IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 25, Issue 6 Ser. 3 (June. 2026), PP 56-62
www.iosrjournals.org

ALK-Positive Histiocytosis Presenting with Leukemoid Reaction and Pleural Mass in a Child: A Rare Case Report

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Abstract
A 10-year-old girl presented with intermittent fever, extreme neutrophilic leukocytosis suggestive of a leukemoid reaction, and positive leptospira serology. Initial imaging showed left lower lobe collapse with pleural effusion that evolved into a pleural-based mass. Bone marrow biopsy revealed atypical epithelioid cell infiltration. Immunohistochemistry demonstrated positivity for ALK (D5/F3), vimentin and myeloperoxidase, confirming ALK-positive histiocytosis. PET-CT demonstrated a hypermetabolic pleural mass with extensive skeletal involvement. Lung biopsy supported ALK-positive epithelioid inflammatory myofibroblastic tumor. This case highlights the diagnostic challenges of this rare pediatric neoplasm and the importance of early tissue diagnosis and immunohistochemistry.

Keywords: *ALK-positive histiocytosis, leukemoid reaction, pediatric pleural mass, PET-CT, inflammatory myofibroblastic tumor*

Date of Submission: 06-06-2026 Date of Acceptance: 17-06-2026

I. INTRODUCTION

ALK-positive histiocytosis is a rare histiocytic neoplasm characterized by ALK gene rearrangements and strong ALK protein expression. Clinical manifestations are heterogeneous and frequently mimic infection, inflammatory disease, or malignancy. Pediatric cases remain uncommon, making diagnosis challenging (1).

II. CASE REPORT

A 10-year-old developmentally normal female child presented with intermittent low- to intermediate-grade fever. Initial investigations revealed marked leukocytosis with neutrophilic predominance and a peripheral smear suggestive of leukemoid reaction. HRCT chest demonstrated left lower lobe collapse with focal bronchial narrowing. Despite broad-spectrum antibiotics and doxycycline for positive leptospira serology, symptoms persisted. Hemoglobin was 8.2 g/dL and leukocyte count rose to 1,98,060/cu.mm. Imaging revealed hepatosplenomegaly, pleural effusion and mesenteric lymphadenopathy. Bone marrow biopsy demonstrated sheets of atypical epithelioid cells. Immunohistochemistry showed positivity for ALK (D5/F3), vimentin and myeloperoxidase, confirming ALK-positive histiocytosis. PET-CT revealed a large hypermetabolic pleural-based mass with disseminated skeletal involvement. Lung biopsy further confirmed ALK-positive epithelioid inflammatory myofibroblastic tumor. The patient received vinblastine and prednisolone, while targeted ALK inhibitor therapy was not initiated because of financial constraints.

Figure 1. Coronal CT image showing left pleural-based thoracic mass.

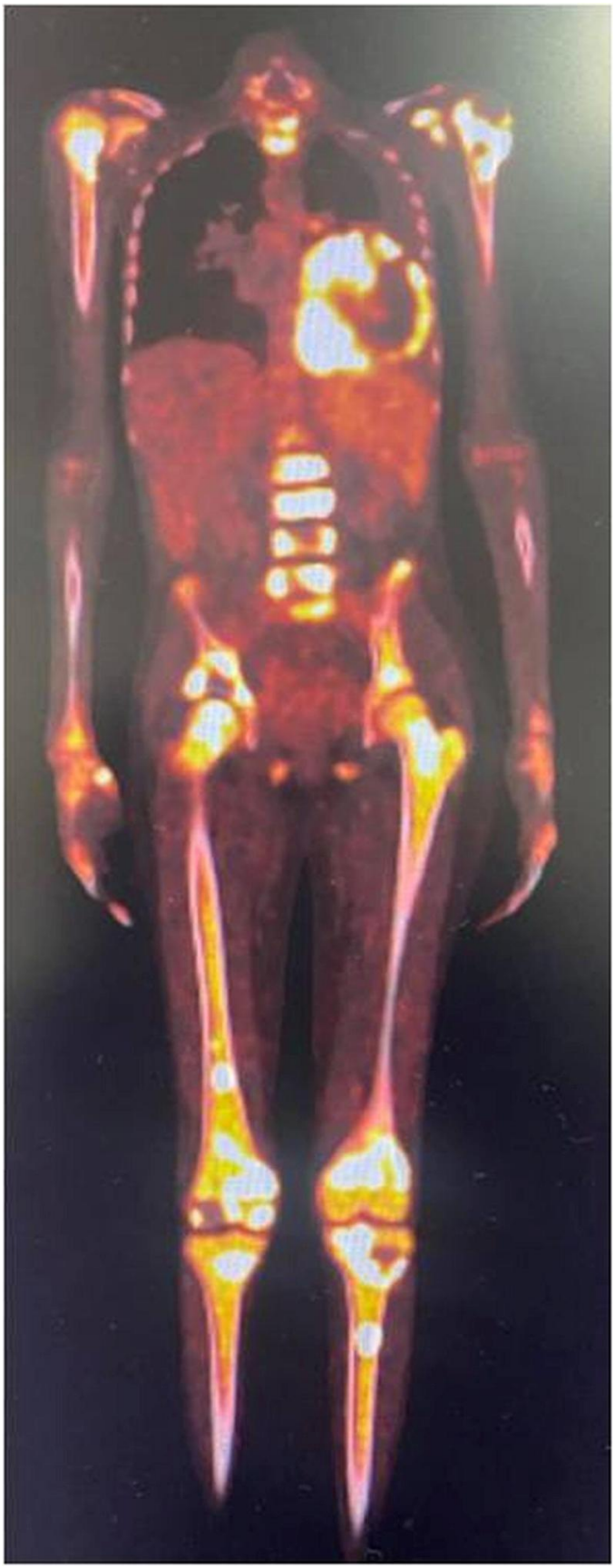

Figure 2. Coronal fused PET-CT demonstrating FDG-avid pleural mass and multifocal skeletal uptake.

Figure 3. Sagittal CT image showing thoracic lesion.

Figure 4. Sagittal PET-CT image showing metabolically active disease.

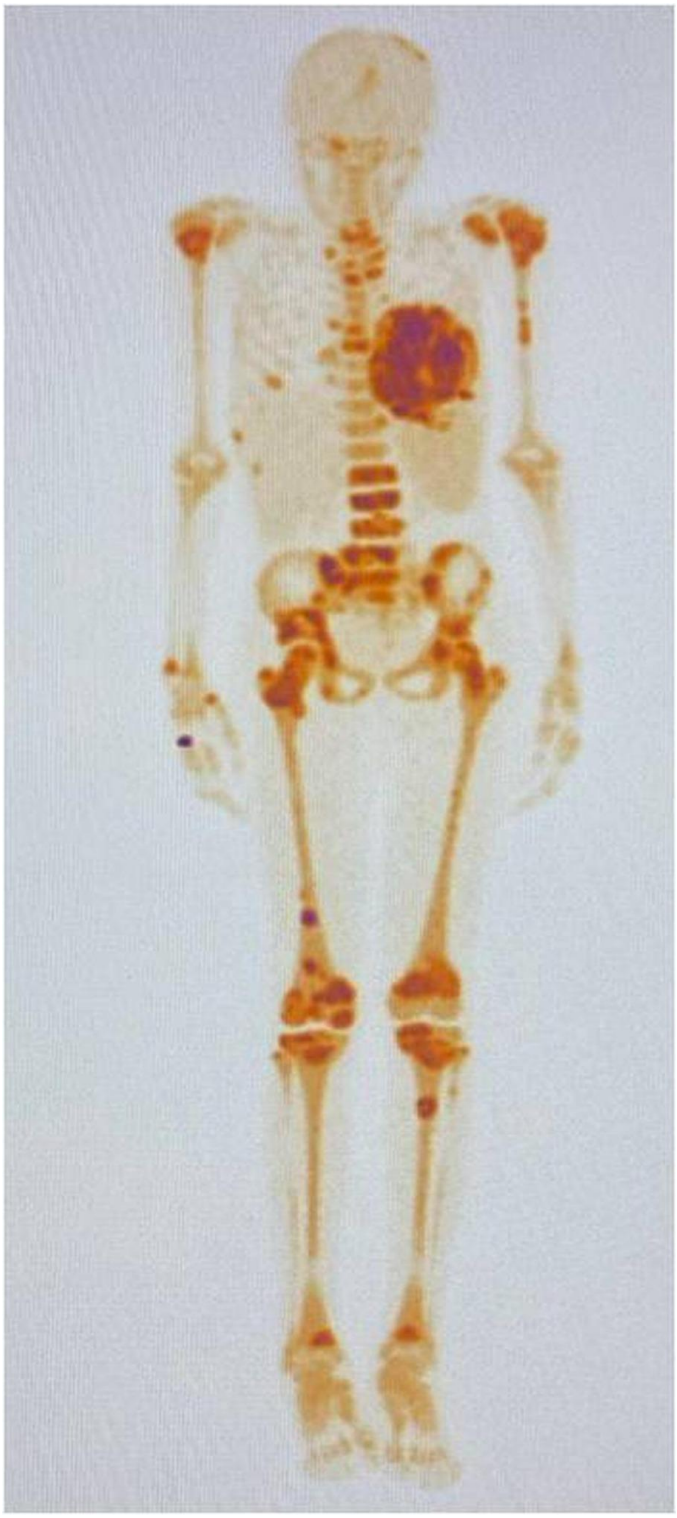

Figure 5. Whole-body MIP image showing disseminated skeletal involvement.

III. DISCUSSION

ALK-positive histiocytosis is a rare and recently recognized histiocytic neoplasm characterized by ALK gene rearrangements. It can present at any age but is extremely uncommon in children. (2,3)**.**The present case is notable for its aggressive presentation with extreme leukemoid reaction, pleural mass, and extensive skeletal involvement. The radiological findings closely mimicked malignant neoplasms, highlighting the diagnostic dilemma. Histopathological examination and immunohistochemistry remain the cornerstone of diagnosis. ALK immunopositivity is a defining feature and helps distinguish this entity from other histiocytic disorders and sarcomas. (2,3)**.**Identification of ALK rearrangements has therapeutic implications, as ALK

inhibitors have shown promising results in reported cases (5)**.**According to the revised classification of histiocytic disorders, ALK-positive histiocytosis is now recognized as a distinct clinicopathological entity (1).

IV. CONCLUSION

ALK-positive histiocytosis should be considered in the differential diagnosis of children presenting with persistent fever, leukemoid reaction and mass lesions. Early biopsy and immunohistochemical evaluation are essential for timely diagnosis and management.

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