

Leukemic pleural effusion in AML-M0: Unusual presentation

Anchal Tandon¹, Seema Acharya², Aparna Bhardwaj², Pooja Kala³

From ¹Junior Resident, ²Professor, Department of Pathology, Shri Guru Ram Rai Institute of Medical and Health Sciences, ³Assistant Professor, Government Doon Medical College, Dehradun, Uttarakhand, India.

Correspondence to: Dr. Anchal Tandon, Department of Pathology, Shri Guru Ram Rai Institute of Medical and Health Sciences, Patel Nagar, Dehradun -248001, Uttarakhand, India. E-mail: anchalofficial@gmail.com.

Received - 02 June 2019

Initial Review - 19 June 2019

Accepted - 01 August 2019

ABSTRACT

Extramedullary disease in AML-M0 is a rare occurrence. It is even rarer to encounter breathlessness as primary presenting complaint secondary to a leukemic pleural effusion. Leukemic pleural effusion occurs rarely in acute myeloid leukemia (AML) with minimal differentiation. Patients with AML usually present with weakness, hemorrhage, and recurrent infections. The highest incidence of extramedullary leukemic manifestation is seen in monocytic AML. Here, we report the case of an 18-year-old male who presented with breathlessness and vocal cord palsy. Examination and investigation established diagnosis of an acute myeloid leukemia M0 with leukemic pleural effusion.

Keywords: Acute myeloid leukemia, Extramedullary, Pleural effusion.

Leukemic pleural effusion occurs rarely in acute myeloid leukemia (AML). Patients with AML usually present with weakness, hemorrhage, and recurrent infections. Frequency of extramedullary disease in AML is less than 1% [1]. The extramedullary disease occurs with or precedes bone marrow involvement. Usual sites for the isolated extramedullary disease are bones, soft tissue, lymphnode, orbit, intestine, mediastinum, epidural region, uterus, and ovary. Very few cases of AML with pleural effusion as initial manifestation have been reported [2].

CASE REPORT

An 18-year-old male patient presented with the complaints of breathlessness, hoarseness of voice and fever on and off since 1-month duration. On examination, the vitals were

stable. Physical examination revealed multiple cervical and supraclavicular firm to hard lymph nodes. Chest examination suggested reduced air entry to the left lower lobe with a dull note on percussion. Laryngeal examination showed evidence of the left vocal cord palsy.

X-ray chest posterior-anterior (PA) confirmed the left-sided pleural effusion with underlying lung collapse/consolidation. Widening of the superior mediastinum and tracheal deviation to the right side were also seen (Fig. 1). Contrast-enhanced computed tomography (CECT) chest done subsequently showed conglomerated lymphnode mass in the superior mediastinum with moderate pleural effusion and the sub-segmental collapse of the basal segment of the lower lobe and lingular lobe.

Thoracocentesis was done, the fluid was exudative (protein was 4.5 g/dl) with increased lactate dehydrogenase (LDH) 4809 U/L and



Figure 1: X-ray chest showed a left-sided pleural effusion with underlying collapse /consolidation.

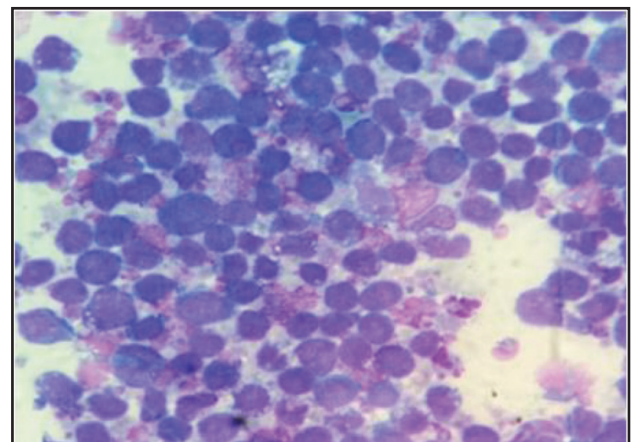


Figure 2: Pleural fluid; MGG; 400x Large atypical cells with high N:C ratio, open chromatin, 1-2 nucleoli. No Auer rods were seen.

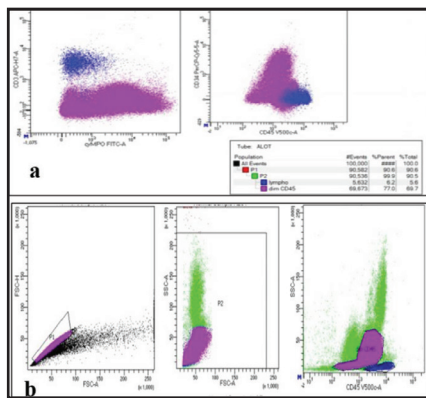


Figure 3: (a and b) Flowcytometry bone marrow aspirate.

adenosine deaminase (ADA)240 U/L. Microscopic examination revealed numerous blast like cells in singles along with mesothelial cells, suggestive of high-grade hematolymphoid malignancy.

The patient refused immunophenotyping of the effusion. The peripheral blood film examination done simultaneously revealed 54% blasts with high nuclear: cytoplasmic (N:C) ratio. No Auer rods were seen (Fig. 2). Bone marrow aspirate and biopsy showed marked suppressed normal hematopoiesis with infiltration by blasts with high N:C ratio, open chromatin, 1-2 nucleoli and scant to moderate granular cytoplasm. Blasts were negative for myeloperoxidase (MPO) stain. Flowcytometry revealed the blasts were positive for CD34, cytoMPO and CD 33 (Fig. 3 and 4). Large atypical blast like cells were seen admixed with small lymphocytes on fine-needle aspiration (FNA) of the left supraclavicular lymphnode (Fig. 5). A final diagnosis of Acute Myeloid Leukemia M0 with leukemic pleural effusion and leukemic infiltration supraclavicular lymphnode was made.

DISCUSSION

Acute Myeloid Leukemia (AML) is a group of hematogenous neoplasms that are characterized by clonal proliferation of myeloid precursors with reduced capacity to differentiate into mature cells. This proliferation of immature cells in marrow leads to various systemic manifestations including anemia, bleeding, and an increased risk of infection [1].

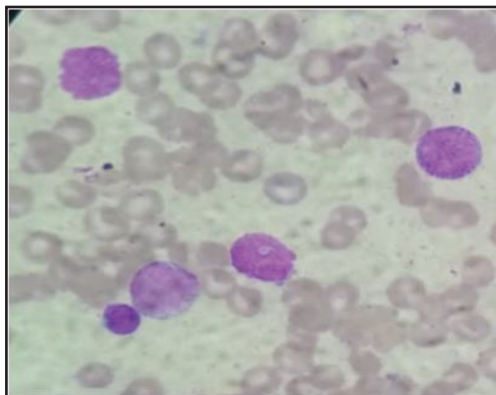


Figure 5: FNA cervical lymphnode; MGG; 1000x. Atypical lymphoid proliferation.

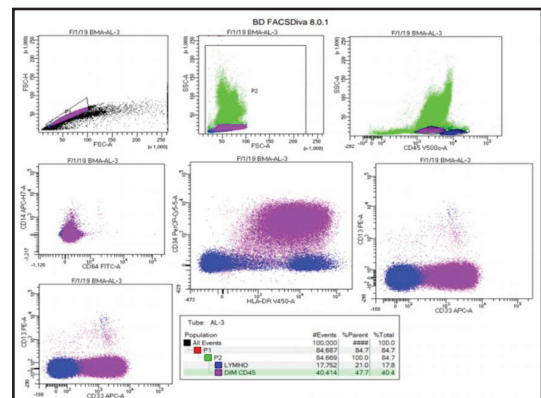


Figure 4: Flowcytometry bone marrow aspirate.

Effusion in AML can be due to various causes including infection, complication of chemotherapy and leukemic infiltration [3]. Leukemic pleural effusion is more commonly a manifestation of acute lymphoblastic leukemia and very few cases of non-lymphoblastic acute leukemia with effusion have been reported [4]. Malignant fluid collection is usually seen associated with solid tumors [5]. Recurrence of AML may occur at an extramedullary site including pleural effusion hence the importance of a correct assessment [6]. Extramedullary recurrence may be isolated without the involvement of the bone marrow [5].

Usually, pleural effusion responds to the treatment of the primary disease, whereas the resistant or the relapsing cases may necessitate a pleurodesis [7]. Faiz et al observed the presence of pleural effusion in 62.5% cases of acute leukemia and myelodysplastic syndromes in their study of which only 36% were malignant [8]. Boodosingh DR reported pleural effusion as the initial presenting complaint in a 66-year-old male, with submandibular, supraclavicular, axillary, and inguinal lymphadenopathy. Pleural fluid showed 62% leukemic myeloblasts [9].

There are limited studies on the role of molecular abnormalities in extramedullary disease in AML. Mutations of NPM-1 gene, FLT3-ITD, and 11q23 mutation have been associated with extramedullary disease manifestation [10].

CONCLUSION

The extramedullary disease is a rare manifestation of AML, of which leukemic pleural effusion is even rarer. These cases may be misdiagnosed as a lymphoproliferative disorder on routine examination especially if unaccompanied by bone marrow involvement. It is important to evaluate effusion with care and to have a high index of suspicion as a malignant pleural effusion may be the only presentation of recurrence of acute myeloid leukemia.

REFERENCES

- Jaffe ES, Harris NL, Stein H, Vardiman JW. World Health Organization of Tumors: Pathology and Genetics of Tumors of Hematopoietic and Lymphoid Tissue, Lyon 2001, France: IARC Press.
- Dores GM, Devesa SS, Curtis RE, Linet MS, Morton LM. Acute leukemia incidence and patient survival among children and adults in United States. *Blood*. 2012;119:34-43.

3. Bibbo M, Wilbur DC. Comprehensive Cytopathology. 3. Philadelphia: Saunders Elsevier; 2008. Pleural, peritoneal and pericardial effusions; p. 566.
4. Ganzel C, Manola J, Douer D, Rowe JM, Fernandez HF, Paietta EM, *et al.* Extramedullary disease in adult Acute Myeloid leukemia is common but lack independent significance: Analysis of patient MECOG-ACRIN Cancer Research Group Trial, 1980-2008. *J Clin Oncology*. 2016; 34:3544-53.
5. Alexarakis MG, Passam FH, Kyriakou DS, Bouros D. Pleural effusions in hematologic malignancies. *Chest*. 2004;125:1546-55.
6. Agrawal R. Acute myeloid leukaemia (AML) presenting as a bilateral pleural effusion. *J Clin Diagn Res*. 2013;7:187.
7. Raina S, Kaul R, Mahesh DM, Kaushal SS, Gupta D, Sharma J. Pleural effusions in acute myeloid leukaemia. *JAPI*. 2008;56:914.
8. Faiz SA, Bashoura L, Lei X, Sampat KR, Brown TC, Eapen GA, *et al.* Pleural effusions in patients with acute leukemia and myelodysplastic syndrome. *Leuk Lymphoma*. 2013;54:329-35.
9. Boodosingh DR, Hernandez LR, Fernandez R, Fernandez F, Velez F, Torrellas P, *et al.* Pleural effusion as the initial extramedullary manifestation of AML. D52 Case Reports AJRCCM Conference. 2012;185:A5901.
10. Mohammadiasl J, Khosravi A, Shahjahani M, Azizidoost S, Saki N. Molecular and cellular aspects of extramedullary manifestations of acute myeloid leukemia. *J Cancer Metastasis treat*. 2016;2:44-50.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Tandon A, Acharya S, Bhardwaj A, Kala P. Leukemic pleural effusion in AML-MO: unusual presentation. *Indian J Case Reports*. 2019;5(4):372-374.

Doi: 10.32677/IJCR.2019.v05.i04.026