Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Hematology

Pleural Effusion as the Initial Manifestation of Chronic Myelomonocytic Leukemia

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DOI: 10.36347/sjmcr.2024.v12i05.096 | **Received:** 19.02.2020 | **Accepted:** 26.02.2020 | **Published:** 30.05.2024

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Abstract **Case Report**

Chronic myelomonocytic leukemia (CMML) is a disease of the elderly, and by far the most frequent overlap myelodysplastic/ myeloproliferative neoplasm in adults. Aside from the chronic monocytosis in both peripheral blood and bone marrow that remains the cornerstone of its diagnosis, the clinical presentation of CMML includes dysplastic features, cytopenias, excess of blasts, or myeloproliferative features including high white blood cell count or splenomegaly. Prognosis is variable, with several prognostic scoring systems reported in recent years, and treatment is poorly defined. Pleural effusion in patients with CMML is a rare occurrence and poorly understood. We report a rare case of CMML patient presenting with pleural effusion as the first clinical sign.

Keywords: Pleural effusion chronic myelomonocytic leukemia.

INTRODUCTION

Chronic myelomonocytic leukemia (CMML) is, by far, the most frequent of myelodysplastic/myeloproliferative entities recognized by World Health Organization (WHO) classifications [1] with an incidence of about 1/100,000 per year. It is a very heterogeneous disease, with hematological characteristics ranging from those of a myelodysplastic syndrome (MDS) with peripheral monocytosis, to very proliferative forms, characterized by high white blood cell (WBC) counts, splenomegaly, and/or other forms of extramedullary disease. Its diagnosis remains largely based on morphology, though recent advances in flow cytometry of blood monocytes may contribute in difficult cases. Somatic mutations in a small subset of recurrently mutated genes can be detected in almost all patients, some carrying a poor prognostic value. Treatment choices remain poorly supported, since, until recently, CMML patients were included in MDS series, whereas only 1 CMML-specific randomized clinical trial has ever been published to date [2].

Pleural effusion in patients with CMML) is a rare occurrence and poorly understood. We report a rare case of CMML patient presenting with pleural effusion as the first clinical sign.

CASE REPORT

A 72-year-old chronic smoking patient who has had a productive cough for one month with resting dyspnea in a febrile environment without hemoptysis or bleeding. There was no history of prolonged medical illness or significant drug therapy.

Clinical examination finds a polypnetic and febrile patient with right pleural fluid effusion syndrome and stable vital parameters. At the abdominal examination absence of splenomegaly. The chest X-ray shows a pleural appearance opacity that is interesting for the lower half of the right thoracic hemichamps [figure 1]. The blood count shows 16,000 leucocytes, 9 g hemoglobin, and 123,000 platelets. There is 1% circulating blastosis and the myelogram shows a myeloproliferative / myelodysplastic border syndrome type LMMC.

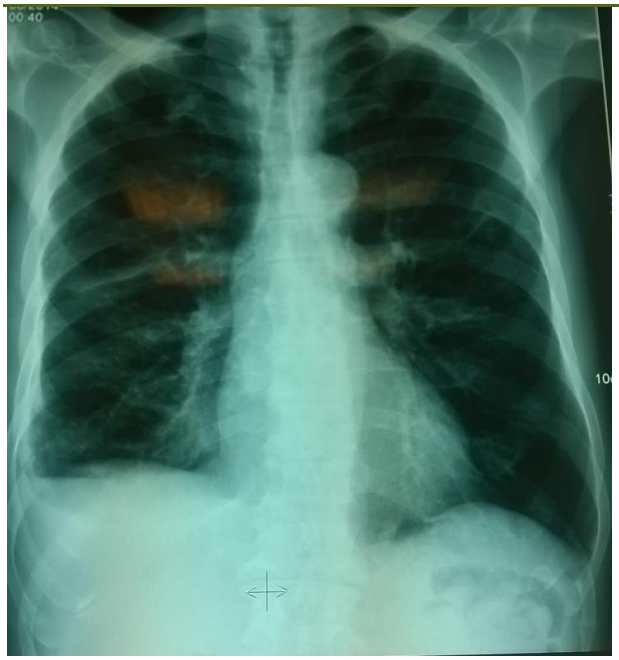

Fig-1: Chest X-ray showed a right pleural effusion

Pleural puncture returned a serumematic fluid with 36% neutrophils and 64% lymphocytes, with no malignant cells. Pleural biopsy could not be performed in the presence of encysted pleurisy of low abundance. The search for koch bacilli in the sputum was negative. The thoracic CT scan shows a right encysted pleurisy. Bronchial fibroscopy is normal and aspiration for malignant cells is normal. The viral hepatitis B, C and HIV serology are negatives. Protein electrophoresis showed a narrow peak of monoclonal appearance with significant hypo albuminemia. The patient was placed on probabilistic antibiotic therapy with protected amoxicillin with clinical improvement and drying of pleurisy. He was referred to the hematology department for further support of his LMMC.

DISCUSSION

CMML patients usually present with increased peripheral and marrow leukocytosis and hepatosplenomegaly which also occur in myeloproliferative disorders. However, the degree of hematopoietic dysplasia, the clinical course, the rate and pattern of leukemic transformation together with the cytogenetic abnormalities are more reminiscent of MDS than of myeloproliferative disorders [3, 4]. It is generally accepted that CMML patients should be kept under observation [5] as they are usually asymptomatic at the time of diagnosis and they may have only a slight monocytosis.

Pleural effusion due to leukemic infiltration in this disease is rare [6]. Still rare is pleural effusion as an initial manifestation of CMML. Analysis of pleural fluid may show increased blasts or in some cases, all stages of granulocytes and a few blasts [7]. In our case, patient presented with exudative pleural effusion with 36% granulocytes and 64% lymphocytes, with no malignant cells.

Several possible mechanisms of pleural effusion in patients with hematopoetic malignancies have been considered. These include:

- Leukemic infiltration into the pleura that usually occurs at the time of or just prior to bone marrow evolution to blast crisis phase [8]. In these cases, the pleural fluid contains a greater proportion of blast cells.
- Nonmalignant causes like infection and hypoproteinemia have also been postulated as the cause of effusion. Therefore, this possibility must be excluded by identification of microorganisms by special stain and/or presence of necrotic debris.

Pleural effusion associated with hematopoetic or lymphoid malignancies are rare. Apart from CMML, there are also reports of multiple myeloma, CML and Non-Hodgkins lymphoma being associated with pleural effusion [9-11]. Pleural effusion being the presenting feature of the same is very rare;

CONCLUSION

Our case offers as a point of interest a revelation on a respiratory mode of an LMMC following a pleural effusion of infectious origin. A focus on this rare pleural manifestation of this pathology is recommended.

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