East African Scholars Journal of Medical Sciences
Abbreviated Key Title: East African Scholars J Med Sci
ISSN: 2617-4421 (Print) & ISSN: 2617-7188 (Online)
Published By East African Scholars Publisher, Kenya

Volume-4 | Issue-8 | September-2021 | DOI: 10.36349/easms.2021.v04i08.001

OPEN ACCESS

Case Report

Ph Positive CML in a 17 Year Old

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Article History
Received: 14.07.2021
Accepted: 17.08.2021
Published: 11.09.2021

Journal homepage:
https://www.easpublisher.com

Quick Response Code

Abstract: We report a case of Chronic Myeloid Leukaemia in a 17 year old female who presented with intermittent fever, generalised body ache, loss of weight and appetite for one month duration. She had no other comorbidities. General examination, systemic examination and ultrasound study of abdomen were normal. Hemogram and peripheral smear revealed normal haemoglobin value (12.7g/dl) with moderate leucocytosis (31,100/ cmm), thrombocytosis (8.02 Lakh/ cmm) and moderate eosinophilia (Absolute eosinophil count 1946/ cmm). She was also found to have an elevated level of Lactate dehydrogenase (599 IU/L). All other lab investigations were within normal limit. ANA was negative.

Keywords: Chronic Myeloid Leukaemia, fever, weight, dehydrogenase.

BACKGROUND

The term myeloproliferative syndrome describes a group of disorders that result from an unchecked, autonomous clonal proliferation of cellular elements in the bone marrow. Chronic myeloid leukaemia is the best defined MPN. Natural course of the disease occurs in three phases, chronic, accelerated and blast crisis. Usually occurs in middle aged and older adults. Most striking abnormality in the peripheral blood is extreme leucocytosis (>100x10^9/L). Blood smears exhibit a shift to left with all stages of granulocyte maturation, predominant cells being segmented neutrophils and myelocyte. We report a rare case of a young female presenting with major (p 210) BCR/ABL1 fusion gene positive Chronic Myeloid leukaemia with moderate leucocytosis, basophilia and minimal shift to left of neutrophils.

CASE PRESENTATION

A 17 year old female presented with intermittent fever, body ache and loss of weight and appetite of one month duration. She had no other comorbidities. No history of connective tissue disorders/ contact with tuberculosis / high risk behaviour.

General examination, systemic examination and Ultrasound examination – abdomen, revealed all normal findings.

Peripheral smear showed moderate leucocytosis with mild shift to left, eosinophilia and basophilia with thrombocytosis.

Bone marrow study showed increased cellularity, myeloid hyperplasia with predominance of mature forms, megakaryocytes increased with frequent dwarf and monolobated forms. Molecular study revealed major (p210) BCR/ABL 1 fusion protein being positive.

Table 1: Relevant investigations performed during hospital stay

Haemoglobin: 12.7 g%	Bilirubin total: 0.2 mg%
Total count: 31,100/cmm	Bilirubin Direct 0.1mg%
Platelet: 8.02 Lakh/cmm	SGOT: 23 IU/L
AEC: 1946/cmm	SGPT: 18 IU/L
ESR : 7 mm/ first hour	ANA negative
Creatinine : 1mg %	LDH: 599 IU/L
Urea: 23 mg %	Total protein: 7.4 g%
Uric acid: 5.3 mg %	Albumin: 4.6g%

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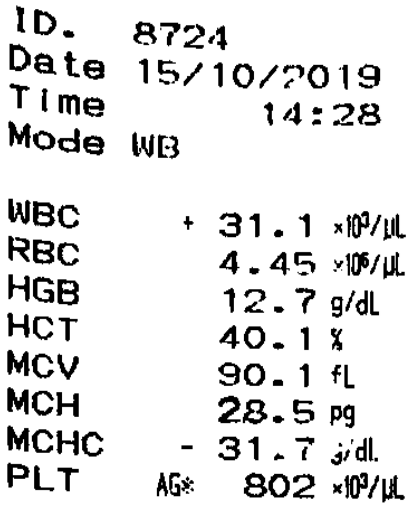

Figure 1: Hemogram

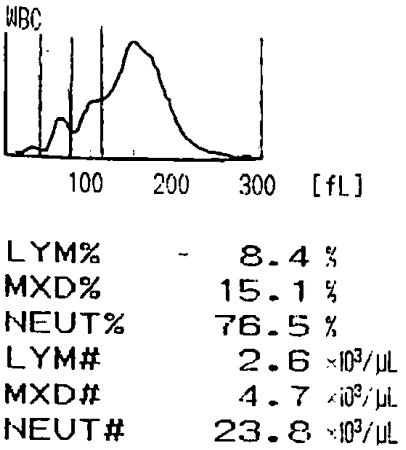

Figure 2: Histogram

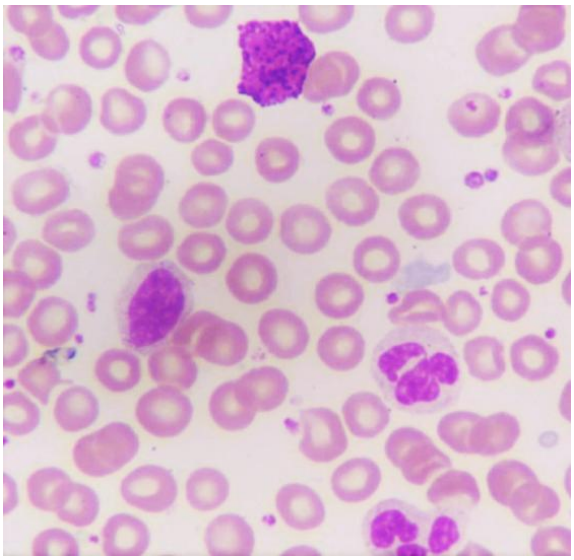

Figure 3: Peripheral smear showing basophil and myelocyte

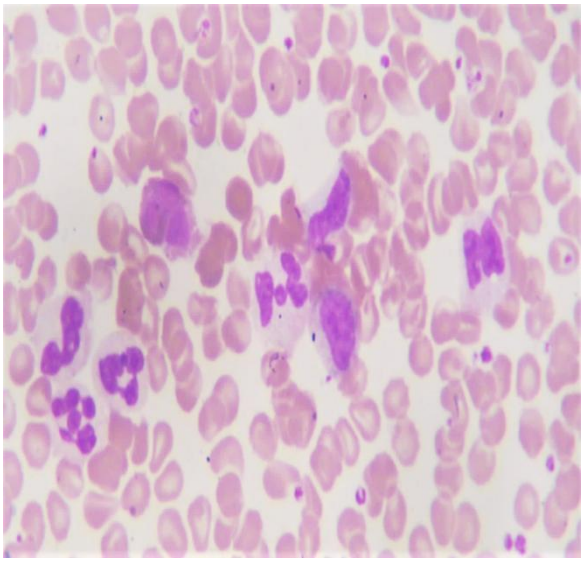

Figure 4: Peripheral smear showing neutrophilic shift to left

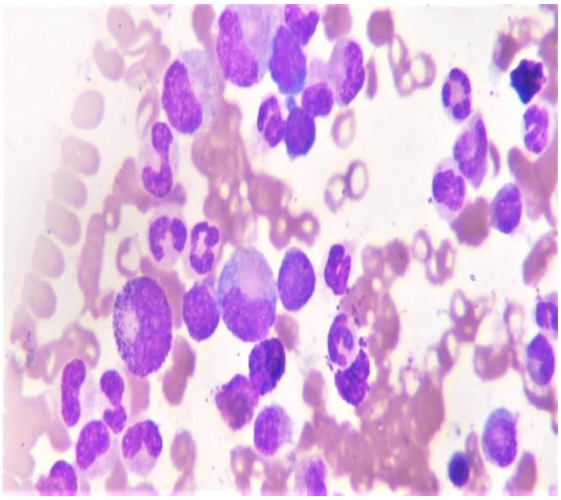

Figure 5: Bone marrow aspirate showing myeloid hyperplasia

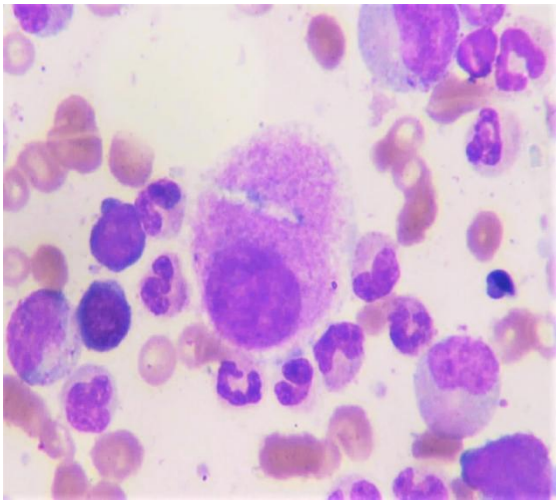

Figure 6: Bone marrow aspirate showing myeloid hyperplasia with monolobated megakaryocyte

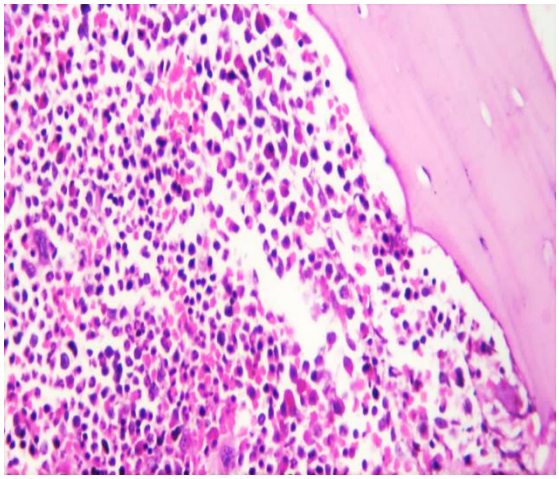

Figure 7: Bone marrow trephine biopsy showing increased cellularity

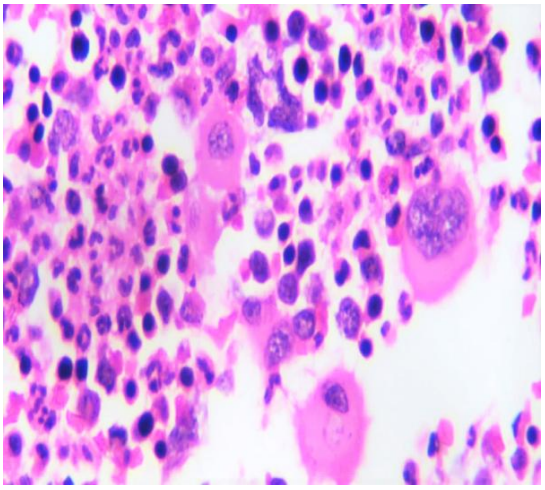

Figure 8: Bone marrow trephine biopsy showing myeloid hyperplasia, increased megakaryocytes with dwarf and monolobated forms

Diagnosis: Chronic myeloid leukaemia – Chronic phase.

Treatment: After report of major (p210) BCR/ABL 1 fusion protein becoming positive, patient was started on Imatinib mesylate.

Outcome and follow up: Patient responded very well to treatment with normalisation of Hemogram.

DISCUSSION

Chromic myeloid leukaemia is most prevalent in seventh, eighth, ninth decades of life. It is characterised by Ph chromosome that result in a fusion gene called BCR/ABL1. Peripheral blood findings are extreme leucocytosis with a shift to left with all stages of granulocyte present. Predominant cells are segmented neutrophils and myelocyte. Eosinophils and basophils are often increased. Thrombocytosis with variation in shape of platelet is seen.

Mohammad Zaid Hussan *et al*., reported 2 cases of CML in 22 year old and 14 year old male patients in whom Imatinib therapy was unsuccessful. Mustafa Yilmaz et al reported 3 cases of CML with isolated thrombocytosis, normal leukocyte count and no organomegaly. No literature on CML with absent myelocyte bulge was noted.

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- Chronic myeloid leukaemia can uncommonly present in young adolescent age group.
- CML can occur with mild to moderate leucocytosis with no organomegaly in the initial stage.
- Myelocyte bulge can be absent in chronic myeloid leukaemia.
- All patients with suspicion of a chronic myeloproliferative neoplasm should be tested for BCR/ABL1 fusion protein.

Cite This Article: Thameem A *et al* (2021). Ph Positive CML in a 17 Year Old. *East African Scholars J Med Sci*, *4*(8), 172-174.