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CASE REPORT

Thoracic cord compression by extramedullary hematopoiesis in thalassemia

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ABSTRACT

Extramedullary hematopoiesis (EMH) refers to hematopoiesis occurring outside the medulla of bone. It may be physiologic or due to pathological conditions like hematopoietic disorders. EMH can involve liver, spleen, thorax, and lymph nodes. It can involve paraspinal tissues with extension and involvement of spinal canal. In our case, the diagnosis was confirmed by the history of the patient stating underlying hematological condition and by magnetic resonance imaging (MRI) findings showing large soft tissue masses in paraspinal areas with involvement of spinal canal and leading to cord compression.

Key words: Extramedullary hematopoiesis, magnetic resonance imaging, thalassemias

Introduction

Extramedullary hematopoiesis (EMH) refers to hematopoiesis occurring outside the medulla of bone. It may be physiologic, for example, during fetal development where it occurs in many different locations such as the liver. However, it is more frequently associated with pathologic processes. For example, it can be caused by disorders like thalassemias, polycythemia rubra vera, myelofibrosis, hemolytic anemia, and other hemoglobinopathies. EMH can involve liver, spleen, thorax, and lymph nodes. It can involve paraspinal tissues with extension and involvement of spinal canal. In our case, the diagnosis was confirmed by the history of the patient, underlying hematological condition, and by magnetic resonance imaging (MRI) findings showing large soft tissue masses in paraspinal areas with involvement of spinal canal and leading to cord compression.

Case Report

A 28-year-old male diagnosed with Beta thalassemia, presented with back pain and weakness of both lower limbs, which had progressed in recent weeks. On examination, he was paraparetic with muscle strength decreased in both lower limbs. Deep tendon reflexes were exaggerated in both lower limbs. Planter response was extensor on both sides. Liver and spleen was enlarged and his higher mental functions and cranial nerves were normal. On hemoglobin electrophoresis, significant percentage of fetal hemoglobin (HbF) was present. MRI of dorsal spine showed lobulated mass lesion not showing any calcification in paravertebral regions on both sides [Figures 1, 2 and 3]. Extradural mass lesion is also seen involving posterior portion of spinal canal from D4 till D8 level [Figure 4] and is causing pressure effect on dorsal cord with post compressive cord signal changes. Our patient did well with radio-therapy, and the power in limbs gradually increased.

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Discussion

EMH is a recognized process which occurs, in chronic anemias in which the body attempts to maintained erythrogenesis. Common site of EMH are spleen, liver, kidney, lymph nodes, or in paravertebral regions.[1] Number of conditions like thalassemia, polycythemia, myelofibrosis, hemolytic anemia, and other hemoglobinopathies leads to such conditions. Gatto in 1954 was the first to report spinal cord compression by EMH.[2] Cases can present with paraparesis, sensory impairment, and occasionally sphincter disturbances. Paraplegia or quadriplegia may ensue.[3] MRI is the diagnostic procedure which shows isointense lobulated mass lesions seen in both T_1 and T_2 weighted imaging involving extradural portion of spinal canal and in paravertebral portions. Most authors do not favor a tissue biopsy in this situation as the lobulated mass with history of hemoglobinopathies is certain for diagnosis.[4] Hemorrhagic signal changes can be seen there.[5]

Figure 1: Coronal T2 weighted images is showing lobulated soft tissue lesion in paravertebral portions of dorsal spine suggestive of extramedullary hematopoiesis

Figure 2: T2 weighted axial images are showing lobulated mass in paravertebral portion with cord compression at D5 level

Figure 3: T2 weighted axial images are showing lobulated mass in paravertebral portion like fig 4 with cord compression at D7-8 level

Figure 4: Saggital T2 weighted images is showing well marginated soft tissue mass lesion involving posterior portion of spinal extradural in location and is causing pressure effect on dorsal cord which is showing post compressive signal changes

EMH is a compensatory mechanism to sustain erythrogenesis. According to Abbassioun *et al*., intrathoracic EMH is either caused by the direct spread of paravertebral localized activated precursor cells producing a paravertebral mass lesion or by the existence of hematopoietic precursor cells in the extradural area locally which later expand under hematological stress in chronic anemia. Rare complication of cord compression due to EMH has been reported with patient with thalassemia,[6] myelofibrosis, sickle cell anemia, and polycythemia vera. In our case, cord compression was because of EMH due to thalassemia. Intrathoracic extramedullary hematopoiesis is generally localized at the posterior mediastinum, and the middle and lower paravertebral areas. The mechanism of spinal cord compression at this site is the localization of the mass lesion as well as limited mobility of the spinal cord at the same localization. The signal characteristics of these lesions on MRI suggest the presence of blood/iron products. Iron in ferrous and ferric state is paramagnetic and can shorten T_1 and T_2 relaxation. As a result, signal intensity in both T_1 and T_2 of adjacent soft tissues can change depending upon whether there is presence of deoxyhemoglobin, methhemoglobin, hemosiderin, or a combination of above. Decrease in T_2 relaxation time will lead to decrease intensity in T_2 weighted imaging. Treatment option of EMH can be radiotherapy or surgical procedure. Advantage of use of radiation therapy includes the avoidance of surgical procedure,[7] but tissue biopsy cannot be done. Radiation may produce bone marrow suppuration in already anemic patient.[8] In our case, patient was still anemic, requiring blood transfusion, but the pressure effect on cord decreased and the powers in limbs gradually increased. Few clinical situations like in pregnancy transfusion therapy is advised as both have got risk. Cytostatic agents like hydroxy urea may also be used.

Conclusion

Patients' with chronic anemia can present with intra-thoracic spinal lesions and paravertebral mass lesions, and can be a cause of paraparesis. MRI procedure is very helpful in seeing the above lobulated mass lesion in such hematological condition, and a follow-up study is helpful.

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