

Isolated Mediastinal Langerhans Cell Histiocytosis in a 7-Year-Old Child

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Abstract- We describe the case of a 7-year-old boy who presented with suprasternal swelling. The chest computed tomography scan showed a large mass of mediastinum. Biopsy from the mediastinal mass was performed. Histologic and immunohistochemistry findings were consistent with Langerhans cell histiocytosis [LCH]. LCH should also be considered in the differential diagnosis of anterior mediastinal tumor in children.

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Introduction

Langerhans cell histiocytosis (LCH) encompasses a disparate group of diseases and distinct clinical syndromes, including solitary eosinophilic granuloma, Letterer-Siwe disease, and Hand-Schuller-Christian disease (1). The clinical spectrum of LCH ranges from a trivial single lesion to aggressive and potentially lethal disseminated disease (2). In most cases, LCH presents as the unifocal disease that typically affects bone; however, the skull bone is the most common site, and less commonly, lymph node, bone marrow, central nervous system, spleen, skin, or lung may be involved (3). Multifocal disease commonly affects young children and can be either single system, as in Hand-Schuller-Christian disease, or multisystem, as in Letterer-Siwe disease. Children with liver, spleen, lung, or bone marrow involvement are at the highest risk for death from LCH and are therefore classified as having high-risk LCH (4). Thymus and mediastinal lymph node involvement in LCH have been rarely reported (5). To the best of our knowledge, an isolated LCH originating from the mediastinum in late childhood has not been previously described.

Case Report

We describe 7-year-old boy who admitted to pediatric

oncology department with suprasternal mass. He had not fever, fatigue, pallor, lymphadenopathy, hepatosplenomegaly, bone pain and swelling, skin rash, weight loss. Hematologic investigations showed white cell blood count (WBC) of $5.9 \times 10^3/\mu\text{L}$, hemoglobin of 12.2 g/L, and platelets of $388 \times 10^3/\mu\text{L}$. Other laboratory tests, including erythrocyte sedimentation rate, lactate dehydrogenase, blood urea nitrogen, creatinine, aspartate aminotransferase, alanine aminotransferase, Uric acid, calcium, phosphorus, ferritin, showed normal results.

Thoracic computed tomography (CT) revealed an isodense lobulated anterior superior mediastinal and thoracic inlet mass of 64x28 mm in diameter with contrast enhancement, which caused the displacement of cervical vessels without encasement. Adjacent rib and sternum invasion was not seen. There was no apparent fat plane between the mass and thymus gland (Figure 1).

Biopsy from the mediastinal mass showed infiltration with histiocytes and eosinophils consistent with the diagnosis of LCH. The diagnosis was confirmed with immunohistochemistry (IHC) staining that was positive for S100 and CD1a.

As LCH is often a multi-system disease, an abdominal CT scan, skeletal survey, and technetium bone scan were obtained, which were normal. Also, bone marrow aspiration and biopsy were unremarkable.

Subsequently, the patient underwent chemotherapy

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Mediastinal langerhans cell histiocytosis

with Histiocyte society LCH III protocol. Over the next 6 months, his mediastinum appeared to be normal on a chest CT scan (Figure 2).

He remains in clinical, hematological, and radiological remission for the last 4 years with no evidence of the development of multi-system LCH or recurrence of mediastinal mass.

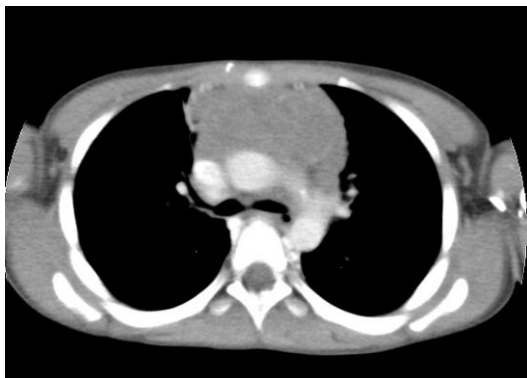


Figure 1. CT scan shows an isodense lobulated anterior superior mediastinal mass of 64x28 mm in diameter with contrast enhancement



Figure 2. Chest CT scan shows complete resolution of mediastinal mass after 6 months of chemotherapy

Discussion

The main clinical and radiological features of LCH in children are due to bone disease and extraosseous involvement is less common (6). Langerhans histiocytosis with mediastinal involvement is rarely reported (7). A review of literature by Ducassou S, et al. from 1963 to 2010 identified 50 childhood cases of LCH with thymic and/ or mediastinal involvement (8). Congenital cysts, lymphoma, Germ cell tumor, thymoma, intrathoracic thyroid tissue, and lymphangioma are the most frequent causes of anterior mediastinal mass in children (9).

In our case the chest CT scan revealed a large mass of the anterior mediastinum. The histological findings

demonstrated an LCH originating from the mediastinum.

To our knowledge, an LCH as an isolated mediastinal mass in late childhood has not been previously described. However, a similar presentation has been reported in an adult by Fahner *et al.*, (10). Although many cases of mediastinal mass with multisystem involvement have been reported, isolated mediastinal LCH is rare (6,7,8,11). Recently, the French LCH registry enrolled 1426 patients with LCH over the past 20 years; 37 (2.6%) had mediastinal mass, and the majority were infants (8). In conclusion, LCH should be considered in the differential diagnosis of isolated mediastinal mass in children. Prompt diagnosis and treatment can lead to excellent results in such children.

Acknowledgments

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