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Case Report

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Spinal Cord Compression Secondary to Extramedullary Hematopoiese in a Patient with a Beta-Thalassemia

Bekralas H[1]*, Bouallag M[1], Khalfi Kh[1], Habchi N[1,2], Chitti M[1], Boulaouad W[1] and Djaafer M[1]*

**Head of Department of Neurosurgery, Mustapha PACHA Hospital Algiers, Algeria*

[1]*Department of Neurosurgery, Mustapha PACHA Hospital Algiers, Algeria*

[2]*Laboratory of Endocrinology, FSB-USTHB, Algiers, Algeria*

***Corresponding author**

Bekralas Houria, Department of Neurosurgery, Mustapha PACHA Hospital Algiers, Algeria. E-mail: nawelmedbio@yahoo.fr

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Abstract

Extramedullary hematopoiesis (EMH) is a rare cause of spinal cord compression (SCC). EMH represents the growth of blood cells outside of the bone marrow and occurs in a variety of hematologic illnesses, including various types of anemia and myeloproliferative disorders. Although EMH usually occurs in the liver, spleen, and lymph nodes, it may also occur within the spinal canal. When this occurs, the mass effect can compress the spinal cord, potentially leading to the development of neurological deficits. We present a case of SCC secondary to EMH. Our patient is a 26-year-old male with beta-thalassemia who presented with both upper thoracic and lower extremity symptoms of spinal cord compression. This report illustrates the importance of considering EMH in the differential diagnosis of SCC, even in the absence of signs of its most common etiologies.

Keywords: Beta-Thalassemia, Extramedullary Hematopoiesis, Spinal Cord Compression

Introduction

The extramedullary hematopoiesis is a rare entity found in chronic hemolytic disorders, secondary to a compensatory phenomenon of anemia in long short. Its development at the epidural space is rare and may cause spinal cord compression [1-3].Being a non-well defined, its treatment is unclear.

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BF patient aged 26 years carrying a homozygous B thalassemia admitted in neurosurgery for motor deficit lasting for 5 months. The review found paraparesis without sensory or sphincter disorders.

NSF found microcytic anemia; SEPs: spinal cord in the neck. MRI: posterior epidural mass with moderate contrast enhancement and heterogeneous extending from D3 to D8 measuring 110x100 mm without bone loss next (Figure 1).

Figure 1: Pre operative MRI shows a posterior epidural mass heterogeneous extending from D3 to D8

The patient underwent a decompression laminectomy from D3 to D7 with a wide excision of the tumor foci nickname.

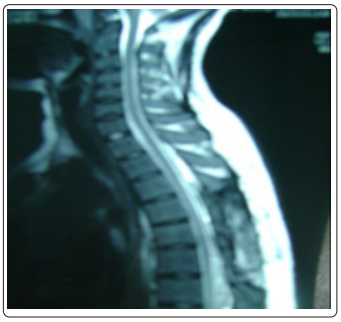

Figure 2: Post operative MRI

The postoperative course was uneventful with rapid recovery (Figure 2).

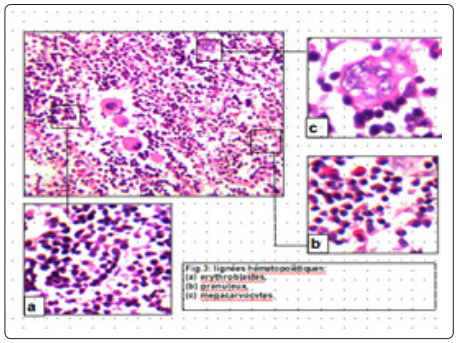

Figure 3: Histological features of hematopoietic cells

Pathological examination of the resection material showed a proliferation of hematopoietic cells of erythroid, granulocytic and dystrophic mégaryocytaire regular or at all stages of maturation (Figure 3).

Discussion

The hematopoiesis is a compensatory mechanism in response to persistent chronic anemia usually occurs between 40 and 60. Although relatively common in Beta thalassemia location in the epidural space is rare [4]. It is often asymptomatic, symptomatic changes to spinal cord compression is exceptional, a fifty cases have been reported, the first being in 1954 by GATTO and al.

EMH is a physiological compensatory mechanism which occurs when bone marrow cannot meet the circulatory needs. Therefore, EMH occurs in patients with thalassemia whose chronic severe anemia has not been corrected by blood transfusion. Typical sites of EMH can be any organ that participates in hematopoiesis during fetal development such as the spleen, liver, kidneys, and lymph nodes as well as other less commonly known sites such as heart, breasts, prostate, broad ligaments, pleura, cranial nerves, and the spinal canal [5,6]. Particularly, in patients with thalassemia, paraspinal involvement is seen in approximately 15% of the cases [1].

Ethiopathogénie: The origin of hematopoietic tissue is still a matter of controversy expulsion of trabecular bone Embryological remnants of hematopoietic cells in the epidural space Diagnosis is made mostly spinal MRI shows hyper intense foci on T1 and T2 [1,2].

Histology showing normal or dystrophic hematopoietic cells at different stages of maturation associated with fibrosis. The treatment is not well codified given the small number of cases asymptomatic for the authors propose a hyper transfusion followed by radiotherapy if it remains refractory. Treatment with hydroxyurea cytostatic agent reduces the amount of hematopoietic tissue appears to have given good results. In symptomatic cases, surgical resection is in order followed by radiotherapy. Both surgery and radiation were usually well tolerated by patients whose spinal involvement was not as extensive as this case. Resection of the mass can lead to quick decompression [5].

Conclusion

The diagnosis of hematopoiesis should be considered in any array of spinal cord compression in a patient with a chronic blood disorder. Its management remains open to debate given the low number of reported cases.

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