Case Report

Paravertebral Extramedullary Hematopoiesis In A Pregnant Thalassemic Patient: A Case Report

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ABSTRACT

Spinal cord compression due to extramedullary hematopoiesis is a well-described and rare syndrome encountered in several hematological disorders including β-thalassemia. Hereby, a 37-year old pregnant woman with intermediate β -thalassemia with paraparesis and lower limb hypoesthesia was presented. MRI showed soft tissue masses in both sides of thoracic paraspinal area. Histologically, a mixture of all hematopoietic cell lines was present at different stages of maturation. Medical literature is also reviewed in this report.

Key words: **Extramedullary hematopoiesis, Intermediate thalassemia, Paraspinal mass**

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Introduction

Intrathoracic tumors usually suggest of a malignant process in the first instance. In a patient with hematological problems, extramedullary hematopiesis (EMH) is a high possibility (1). EMH is a compensatory phenomenon that occurs in patients with hematological disorders when bone marrow function is not sufficient to maintain the circulatory demand (2). EMH accompanies a wide variety of diseases including thalassemia, sickle cell anemia, polycytemia rubra vera, chronic myelogenous leukemia, agnogenic myeloid metaplasia, and hereditary spherocytosis (3). The most common sites of EMH are organs that have physiological hematopoiesis during emberyonic life, especially liver and spleen. Other reported sites of diffuse compensatory EMH include lymph nodes, adrenal glands, kidney, breast, dura mater, adipose tissue, and skin (2, 4). EMH causing spinal cord compression is a rare manifestation of thalassemia during pregnancy with one previously reported case in a 24-year old thalassemic woman from Thailand presenting with paraparesis at second trimester of pregnancy (5). The only previously reported case in Iran was in a non-pregnant woman (6).

Case Report

A 37-year old pregnant woman (24 weeks) and a known case of intermediate β-thalassemia referred with paraparesis and lower limb hypoesthesia which started from 5 months ago and worsened 1 month before admission. No urinary incontinence was present. In her past medical history, splenectomy was done 17 years ago. She had got no blood transfusions. The only

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medication she was taking was Desferal. On physical examination, upper limb forces were 5/5 and lower limb forces were 2/5 and 3/5 in proximal and distal parts respectively. Hypoesthesia was also present in both lower limbs. Babinski reflex was also flexor at both sides. Other organs were normal on physical exam. Hemoglobin was 9.4 g/dl (normal range:12-14 g/dl) on admission. MRI of thoracic spine without Gadolinium injection (sagittal T1, T2, axial T2) revealed soft tissue masses on both sides of the thoracic spinal area at T4-T9 level (Figure 1). All discs were normal in signal intensity without herniation. Bony spinal canal was normal in alignment and size without abnormal signals. Extramedullary hematopoiesis was radiologically suggested according to patient's history. She underwent laminectomy and intraoperative pathology consultation was done which showed heterogeneous population of hematopoietic precursor cells including erythroid, myeloid, and megakaryocytic series (Figure 2). She was discharged in good condition two weeks after operation with Desferal mesylate 1000 mg twice weekly.

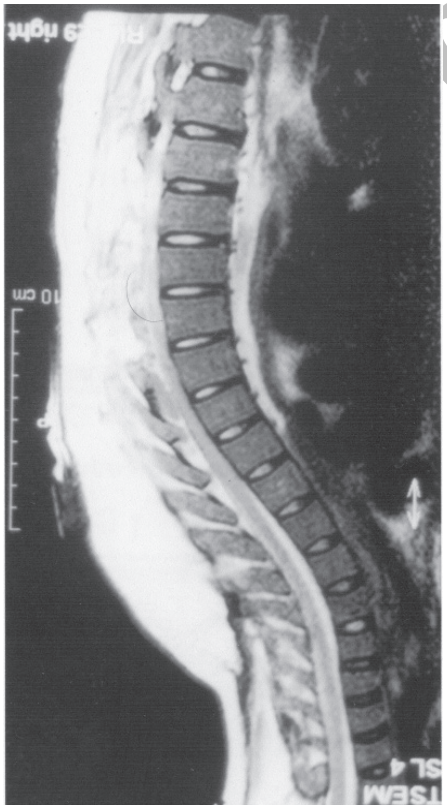

Figure 1: MRI (sagittal view) showing paravertebral soft tissue mass involving T4-T9

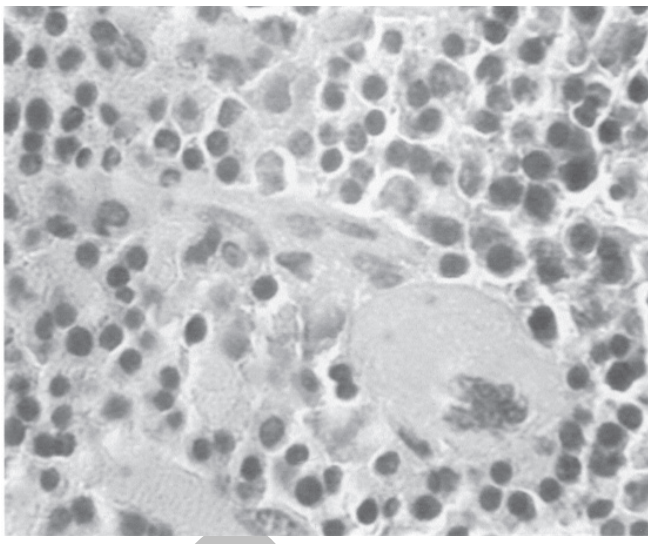

Figure 2: Light microscopy showing a mixture of hematopoietic cells (H&E, x 400)

Discussion

Bone marrow and extramedullary sites attempt to increase the production of erythrocytes to compensate the chronic hemolytic anemia in individuals with thalassemia. EMH is commonly seen at sites such as abdomen, chest and/or epidural space (3, 7). There are two forms of EMH, namely, para-osseous in which the normal medullary tissue of the bone marrow ruptures through the bone to present as a para-osseous mass and extra-osseous in which it occurs within the soft tissue. Para-osseous EMH occurs more frequently in hemoglobinopathies whereas extra-osseous type predominantly accompanies with myeloproliferative disorders (4). Patient with intermediate thalassemia are not usually treated with regular transfusion therapy as the medullary and EM hematopoiesis are able to maintain hemoglobin concentration above a reasonable level (3). However, in conditions when the demand increases, such as in pregnancy, new sites of EMH are produced. Our patient's symptoms started during pregnancy, which may be due to the above reason.

First description of spinal cord compression by EMH dates back to 1954 (8). Since then, about 60 cases have been reported, mainly with intermediate β-thalassemia. This complication has mainly been observed in the thoracic segment of spinal cord,

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as our case did, but the reason for this predilection remains uncertain (1, 2, 5-7, 9, 10).

MRI is the imaging modality of choice for the primary diagnosis of EMH (10). Surgical decompression has been the method of choice for the management of the disease because the histological diagnosis can be established and immediate decompression can be achieved (6). Because of some disadvantages of surgical intervention including excessive bleeding, a higher incidence of recurrence and clinical decompensation and deterioration, incomplete resection and low doses of radiation bring about a good clinical response (3). Repeated blood transfusion until delivery is another treatment choice in pregnant women (5). In our patient, surgical removal of the lesions improved her condition and no radiation was given.

Differential diagnosis of the paraspinal location of EMH includes tumors, lymphoma, paravertebral abscess and metastatic carcinoma (3). Histological diagnosis of EMH is made by the presence of heterogeneous population of erythroid and myeloid precursors and megakaryocytes (11, 12). Megakaryocytes can be confused with Reed-Stenberg cells of Hodgkin's lymphoma, especially in frozen sections, though formers are greater in size, multinucleated, and devoid of nucleoli. Furthermore, the cytoplasm of megakaryocytes is strongly periodic acid Schiff (PAS) positive. Immunohistochemically, megakaryocytes are reactive for factor VIII-related antigen, whereas Reed-Sternberg cells are reactive for CD30 and CD15 (12). In geographic areas where thalassemia is prevalent, EMH should be considered in differential diagnosis of patients who have chronic anemia with a paraspinal mass (3). Combination of pathologic features and patient's history help the pathologist to avoid incorrect diagnosis. This case has been the second reported one of spinal cord compression due to EMH in thalassemia and the first pregnancy-associated case in Iran (6).

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