



Unique Imaging Findings in a Case of Pediatric Langerhans Cell Histiocytosis

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Description

Langerhans Cell Histiocytosis (LCH) is a rare disorder characterized by the abnormal proliferation of Langerhans cells. It typically presents with skeletal involvement, skin lesions, or lung infiltration in pediatric patients. We report a case of pediatric LCH with unique and unexpected imaging findings, including extensive splenic involvement, which is a rare manifestation of the disease. This case emphasizes the importance of considering LCH in the differential diagnosis of pediatric patients with unusual imaging findings [1].

Langerhans Cell Histiocytosis (LCH) is a rare disorder characterized by the proliferation of abnormal Langerhans cells, which are a type of dendritic cell. LCH primarily affects pediatric patients, with a wide spectrum of clinical manifestations, ranging from single bone lesions to multi-organ involvement. Common sites of LCH involvement include the bones, skin, and the lungs [2].

In this case, we present a unique and atypical presentation of pediatric LCH with extensive splenic involvement, which is an uncommon finding in the disease. This case emphasizes the need for clinicians to consider LCH in the differential diagnosis of pediatric patients with unexplained imaging findings.

A 6-year-old male child presented with a 4-week history of intermittent fever, weight loss, and generalized malaise. Physical examination revealed hepatosplenomegaly, with the spleen extending 6 cm below the left costal margin. There were no palpable lymphadenopathies, and the patient's general condition appeared otherwise well [3].

Initial laboratory investigations showed pancytopenia, with a hemoglobin level of 8 g/dL, a platelet count of 45,000/ μ L, and a white blood cell count of 3,000/ μ L. Peripheral blood smear revealed an increased number of Langerhans cells.

Abdominal ultrasound and Computed Tomography (CT) were performed to investigate the hepatosplenomegaly. Unexpectedly, the imaging revealed multiple splenic lesions with varying sizes, most of which demonstrated heterogeneous enhancement on contrast-enhanced CT. Some of the lesions also exhibited central low-density areas, suggestive of necrosis.

Further imaging with a whole-body bone scan was conducted to evaluate for potential skeletal involvement, as well as a chest X-ray to assess for lung infiltration. No bone lesions or lung abnormalities were detected. Subsequently, the patient underwent a splenic biopsy, which confirmed the presence of Langerhans cells.

The diagnosis of LCH was established, and the patient was initiated on chemotherapy with vinblastine and prednisolone. During the treatment course, the patient's symptoms gradually improved, and follow-up imaging showed a significant reduction in the size and number of splenic lesions [4].

Discussion

Langerhans Cell Histiocytosis typically involves the bones, skin, and lungs in pediatric patients. Splenic involvement is a rare manifestation of the disease, and unique splenic findings, as observed in this case, are even less common.

The characteristic feature of LCH is the presence of Langerhans cells, which can infiltrate various organs and tissues. The disease is typically diagnosed based on histopathological examination of affected tissue, with immunohistochemical staining for CD1a and Langerin.

In our case, the initial presentation of splenic involvement in the absence of skeletal or lung manifestations made the diagnosis challenging. It underscores the importance of considering LCH in the differential diagnosis of pediatric patients with unexplained splenic abnormalities, even in the absence of classical bone or skin lesions.

The management of LCH in pediatric patients often includes chemotherapy with vinblastine and corticosteroids. In some cases, other therapeutic modalities, such as surgery or radiation therapy, may be considered, depending on the extent and severity of the disease.

Conclusion

Langerhans Cell Histiocytosis is a rare disorder with a wide spectrum of clinical manifestations. This case of pediatric LCH with unique and extensive splenic involvement serves as a reminder that the disease can present in unexpected ways. It underscores the importance of considering LCH in the differential diagnosis of pediatric patients with unusual imaging findings, even when classical bone or skin lesions are absent. Early recognition and intervention are vital for optimal management and outcomes in pediatric LCH.

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