Hematology clinicopathological exercise

Case Presentation

A 55-year-old male presented with hemoptysis and clubbing of fingers with no organomegaly. His full blood count revealed normochromic normocytic anemia (Hb: 8.6 g/dl), absolute neutrophilia (8.3 × 10^9/I), normal platelet count, and high erythrocyte sedimentation rate (110 mm/h). chest X-ray shows bilateral hilar lymphadenopathy, computed tomography (CT) scan of chest show diffuse ground glass appearance and reticulonodular shadow. Sputum for acid-fast bacillus was negative.

The patient was admitted to the hospital and given two pints of blood with trial of antituberculosis drugs.

After 2 weeks, the patient came with severe anemia and jaundice, splenomegaly, Hb: 5.4 g/dl, leukoerythroblastic blood picture, spherocytosis, reticulocyte count after correction with positive crankcase ventilation 11%, direct antiglobulin test positive, total serum bilirubin: 5 mmol/I, nearly equal fractions of direct and indirect types. Bone marrow aspiration findings are illustrated in Figure 1a and b.

- Describe the bone marrow findings and your provisional diagnosis?
- What other investigations should the patient send for to confirm the diagnosis?

Answers

- B. M slides show hypercellular marrow with erythroid hyperplasia, hemophagocytosis, and presence of nonhemopoietic cells in clusters and scattered throughout the marrow cells. The provisional diagnosis is nonhematopoietic metastatic cancer with hemophagocytic syndrome [Figure 2]
- Tumor markers, biopsy from suspected lesion (in this case via mediastinoscopy), serum ferritin, soluble CD25, serum triglycerides, plasma fibrinogen. CT neck, chest, and abdomen.

Hemophagocytic lymphohistiocytosis (HLH) represents a spectrum of hyperinflammatory disorders associated with activation of cytotoxic T- and natural killer (NK) cells, and macrophages. The excessive immune activation results in the clinical hallmarks of HLH, including fever, hepatosplenomegaly, and cytopenias, combined with a characteristic set of laboratory parameters (elevated ferritin, triglycerides, soluble CD25, transaminases, lactate dehydrogenase, d-dimers; decreased fibrinogen, albumin, and sodium).[1]

The triggers for HLH are infections such as Epstein–Barr virus, autoimmune diseases (rheumatoid arthritis or systemic lupus erythematosus), and malignancies. The proportion of each tumor type in adult patients with HLH in the context of a malignancy is reported to be 35% for T-cell or NK lymphomas, 32% for B-cell lymphomas, 6% for leukemias, 6% for Hodgkin lymphomas, 14% for other and nonspecified hematologic neoplasm, and 3% for not specified neoplasms.[2]

Solid tumors are not commonly associated with HLH with only 3% prevalence in adults.[2] In particular, mediastinal germ-cell tumors have been reported.[3]

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Conflicts of interest
There are no conflicts of interest.

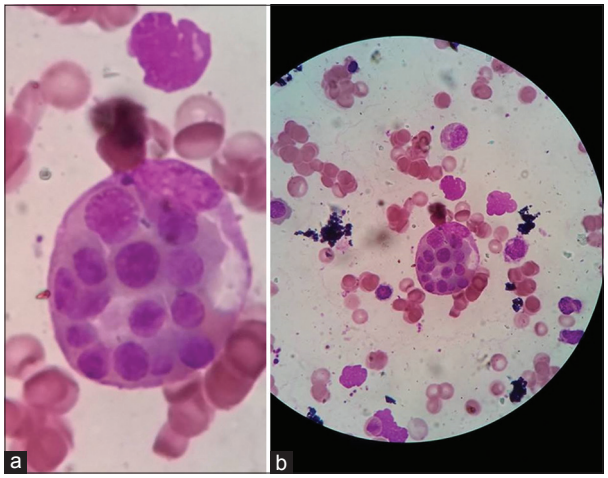

Figure 1: (a) Hemophagocytosis (b) clusters of nonhematopoietic cells

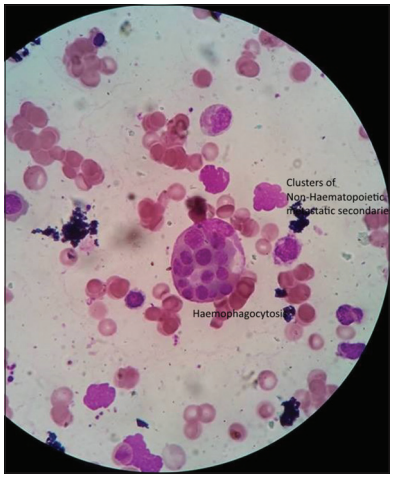

Figure 2: Nonhematopoietic metastatic cancer with hemophagocytic syndrome

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References

1. Lehmberg K, Nichols KE, Henter JI, Girschikofsky M, Greenwood T, Jordan M, *et al*. Consensus recommendations for the diagnosis and management of hemophagocytic lymphohistiocytosis associated with malignancies. Haematologica 2015;100:997-1004.
2. Ramos-Casals M, Brito-Zerón P, López-Guillermo A, Khamashta MA, Bosch X. Adult haemophagocytic syndrome. Lancet 2014;383:1503-16.
3. Nichols CR, Roth BJ, Heerema N, Griep J, Tricot G. Hematologic neoplasia associated with primary mediastinal germ-cell tumors. N Engl J Med 1990;322:1425-9.

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