ISSN : 2309-3234

TMSS Medical College Journal (TMCJ)

Volume 24, No. 01, January 2025

Contents

An Official Publication of TMSS Medical College

TMSS Medical College Journal (TMCJ)

January 2025

An Official Publication
of
TMSS Medical College, Bogura, Bangladesh

TMSS Medical College Journal (TMCJ)

Vol. 24, No 1, January 2025

An Official Publication of TMSS Medical College, Bogura, Bangladesh

The TMSS Medical College Journal (TMCJ) is published two times a year (January and July). The TMSS Medical College Journal (TMCJ) is approved by Bangladesh Medical & Dental Council (BM&DC/4-D-2016/1478: Dated 15.01.2017). It accepts original articles, review articles and case reports. While every effort is always made by the Editorial Board and the members of the Journal Committee to avoid inaccurate or misleading information appearing in the TMSS Medical College Journal, information within the individual article are the responsibility of its author(s). The TMSS Medical College Journal, its Editorial Board and Journal Committee accept no liability whatsoever for the consequences of any such inaccurate and misleading information, opinion or statement.

Printed by

TMSS Printing Press
Bogura, Bangladesh
e-mail: tmssprintingpress@gmail.com

Address of Correspondence

Editor-in-Chief, TMSS Medical College Journal, TMSS Medical College
Rangpur Road, Thengamara, Bogura, Bangladesh.
Phone: 051-61830, 01775-316617
e-mail: editortmcjournal@gmail.com, Web: www.tmcjournal.org, www.tmssmedicalcollege.com

Case Report

Thoracic Extramedullary Hematopoiesis in a 35- Year- Old Male: A Case Report

Dey MR[1*], Bondopadhay D[2]

1. Mano Ranjan Dey, Medical Student, TMSS Medical College, Bogura.
2. Disha Bondopadhay, Medical Student, TMSS Medical College, Bogura.

***Corresponding Author**

Abstract

Extramedullary hematopoiesis (EMH) refers to hematopoiesis that occurs outside the bone marrow. It may be physiological, like during fetal development, or may be pathological, like in hematopoietic disorder. The typical sites of EMH involve the liver,spleen,thorax and lymph nodes. In our case, the diagnosis was confirmed by the patient's history stating past hematological disorder, computed tomography (CT) scan findings and by fine needle aspiration biopsy (FNAB) test.

Keywords: Extramedullary hematopoiesis, Thalassemia, Fine needle aspiration biopsy, Computed tomography scan.

Introduction

Hematopoiesis- Hemato (blood) and poiesis (formation) is the process in which all blood cells (Red blood cells, white blood cells and platelets) are formed. In a healthy person, approximately 10 billion new blood cells are Produced Per day in the body to maintain blood cells. During embryonic life, the yolk sac is the main site of hematopoiesis from the 3rd week of gestation. The liver and Spleen are the main sites of hematopoiesis from the 3rd month of embryonic life till birth. By the 4th and 5th month of embryonic life bone marrow will also Start Producing blood cells. After birth, the liver and Spleen stop producing blood cells, and bone marrow is the only source of new blood cell formation. In normal healthy adult hematopoiesis occurs from the bone marrow is termed as medullary hematopoiesis.[1] If hematopoiesis occurs other than in the bone marrow, this is termed extramedullary hematopoiesis. It is an abnormal phenomenon. Initially, it is microscopic and asymptomatic, but when it enlarges sufficiently, it produces a tumor-like mass. It is typically seen in the spleen, liver, and lymph nodes, but also can be seen in some atypical sites like adipose tissue, breast, adrenal gland, kidney, thymus, heart, peripheral nerve, lung, cartilage, broad ligament, retroperitoneum, epidural space, and epididymis, and some soft tissues of the body.[2, 3] In this report, we discuss the case of a 35-year-old male who presented to the OPD with exertional dyspnea. After considering its radiological findings, we diagnosed it as a benign neurogenic tumor however later, we performed a fine needle aspiration biopsy for confirmation, and we finally diagnosed it as an EMH.

Case presentation

A 35-year-old pale Bangladeshi male presented to the local hospital with exertional dyspnea. He denied any history of cough, hemoptysis, chest pain, fever, chills, night sweats or weight loss.

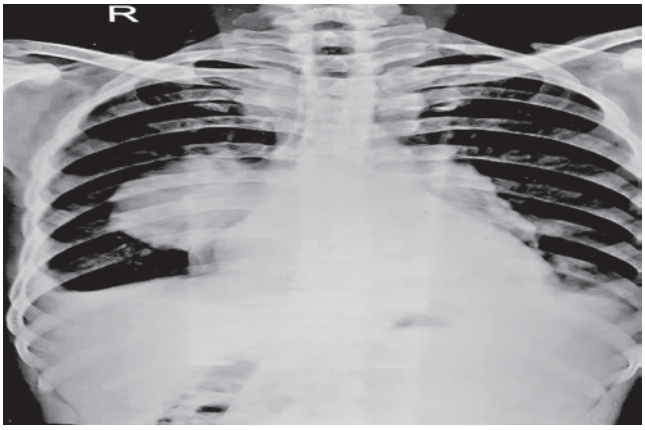

Figure-1: X-Ray of The Chest PA View

Initial workup reveals a lung space-occupying lesion on X-ray(Figure-1).

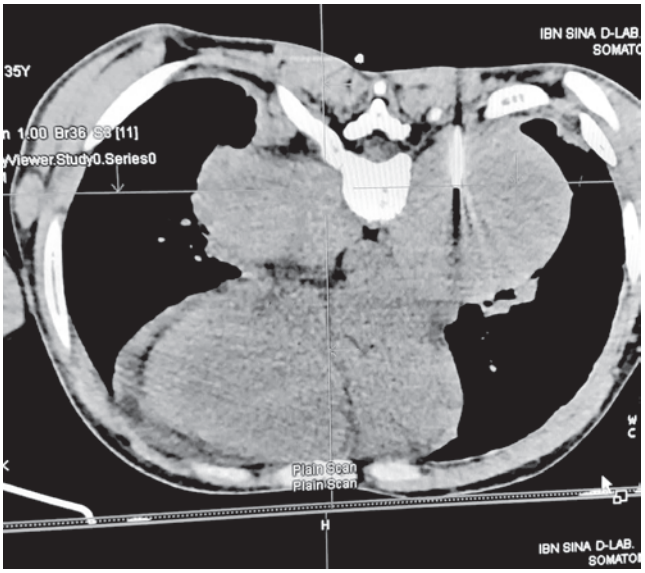

Figure-2: Contrast-enhanced axial CT of the chest

Computed tomography (CT) revealed a huge bilateral posterior mediastinal solid mass (48HU) measuring 80x76x75mm on the right side and 66x54x52mm on the left side. It was considered benign such as neurogenic tumors (Figure-2).

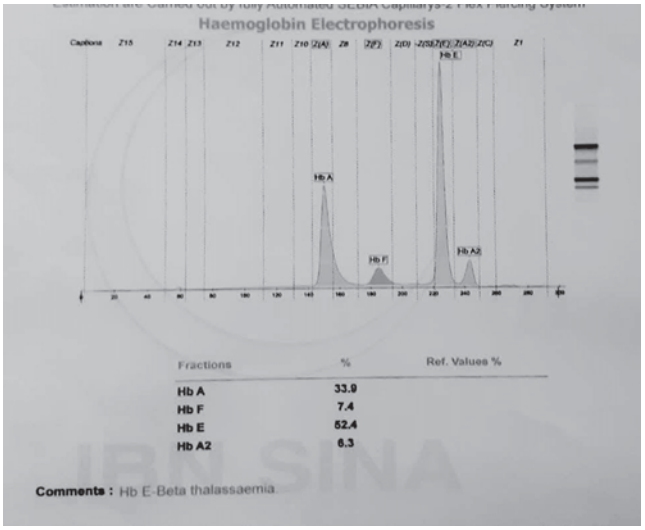

Figure-3: Hemoglobin Electrophorosis

Past medical history reveals he is a case of transfusion-dependent hemoglobin E-Beta thalassemia which was diagnosed by hemoglobin electrophoresis (Figure-3).

On physical examination, he was afebrile and had a blood pressure of 137/64 mmHg, a heart rate of 75 beats/min, and a oxygen saturation of 96% on ambient air. Ultrasonogram reveals mild hepato- spleenomegaly.

After admission, relevant laboratory examination was completed. White blood cell count 6,700/cmm, Hemoglobin 6.2 g/dl, Red blood cell count 3.99 m/µl, Hematocrit 21%, Mean corpuscular volume 51.0fL, Mean corpuscular hemoglobin 14.0pg, Mean corpuscular hemoglobin concentration 27.0g/dl. Other laboratory tests were within normal limits.

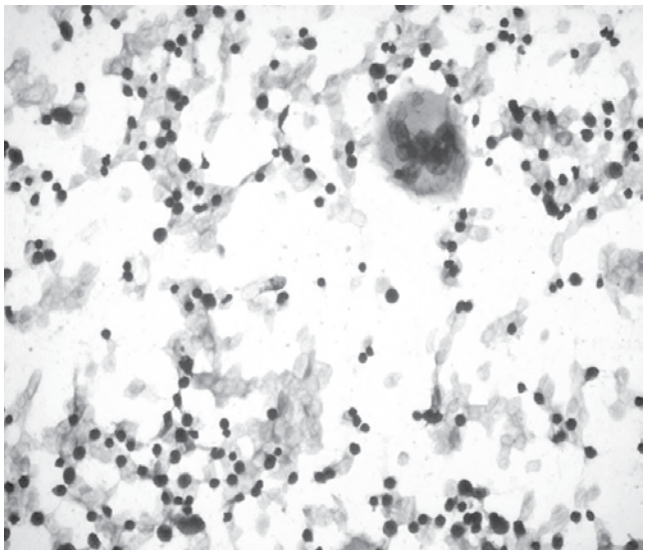

Figure: 4

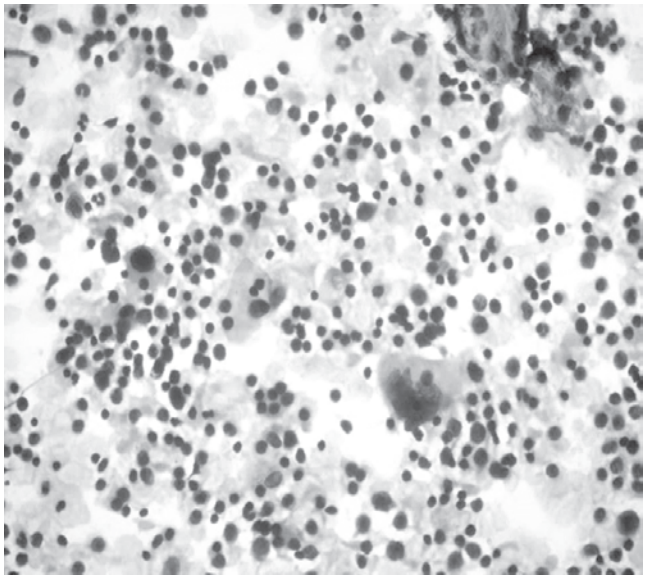

Figure:5

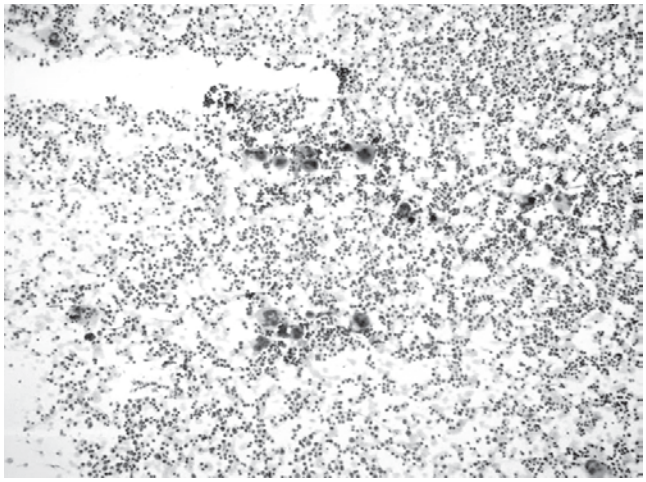

Figure: 6

The Fine needle aspiration biopsy (FNAB) specimen revealed cellular marrow elements with hematopoietic cells composed of megakaryocytes, erythroblasts, and myeloid cells. Finally, he was diagnosed with extramedullary hematopoiesis (EMH) (Figure-4,5,6).

Discussion

According to the hypothesis, extramedullary hematopoiesis occurs as a compensatory mechanism, when the bone marrow is unable to maintain the rate of erythrocyte formation, or erythrocytes are removed and destroyed rapidly.[4] It tends to show generally male dominance at the age of 28 to 74, with a male-to-female ratio of 5:1. Patients with thalassemia intermedia this incidence of extramedullary haematopoiesis have high risk of 20%, patient with transfusion dependent thalassemia (TDT) the risk is about 1%. Then 11–15% of individuals experiencing extramedullary hematopoiesis show a paravertebral location of hematopoietic tissue.[5, 6] Extramedullary hematopoiesis may be found in some pathological conditions such as thalassemia, Spherocytic anemia, Erythroblastosis of newborn, Hodgkin's disease, Myelofibrosis, Carcinoma with bone marrow invasion, Sickel cell anemia, carcinomatosis, Myeloproliferative syndrome, increased renal release of erythropoietin, Escape of progenitor cells from marrow and their lodging into different organs.[2] Presence of intra-thoracic extramedullary hematopoiesis is very rare. When it occurs, it is usually observed as round, smooth, lobulated, with dense soft parts in the posterior mediastinum, more common in the lower paravertebral regions but sometimes it is also noticed in the anterior mediastinum and the pleura.[7- 10]

More than 80% of the patients show asymptomatic findings that are accidentally detected by radiology or the patient presents with complications of chylothorax, hemothorax and pneumothorax and neurologic symptoms like sensory impairment, paraparesis, quadriplegia and occasionally sphincter disturbances, most frequently being mentioned.[11,12]

Previous reports of tumor-like masses produced by intrathoracic extramedullary hematopoiesis were based on autopsy findings, recent reports of the antemortem diagnosis of this condition have been presented.[3]

Diagnosis of intrathoracic extramedullary hematopoiesis is usually done by the history of hematological disease, x-ray of the chest, computerized tomography scan of the chest, Magnetic resonance imaging is considered to be the best technique for the diagnosis, non-invasive methods also used for diagnosis like 18F-fluoro- 3′deoxy- L-thymidine PET/CT and 99mTc-SC scintigraphy and invasive procedures such as needle aspiration biopsy, bronchoscopy with transbronchial biopsy, endobronchial ultrasound-guided fine needle aspiration of the tissue mass and mediastinoscopy, medical thoracoscopy and surgical techniques like median sternotomy, video-assisted thoracoscopic surgery and thoracotomy. Invasive methods are not that much necessary and are potentially dangerous because of the highly vascular consistency of extramedullary hematopoiesis.[13-23].In this case, we diagnosed it by considering its past medical history and confirmed it by performing a Fine Needle Aspiration Biopsy test.

Treatment is suggested when there is an occurrence of complications. The surgery is not recommended because of the recurrence of the masses and the diffuse nature of the process. In our case we combine blood transfusions, corticosteroids and hydroxyurea

administration, which are the good treatment option for the inhibition of haematopoiesis and to prevent the recurrence.[24,17]

However it has poor prognosis when it recurs or invades other organs and ultimately it may cause death of the patients by producing some serious complications like pulmonary embolism, conversion to acute leukemia, renal failure, and cerebral infarction.[25]

Differential diagnosis of intra-thoracic extramedullary hematopoiesis includes Neurogenic tumors which is are the most common cause of a posterior mediastinal mass, Hodgkin's disease usually located in the middle or anterior mediastinum, lymphomatous masses are sometimes present predominantly in the paraspinal region of the thorax, Abscess arising from the spine here no lobulation is present, Extrapleural cysts present with single arc at their anterior and lateral margins, Primary and metastatic malignancies do not show a lobulation and show bone involvement, Intrathoracic meningocele presents as a single, occasionally bilateral, round density and Localized mediastinal lymph node hyperplasia.[26,27]

Conclusion

EMH is a rare cause of intrathoracic mass formation, and it's a challenge to differentiate from a benign tumor. Extramedullary hematopoiesis should be considered in the differential diagnosis of a posterior mediastinal mass, particularly in a thalassaemia patient. Use of CT and FNAB provides detailed information for evaluation of EMH and a follow-up study is helpful.

References

1. Guyton and hall text book of medical physiology,13th edition,chapter-33
2. Smith PR, Manjoney DL, Teitcher JB, Choi KN, Braverman AS. Massive hemothorax due to intrathoracic extramedullary hematopoiesis in a patient with thalassemia intermedia. Chest. 1988 Sep 1;94(3):658-60.
3. Parsons Jr L, Thompson JE. Symptomatic myelolipoma of the adrenal gland: report of a case and review of the literature. New England Journal of Medicine. 1959 Jan 1;260(1):12-5.
4. O'Malley DP. Benign extramedullary myeloid proliferations. Modern Pathology. 2007 Apr 1;20(4):405-15.
5. Haidar R, Mhaidli H, Taher AT. Paraspinal extramedullary hematopoiesis in patients with thalassemia intermedia. European Spine Journal. 2010 Jun;19:871-8.
6. Subahi EA, Ata F, Choudry H, Iqbal P, AlHiyari MA, Soliman AT, De Sanctis V, Yassin MA. Extramedullary haematopoiesis in patients with transfusion dependent β-thalassaemia (TDT): a systematic review. Annals of Medicine. 2022 Dec 31;54(1):764-74.
7. Yamato M, Fuhrman CR. Computed tomography of fatty replacement in extramedullary hematopoiesis. Journal of computer assisted tomography. 1987 May 1;11(3):541-2.
8. Pornsuriyasak P, Suwatanapongched T, Wangsuppasawad N, Ngodngamthaweesuk M, Angchaisuksiri P. Massive hemothorax in a beta-thalassemic patient due to spontaneous rupture of extramedullary hematopoietic masses: diagnosis and successful treatment. Respiratory care. 2006 Mar 1;51(3):272-6.
9. Wyatt SH, Fishman EK. Diffuse pulmonary extramedullary hematopoiesis in a patient with myelofibrosis: CT findings. Journal of computer assisted tomography. 1994 Sep 1;18(5):815-7.
10. Fielding JR, Owens M, Naimark A. Intrathoracic extramedullary hematopoiesis secondary to B12 and folate deficiency: CT appearance. Journal of computer assisted tomography. 1991 Mar 1;15(2):308-10.
11. Papakala E, Porpodis K, Fouka E, Lagoudi K, Efthymiou C, Papakosta D. Pleural effusion as the main manifestation of intrathoracic extramedullary heamatopoietic masses in a patient with beta thalassemia minor. www. pneumon. org. 2016 Apr 1;29(2):156-60.
12. Kubokura H, Koizumi K, Yoshino N, Yamagishi S, Mikami I, Hirata T, Harada A, Kawamoto M, Shimizu K. A case report: thoracic

extramedullary hematopoiesis found by occurring spontaneous pneumothorax. Ann Thorac Cardiovasc Surg. 2008 Dec 1;14(6): 382-5.

13. Monville Jf, Fillet G, Ghaye B. Thoracic Extramedullary Hematopoiesis Secondary To Enzy-matic Deficiency. Jbr–btr. 2007 May 1; 90: 198-9.
14. Hassoun H, Lawn-Tsao L, Langevin Jr ER, Lathi ES, Palek J. Spinal cord compression secondary to extramedullary hematopoiesis: a noninvasive management based on MRI. American journal of hematology. 1991 Jul;37(3):201-3.
15. Zade A, Purandare N, Rangarajan V, Shah S, Agrawal A, Ashish J, Kulkarni M. Noninvasive Approaches to Diagnose Intrathoracic Extramedullary Hematopoiesis: 18: F-FLT PET/CT and: 99m: Tc-SC SPECT/CT Scintigraphy. Clinical Nuclear Medicine. 2012 Aug 1;37(8):788-9.
16. Özdemir A, Güngör F, Tunçdemir F, Çubuk M, Karayalçin B. Scintigraphic diagnosis of intrathoracic extramedullary hematopoiesis in a patient with β-thalassemia. Annals of nuclear medicine. 1998 Jun;12:149-51.
17. Ketata W, Msaad S, Kwass H, Gargouri I, Gheriani MM, Gouiaa N, Dabbech C, Boudawara T, Ayoub A. Thoracic extramedullary hematopoiesis: A diagnosis to not forget in a patient with posterior mediastinal mass with anemia. Respiratory Medicine CME. 2009 Jan 1;2(3):118-20.
18. Yusen RD, Kollef MH. Acute respiratory failure due to extramedullary hematopoiesis. Chest. 1995 Oct 1;108(4):1170-2.
19. Ha D, Liao X, Wang HY, Jamieson C, Magaña M, Makani S. Thoracic extramedullary hematopoiesis mimicking metastatic cancer. Journal of Bronchology & Interventional Pulmonology. 2016 Oct 1;23(4):343-6.
20. Da Costa JL, Loh YS, Hanam E. Extramedullary hemopoiesis with multiple tumor-simulating mediastinal masses in hemoglobin E-thalassemia disease. Chest. 1974 Feb 1;65(2):210-2.
21. Murthy V, Bessich JL. Medical thoracoscopy and its evolving role in the diagnosis and treatment of pleural disease. Journal of thoracic disease. 2017 Sep;9(Suppl 10):S1011.
22. Luciani GB, Lucchese G. Minimal-access median sternotomy for aortic valve replacement. Journal of Thoracic Disease. 2013 Nov;5(Suppl 6):S650.
23. Yao WQ, Wang WY, Liu XJ, Gao LM, Wang Z, Xiang CX, Tao Q, Zhang WY, Liu WP. Mediastinal myelolipoma/extramedullary hematopoiesis presenting as a mass: rare differential diagnosis among mediastinal tumors. International Journal of Clinical and Experimental Pathology. 2018;11(5):2714.
24. Russo D, Pileri S, Barbieri E, Bandini G, Zaccaria A, Benfenati D, Bernardi B, Trevisan C, Busacca M, Babini L. Spinal cord compression by extramedullary hematopoietic tissue in a thalassemic patient: prompt effect of radiotherapy. Haematologica. 1989 Sep 1;74(5): 495-8.
25. Koch CA, Li CY, Mesa RA, Tefferi A. Nonhepatosplenic extramedullary hematopoiesis: associated diseases, pathology, clinical course, and treatment. InMayo Clinic Proceedings 2003 Oct 1 (Vol. 78, No. 10, pp. 1223-1233). Elsevier.
26. Yang X, Chen D, Long H, Zhu B. The mechanisms of pathological extramedullary hematopoiesis in diseases. Cellular and Molecular Life Sciences. 2020 Jul;77:2723-38.
27. Sohawon D, Lau KK, Lau T, Bowden DK. Extra-medullary haematopoiesis: a pictorial review of its typical and atypical locations. Journal of medical imaging and radiation oncology. 2012 Oct;56(5):538-44.