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Original Research Paper

Haemothorax in Thalassaemia May be a Complication of Rupture of Intrathoracic Extramedullary Haematopoiesis

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Abstract-
Intrathoracic extramedullary hematopoiesis is reported in the literature but rare; typically found in the chronic hemolytic anemia. We report a beta-thalassaemia major patient, 11-y-girl, presented with hemothorax. Computed tomographic scan of the thorax showed many paravertebral and extra-pulmonary tumors. Biopsy revealed extramedullary haematopoiesis. Hemothorax in beta thalssemia should be suspicion about Intrathoracic extramedullary hematopoiesis because surgical intervention is first-line management.

Keywords*: extramedullary haematopoiesis; haemothorax; thalassaemia*

Introduction

Extramedullary hematopoiesis (EMH) is production of normal blood cells outside of the bone marrow. It is chiefly a compensatory mechanism for chronic hemolytic diseases but also found malignant or fibrotic diseases of bone marrow[1,2]. The EMH affect mainly the spleen, liver, and lymph nodes and, less commonly retroperitoneal, retropleural sites, pleura, and peritoneum.

Intrathoracic EMH is common in thalassemia, because irregular transfusion fails to suppress erythropoiesis. That EMH is small nodule in posterior mediastinal along paravertebral areas, usually asymptomatic plural effusion(1–3)

Hemothorax is caused by trauma, tuberculosis, malignancy. But rapture of EMH can raise hemothorax - reported in literature.(4)

Case report

An 11-year-old girl presented in emergency department with dyspnoea which has been progressive since three days. There was no history of trauma or fever with cough. She was diagnosed as beta–thalassaemiamajor by haemoglobin electrophoresis at age of eight months. She was on a suboptimal blood transfusion program in her local town. She was started on desferoamine six months prior to admission in this institute. On examination, she had a hemolytic facies, weight and height both below 3rd percentile for the age. Physical examination revealed a blood pressure of 106/66 mm Hg, pulse rate of 95/min, pale conjunctiva, icteric sclera, diminished left sided breathing sounds, and marked hepatosplenom-egaly. Initial haematological examination showed a haemoglobin level of 7 g/dl, haematocrit value of 22.1%, mean corpuscular volume of 70fl,meancorpuscular haemoglobin concentration of 22 g/dl, red blood cell count of 3.1 × 10 6/mm^3, white blood cell count of 16,400/mm3 and platelet count of 4,20,000/mm^3

Chest radiography showed homogeneous opacity of left hemithorax. Thoracocentesis revealed a bloody effusion with a protein level of 6gm/dl, sugar 5 mg/dl, red blood cell count 1.8×10^6/mm3 , white blood cell count 2 × 10^4/mm3 and negative cytological results.And haematocrit of Pleural fluid to Blood haematocrit Ratio was more than 55%. Culture was negative. Three times Cell-block examination were no malignant cell found. Montoux test and AFB for sputum excludes tuberculosis.

Computed tomographic scanning of the chest revealed many extrapulmonary paravertbral tumors beside the thoracic spine. A CT-guidedbiopsy of the mass showed normal haematopoietic tissue with normal maturation.

Repeated CT-guided aspiration of hemothorax was done, fluid was accumulating very fast. Tube thoracostomy was approached, approximately total 4 liters was collected. Repeated Blood transfusions was given. Hydroxyurea was started. Patient was transferred to Department Cardiothoracic Surgery for further management. Unfortunately patient could not survive due to nosocomial infection

Discussion

EMH is an appearance of hematopoietic elements outside of the bone marrow.EMH is a known complication of diseases in which there is an encroachment of marrow spaces or a chronic increase in rate of production of Red Blood Cells[6]. It is because of a compensatory bone marrow hyperplastic condition, with seeding of circulating hematopoietic progenitors at various sites.

EMH is commonly found in hemoglobinopathies like thalassemias, hereditary spherocytosis [1-5]. In addition, neoplastic disease like chronic myeloproliferative disorders [6] or leukemias [2] may produce EMH . It may appear in two sites[7]: (a)Paraosseous - rupture of normal bone marrow from bones, commonly in hemoglobinopathies;

(b)Extraosseous- blood cell production in soft tissues, mainly in myeloproliferative disorders. Intrathoracic EMH most commonly occurs in posteroinferior mediastinum areas, but may be found in anterior mediastinom and pleura [8]. Pleural involvement is usually asymptomatic and microscopic and is noted only at autopsy [8]. Pleural effusion due to pleural involvement by EMH is rarely reported and underlying cause is myleofibrosis [1,3,8]

The preference of EMH for the intrathoracic site is not clearly understood; a possible explanation is that the negative pressure at that site could facilitate the extrusion of hyperplastic hematopoietic tissue from the vertebral marrow [9]. In the case of mediastinal EMH, the mediastinal masses present on plain thorax X-ray as well demarcated and lobulated masses in the posterior mediastinum, usually below the level of T6. It may be unilateral or bilateral, do not contain calcification, and are not associated with bone destruction [6].CT studies of EMH have described homogenous, well demarcated soft tissue masses

usually in the paravertebral region, typically absence of calcification and bone destruction [3].

The differential diagnosis of posterior mediastinal tumor are considered in our case. Those are neurogenic tumor, lymphoma, abscess. Index patient had posterior mediastinal masses without evidence of calcification or bone destruction on CT scanand characteristics lobulation are the main radiological features to distinguish EMH from neurogenic tumorsin the posterior mediastinum [10]. EMH may also be detected by radionuclide scanning by $^{99}Tc^{m}$ sulfur colloid or ^{111}Indium chloride [6]. Whatever radiological technique is used to diagnose the EMH , biopsy confirmation is must. Although invasive diagnostic procedures have been used, including thoracotomy and CT guided needle aspiration biopsy, they are potentially hazardous because of highly vascular nature of the masses, and so an increased risk of bleeding [10].

The first-line of treatment of fatal EMH ruptured hemothorax is surgical intervention.[3]. Surgery is also advocated for clot extraction, compressive mass debulking , diagnostic purposes.[8] Repeated aspiration or tube thoracostomy and repeated transfusion may be an alternative for hemothorax. When thoracic surgery is avoided due to increased risk of bleeding. In these cases, the treatment of choice is radiotherapy [8].

Treatment of patients with EMH is only required in the presence of complications. EMH tissue is highly radiosensitive and relatively small doses of radiation are effective[1,10]. Regression of asymptomatic EMH can have done by transfusion, chelation therapy , and hydroxyurea [11]. Although Pleurodesis may be satisfactory therapy in EMH related Pleural effusion,[3] pleural sclerosing agents is sometimes contraindicated because these agents can worsen bleeding [8].

In our country where thalassemia is a very common cause of chronic hemolytic anemia, which is often inadequately treated, ineffective erythropoiesis and extramedullary hematopoiesis are very common. Therefore a child with thalassemia presenting with hemothorax may have underlying intrathoracic extramedullary hematopoiesis. This possibility has to be kept in mind while elucidating a differential diagnosis of hemothorax in such a patient, apart from the common entities such as tuberculosis and malignancy. And management is importantly surgical.

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