We reported a case of anterior mediastinal Langerhans cell histiocytosis in a 3-month-old male infant. CT scan showed a huge heterogeneously enhanced mass with cystic compartments and punctuated calcification in the anterior mediastinum, interstitial reticulonodular infiltration with scattered cysts in both lungs, and several hypodense nodules in the liver.

**Key words:** Langerhans cell histiocytosis; Anterior mediastinum; Computed tomography

The presence of anterior mediastinal masses has rarely been reported in Langerhans cell histiocytosis [1,2]. Only a few cases of histiocytic infiltration that produce anterior mediastinal masses have been reported, and they are commonly associated with varying degrees of lung involvement [3]. Solid mediastinal masses may cavitate and cause thymic cysts [2]. We reported a case of Langerhans cell histiocytosis in a 3-month-old male infant, in whom a huge anterior mediastinal mass with bilateral pulmonary reticulonodular infiltration and hepatic hypodense nodules was detected by CT images. CT-guided biopsy of the huge anterior mediastinal mass was performed for tissue proof to achieve the diagnosis.

**CASE REPORT**

A 3-month-old male infant (full-term, BBW 3228gm, Apgar score: 1'-9, 5'-10) had suffered from intermittent fever (body temperatures of 38°C) and cough since the preceding month. He had been treated with medication for upper respiratory tract infection in a community clinic, then was sent to our hospital due to aggravation of the symptoms and development of hemoptysis and shortness of breath. On physical examination, the conjunctiva was pale and the chest was symmetrically expanded. Crackle breathing sound was noted in both lungs. The liver and spleen were impalpable, and there was no rash or ecchymosis over the skin. Blood examination showed hemoglobin of 9.7 gm/100ml, hematocrit of 29.1%, and elevated serum alpha-fetoprotein of 201 per unit. Otherwise laboratory studies revealed negative findings. Urine analysis of vanillylmandelic acid and homovanillic acid was negative as well.

Chest radiograph revealed enlargement of the mediastinum with diffuse interstitial infiltration in both lungs (Figure 1). CT scan revealed a huge, heterogeneous enhanced mass in the anterior mediastinum with...
a cyst and punctate calcification. The adjacent great vessels were encased (Figure 2a & 2b). Diffuse interstitial lung infiltration with nodularity, reticulation, and multiple cysts in both lungs was also noted (Figure 3). In addition, a few hypodense nodules were found in the right lobe of the liver (Figure 4). Under general anesthesia, CT-guided biopsy of the anterior mediastinal mass was done. Microscopically, medium-sized hyperchromatic cells with abundant pinkish cytoplasm and heavy eosinophilic infiltration were noted (Figure 5). Immunohistochemical stains revealed that the cells were positive for S-100 and LCA. The pathologic diagnosis was Langerhans cell histiocytosis. A whole body bone scans (Tc-99m) was performed for searching any other involved focus, but no positive finding was obtained. This patient received chemotheraphy with Mercaptopurine, Prednisolone, Vinblastine and Etoposide. Partial response of the lesion to the treatment was noted at the 10-month follow up.

DISCUSSION

Langerhans cell histiocytosis is described as a disease of unknown etiology. It is believed to be due to immunodysregulation [4] or a specific dendritic antigen-presenting cell [5]. Clinically, it is an illness with insidious onset. Initially, there may be a cutaneous rash or upper respiratory infection. Progressive weakness, weight loss, anemia, lymphadenopathy and hepatosplenomegaly usually developed. Imaging studies frequently demonstrate infiltrative lesions of the lungs and the bones. Pathologically, bone marrow, lymph nodes, liver and spleen, and, less frequently, other tissue are infiltrated with nonlipid histiocytic and reticuloendothelial cells. In general, the organs of the reticuloendothelial system are involved earlier and to a greater extent than other organs. Therefore, lymphadenopathy, hepatosplenomegaly, and bone marrow infiltration tend to dominate the disease picture.

Although Langerhans cell histiocytosis may involve various organs with different presentations, its manifestation as an anterior mediastinal mass with calcification is rare [3,6]. The calcifications have unknown etiology, but may be indicative of thymic infiltration with histiocytosis [3]. Thymus involvement Langerhans cell histiocytosis may bring into
being a large anterior mediastinal mass, usually in association with varying degree of lung involvement [1-3, 6].

The most common lung abnormalities are cysts, nodules, and reticulation [3]. They may be primary, or parts of a systemic process. Development of pulmonary cysts occurs either in areas of histiocytosis necrosis or in the relatively normal lung distal to bronchial infiltration or obstruction [2]. Clark postulated that pulmonary cysts may be resulted from direct communication of destructed lung parenchyma and bronchi [7]. End-stage lung involvement by Langerhans cell histiocytosis may have a honeycombed appearance, corresponding to joined and confluent cysts. Rarely, ground-glass opacities are encountered, corresponding to alveolitis on histologic examination [3]. A few autopsy proven cases of liver involvement by Langerhans cell histiocytosis have been reported, but none of them had imaging documentation [4,8]. In this case, several hypodense nodules in the liver were clearly shown on CT.

In our case, CT demonstrated a widespread lesion involving the anterior mediastinum, both lungs and the liver. Cystic areas were noted in both the mediastinal lesion and both lungs. The differential diagnosis is quite limited in this 3-month-old infant. Teratoma, thymic hyperplasia or cysts, lymphoma and lymphangioma are the common anterior mediastinal tumors in pediatric patients [9] but all of them are rarely associated with pulmonary involvement or liver invasion. Although Langerhans cell histiocytosis involving the anterior mediastinum is rare and remains a diagnostic challenge, the CT findings in this case may offer some clues while CT-guided biopsy is feasible in attaining the histologic diagnosis.

![Figure 3. CT scan of chest with lung window displayed diffuse lung infiltration, nodularity, and multiple cysts in the both lungs.](image1)

![Figure 4. Enhanced CT scan of upper abdomen showed two ill-defined heterogeneous masses in the right lobe of the liver.](image2)

![Figure 5. Photomicrography of histologic specimen showed medium-sized hyperchromatic cells with abundant pinkish cytoplasm and heavy eosinophilic infiltration. (200X)](image3)

REFERENCES

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侵犯前縱隔腔，肺及肝的Langerhans細胞組織症：
電腦斷層的表現

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我們報告一例罕見的前縱隔腔Langerhans細胞組織症，發生於一位三個月大的男嬰，電腦斷層掃描檢查在前縱隔腔可見一巨大、龐雜造影的腫瘤，內含有水囊樣及鈣化成份。兩側肺部有組織間腺與網狀結節的浸潤及水囊生成，同時在肝臟發現有多個低密度結節。

關鍵詞：前縱隔腔，Langerhans細胞組織症，電腦斷層掃描