round Cell Tumor: CT and MR Findings: a case report

Wen-Yu Lee, Chin-Ming Jeng, Chih-Yu Hsu, Yung-Chuan Sung, Shih-Hung Huang, Young-Chen Wang, Chin-Hue Kung, Chan-Ying Wu

Department of Radiology, Cathay General Hospital

Desmoplastic small round cell tumor is a rare malignant small cell neoplasm which tends to occur in adolescents and young adults. This tumor usually presents as an abdominopelvic mass with intraperitoneal and hepatic metastasis. We report a case presenting as abdominal masses with unilateral pleural seeding.

A 15-year-old woman without previous history of major diseases came to our hospital because of shortness of breath for 2 weeks. Chest X-ray revealed massive amount of left pleural effusion. CT and MRI suggested two intraabdominal masses with multiple left pleural seeding and massive left pleural effusion. The definitive diagnosis was made with the aid of echo-guided fine needle biopsy.

Key words: CT; Desmoplastic small round cell tumor; Magnetic resonance imaging

Desmoplastic small round cell tumor is an aggressive malignant small round cell neoplasm that mainly affected adolescents and young adults. It usually presents as abdominal discomfort with abdominal mass [1]. The purpose of this paper is to describe a rare case of desmoplastic small round cell tumor presenting as shortness of breath with massive left pleural effusion on CT & MRI.

CASE REPORT

A 15-year-old girl was quite well until two weeks ago. She was sent to a local hospital due to shortness of breath. Fever was also noted. Massive left pleural effusion was found. A pig-tail catheterization for effusion drainage was undertaken at that time. She was then transferred to our hospital for further treatment. Physical examination at emergency department showed diminution of breath sound at left lower lung field. The cytologic findings in pleural effusion revealed many large pleomorphic hyperchromatic cell with prominent nucleus and small lymphocytes.

Plain chest radiographs showed massive left pleural effusion without shifting of mediastinum. Trachea was at the midline (Fig. 1).

Chest CT showed nodular thickening of left pleural surface with massive left pleural effusion (Fig.2a). Abdomen CT revealed two heterogeneous masses over left upper abdomen (Fig.2b). No definite calcification of the masses was found. Neither abdominal lymphadenopathy nor hepatic metastasis was noted. Chest and abdomen MR imagings revealed two masses over left upper abdomen. They were inhomogeneous low signal intensity on T2*weighted image (SPGR), intermediate signal intensity on T1-weighted image, heterogeneous contrast enhancement on Gd-T1-weighted image. Circumferential nodular thickening of left pleura with massive left pleural effusion was revealed (Fig.3a, 3b, 3c, 3d).
The definite diagnosis was made by echo-guided fine needle biopsy from the left infradiaphragmatic area. Histological examination showed desmoplastic round cell tumor (Fig.4). No pleural biopsy was done. According to histological findings, the pleural lesion was regarded as metastasis.

The patient received chemotherapy. She is regularly followed up at OPD. After therapy for 6 months, her general condition is stable.

**DISCUSSION**

Desmoplastic small round cell tumor is a rare, aggressive neoplasm that tends to affect adolescents and young adults. It is characterized by a reciprocal translocation t(11;22)(p13;q12) associated with the EWS-WT1 (Ewing sarcoma-Wilms tumor 1) gene fusion transcript [2]. It has a male predominance in previous reports [3]. But a nearly equal sex ratio is reported by Pickhardt et al [4]. The manifestations of desmoplastic small round cell tumor are related to its location. The presentations of intra-abdominal desmoplastic small round cell tumor are usually non-specific such as abdominal or pelvic symptoms and palpable masses. The typical histological findings of desmoplastic small round cell tumor is variable-sized clusters of small, round or spindled cells lying in a desmoplastic stroma [2].

The characteristic CT presentation of desmoplastic small round cell tumor is single or multiple peritoneal masses without any original organ that are located within the omentum or the mesentery, or adjacent to the bladder [4]. Tumors are often inhomogenenous on CT due to focal hemorrhage or necrosis. Lymphadenopathy, punctate calcification of tumor, diffuse peritoneal thickening, hydronephrosis and hepatic metastasis are less common findings [3,4,5,6]. Pleural dissemination is accompanied by disease spreading in one study [3]. In our case, two intraperitoneal masses without other intraperitoneal manifestation were found. Obvious pleural rind with massive pleural effusion was noted.

Characteristics MRI findings of desmoplastic small round cell tumor are intermediate signal intensity on T1-weighted images, high signal intensity

![Figure 1. CXR shows massive left pleural effusion without shifting of the mediastinum. The trachea was at the midline.](image1)

![Figure 2. a. Contrast-enhancing CT at the level of hepatic dome shows nodular thickening (white arrowheads) of left pleura with massive left pleural effusion, a pig-tail catheter in place. b. Contrast-enhancing CT at the level of celiac trunk shows two heterogeneous masses (black arrows) over left upper abdomen.](image2)
MRI can well depict tumor location and extension by multiplanar images than other modalities.

Because no specific features of intra-abdominal desmoplastic small round cell tumor. It must be differentiated with multiple peritoneal masses without obvious original organ. The differential diagnosis includes malignant mesothelioma of peritoneum, non-Hodgkin’s lymphoma, malignant gastrointestinal stroma tumor, pseudomyxoma peritonei, peritoneal metastasis, tuberculosis. The malignant mesothelioma of peritoneum seems to present as small modules lining the surface of parietal peritoneum with a dominant mass in one part of abdomen. The non-Hodgkin’s lymphoma usually mimics carcinomatosis with mesenteric and retroperitoneal lymphadenopathy. The pseudomyxoma peritonei is low-attenuation masses with scalloping parenchymal organ margins on CT. The peritoneal metastasis comes from ovarian and gastrointestinal primaries with masses and soft tissue infiltration and ascites. The tuberculosis may show low attenuation lymphadenopathy, exudative ascites, mesenteric masses on CT.

The treatment of desmoplastic small cell is difficult including chemotherapy, radiotherapy,
surgical resection. Outcome of the majority of cases are poor despite of aggressive therapy. The patients usually die within the first 2 years after diagnosis. However, recent studies of combined modality protocol associated with autologous peripheral blood stem cell transplantation have been tried [8].

**Figure 4.** Photomicrography of the specimen shows the tumor composed of cohesive epithelial-like nests and trabeculae of small to medium-sized, round hyperchromatic cells, with surrounding desmoplastic stroma (Hematoxylin and eosin stain, x 200).

---

**REFERENCE**


5. Varma DG, McDaniel K, Ordenez NG, et al. Primary malignant small round cell tumor of the abdomen: CT findings in five cases. AJR 992; 158: 1031-1034


腹腔內促纖維性小細胞瘤：電腦斷層及磁振影像之病例報告

李紋瑜¹  鄭慶銘¹  徐志育²  宋詠娟²  黃世鴻³  王永成¹  孔慶惠¹  吳昭瑩¹

國泰綜合醫院  放射線科¹  內科²  病理科³

促纖維性小細胞瘤是一種十分罕見的惡性腫瘤，好發於青少年及年輕人。通常以腹部骨盆腫瘤併腹膜及肝轉移表現。我們要報告一例以腹部腫瘤表現併單側肋膜轉移。

一位 15 歲之女性，以前沒有疾病史，由於兩週呼吸急促來本院就診。胸部 X 光片發現大量左側肋膜積水，電腦斷層及磁振影像發現腹腔內腫瘤合併多處左側肋膜轉移及大量肋膜積水。確定診斷是靠超音波下細針切片。

關鍵詞：電腦斷層；促纖維性小細胞瘤；磁振影像