

## Case Report

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# Anterior Chest Wall Bulging: A Rare Initial Manifestation of Acute Lymphoblastic Leukemia in a Child

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## Abstract

Acute lymphoblastic leukemia (ALL) has several clinical manifestations on the base of bone marrow infiltration and the extent of extra medullary involvement. Anterior chest wall bulging as an initial presentation is rare. The aim of this case report study is to present a 3.5 year-old-boy presented with anterior chest wall bulging without history of trauma since 1 month ago. Tc99m-MDP bone scan showed multifocal active bony pathology in sternum, lower thoracic and all lumbar vertebra and right sacral ala. Bone marrow aspiration and biopsy was in favor of acute precursor lymphoblastic leukemia, B cell type. Therefore, acute lymphoblastic leukemia has different initial presentation and anterior chest wall bulging can be a rare initial sign of acute lymphoblastic leukemia in children.

**Keywords:** Acute lymphoblastic leukemia; Sternum; Chest wall; Bulging

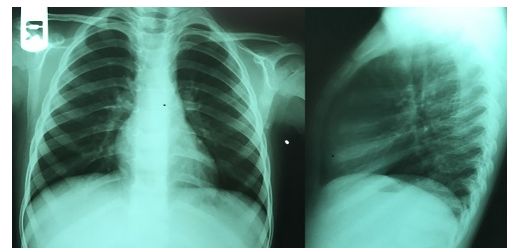
## Introduction

Acute Lymphoblastic Leukemia (ALL) is the most common pediatric malignancy with a cure rate of about 80-90%. [1,2]. The clinical manifestations of ALL could be gradually or acutely. The signs and symptoms of ALL are commonly due to bone marrow failure, including thrombocytopenia, anemia and neutropenia. So, the patients can present with bleeding, petechia, purpura, fatigue, anorexia, malaise, bone pain, pallor and fever. Fever is the most common symptom. Extramedullary leukemic infiltration can be presented by lymphadenopathy, hepatomegaly and splenomegaly [1,3].

Unusual sites of extra medullary involvement by leukemic cells including ocular, bladder and pancreas, are extremely rare as a first presentation [4]. Bone pain in ALL is due to leukemic infiltration of bone and periosteum, and often occurs in long bones [5]. Several unusual initial presentations of ALL in children were reported as rare presentations. Cardiac tamponade, spontaneous humerus fracture and osteoporosis, hypercalcemia symptoms, back pain and vertebral compression, acute renal failure and acute abdomen due to splenic infarction are the rare unusual first presentations of pediatric ALL [6-12]. Here in, we reported a case of anterior chest wall bulging as a rare initial manifestation of acute lymphoblastic leukemia in a child.

## Case Report

A 3.5 years old boy presented initially with anterior chest wall bulging since 25 days prior to admission without any history of trauma or injury (Figure 1). The patient also had occasionally generalized body pain and fever since 15 days prior to admission that referred to general practitioner and received antibiotic but without improvement.



**Figure 1:** 3.5 years old boy with anterior chest wall bulging.

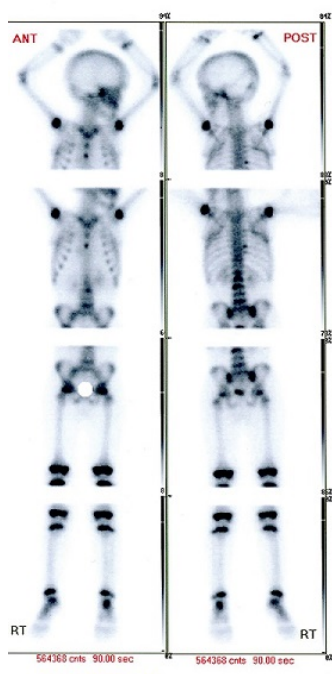
After that, he developed fever and bone pain especially at sternum. The patient had history of glucose-6-phosphate dehydrogenase deficiency. Finally, he admitted for further evaluation in Amir Hospital, affiliated to Shiraz University of Medical Sciences, Shiraz, Iran. In physical examination, he had normal vital sign (Temp: 37.1 orally, RR: 18, PR: 86 and BP: 90/60). Mild tenderness on sternum, tibia and vertebral bone were revealed but there were no limitation of movement or joint swelling. No lymphadenopathy, hepatomegaly and splenomegaly were detected. Laboratory studies were summarized in (Table 1). Chest X Ray (PA and Lateral) shows sternal protrusion without infiltration or mass in lung parenchyma and, also no signs of osteoporosis were detected in bony parts (Figure 2). Calcium, phosphor and parathyroid hormone were normal (Table 1).

Tc99m-MDP bone scan performed and demonstrated abnormal increased radiotracer activity in the lower part of body of sternum, lower thoracic and all lumbar vertebrae, some ribs and right sacral ala (Figure 3). Bone marrow aspiration and biopsy was performed and showed hyper cellular marrow mainly lymphocyte with about 70% blasts in favor of acute leukemia. Immunohistochemistry profile on formalin fixed paraffin embedded tissue of bone marrow biopsy was in favor of acute precursor lymphoblastic leukemia B cell type (CD10(+), PAX5(+) and TDT (+)).



**Figure 2:** Chest X ray PA and LAT shows sternal protrusion without infiltration or mass in lung parenchyma, and also no signs of osteoporosis

Immunophenotyping by flowcytometry was done on bone marrow of this patient by a panel of antibodies. Populations of blasts were selected on SSC/CD45 and also SSC/FSC scattergrams. This population account for 70.0% of total cells and has ALL phenotype and is positive for HLADR, CD34, CD45(dim), CD19, CD10 and negative for CD13, CD33, CD11b, CD117, CD14, CD64, CD61, CD5, CD2, CD20, the rest of events other than these gates seems to be normal in appearance. The PCR report of bone marrow shows no evidence of t (9; 22), t (4; 11) and t (1; 19).



**Figure 3:** Tc99m-MDP bone scan performed and demonstrated abnormal increased radiotracer activity in the lower part of body of sternum, lower thoracic and all lumbar vertebrae, some ribs and right sacral ala.

Chemotherapy of standard risk ALL started for him (Vincristine, Prednisolone and Pegylated L-asparaginase with triple intrathecal injection). CSF cytology was negative and bone marrow aspiration on day 7 of treatment had about 10% blast with rapid early response and at the end of induction had less than 5% blast. At now, the patient is on regular follow up and on maintenance therapy with vincristine monthly, dexamethasone orally, 6-mercaptoprine and methotrexate orally. Bone scan become normal and chest wall bulging was improved.

Hematology	Result	Unit	Chemistry	Result	Unit
WBC	5.3	$\times 10^3/\mu\text{L}$	FBS	88	Mg/dL
Neutrophils	28%	-	BUN	9	Mg/dL
Lymphocytes	68%	-	Cr	0.8	Mg/dL
Monocytes	4%	-	Na	139	MEq/dL
RBC	3.60	$\times 10^6/\mu\text{L}$	K	4.5	MEq/dL
Hemoglobin	9.9	gr/dL	Ca	10.1	Mg/dL
Hematocrit	30.1	gr/dL	Po4	5.2	Mg/dL
MCV	78.6	fL	PTH	15.6	Pg/mL
MCH	25.5	Pg	Mg	1.9	Mg/dL
MCHC	32.5	gr/dL	Uric Acid	2.8	Mg/dL
Platelet	350	$\times 10^3/\mu\text{L}$	Protein Total	6.3	gr/dL
Serology	Result	Unit	Albumin	4.6	gr/dL
ESR	40		ALT	13	U/L
CRP	Negative		AST	26	U/L
Wright	Negative		ALP	492	U/L
Widal	Negative		Total Bilirubin	0.5	Mg/dL
2ME	Negative		Direct Bilirubin	0.1	Mg/dL
			LDH	475	U/L

WBC: White Blood Cell; RBC: Red Blood Cell; MCV: Mean Corpuscular Volume; MCH: Mean Corpuscular Hemoglobin; MCHC: Mean Corpuscular Hemoglobin Concentration; ESR: Erythrocyte Sedimentation Rate; CRP: C - Reactive Protein; 2 ME: 2-Mercaptoethanol; FBS: fasting blood sugar; ALT: Alanine Aminotransferase; AST: Aspartate Aminotransferase; ALP: Alkaline phosphatase; LDH: Lactic Acid Dehydrogenase; PTH: Parathyroid Hormone

**Table 1:** Laboratory findings in the case patient

## Discussion

Bone pain is a common manifestation in pediatric ALL that mainly involves long bones because of infiltration of leukemic cells in periosteum and bones [13].

But involvement of sternal bone as a bulging of anterior chest wall is extremely rare as an initial manifestation. Bulging of anterior chest wall is almost fifteenth unusual musculoskeletal presenting symptoms in childhood ALL. However, other bony presentations including, long bone fractures, osteoporosis, vertebral compression and hypercalcemia were seen as an initial presentations of ALL [9-12].

Approximately 40% of childhood ALL initially presents with a limping and bone or joint pain [14]. About 25% of childhood ALL have particular radiographic features such as osteopenia and multiple fractures at diagnosis, vertebral compression, chronic recurrent multifocal osteomyelitis, arthritis, and calf pain. These are rare and atypical musculoskeletal presentations of childhood [14].

Leukemic infiltration in sternal bone with bulging of chest wall was seen in solid tumors, like Ewing sarcoma and lymphoma [15,16]. Anaplastic large cell lymphoma of bone also can present with multiple bone involvement [15,17]. Langerhans cell histiocytosis, anterior mediastinal mass like thymomas, germ cell tumours, lymphomas and active pulmonary tuberculosis with bony involvement should be in differential diagnosis of a patient who present with anterior chest wall bulging. Chest X-ray (PA and Lateral) is helpful as a first imaging study [18-20].

So, in such cases we should think of acute leukemia. In our case report, there was no poor prognostic sign either laboratory or clinically, and the response to standard risk therapy was good. So anterior chest wall bulging at initial presentation of ALL may be not a poor prognostic sign but further study may be needed.

## Conclusion

ALL has several clinical manifestation, anterior chest wall bulging can be a rare initial manifestation of acute leukemia due to infiltration of leukemic cells in sternum.

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