Extramedullary hematopoiesis

Formation of blood cells outside the bone marrow. It is a common manifestation of many chronic hemolytic anemias, and typically involves the liver, spleen, and lymph nodes.

Only rarely is the spinal epidural space involved.

The origin of the spinal epidural hematopoietic tissue is still controversial. It has been hypothesized that this tissue could be extruded through the trabecular bone of the vertebral body with a circumferential involvement of the vertebra, or it may have extended through the thinned trabeculae at the proximal rib ends.

Others have proposed some embryological hematopoietic cell remnants within the epidural space, which would be stimulated along the course of chronic anemia. Development of hematopoietic tissue from branches of the intercostal veins has also been suggested, while others still attribute the masses to embolic phenomena.

Early in its evolution, the paraspinal extramedullary site of hematopoiesis reveals immature and mature cells mainly of the erythroid and myeloid series and dilated sinusoids containing precursors of red cells. The lesions eventually become inactive and reveal some fatty tissue and fibrosis or massive iron deposits.

There is some predilection for the site of spinal cord involvement by the hematopoietic tissue. The thoracic region and to a lesser extent the lumbar region are the most commonly involved sites. The reason for this predilection is uncertain, but because the subarachnoid space and the spinal canal are narrow in the thoracic region, which also has limited mobility, small intraspinal hematopoietic tissue may cause compression of the spine at this level. This is in contrast with other regions of the cord in which such tissues must reach larger sizes to exert enough pressure on the spinal cord and cause symptoms.

A paraspinal location for the hematopoietic tissue occurs in 11–15% of cases with EMH.

Since the first case described by Gatto et al. in 1954, a large number of cases has been reported in the literature.

Paraspinal EMH mainly presents as pseudotumors, which may cause a variety of neurological symptoms due to spinal compression. However, it is believed that more than 80% of cases may remain asymptomatic and the lesions are usually discovered incidentally by radiologic techniques. Probably the development of neurologic symptoms depends on the chronicity of the disease with neurologic symptoms most frequently being reported during the third and fourth decades of life , although few reports described presentation as early as the first decade of life. The male to female ratio reaches 5:1. Various clinical presentations have been reported including: back pain, lower extremity pain, parasthesia, abnormal proprioception, exaggerated or brisk deep tendon reflexes, Babinski response, Lasegue sign, paraparesis, paraplegia, ankle clonus, spastic gate, urgency of urination, and bowl incontinence. The size and location of lesions and the extent of spinal cord involvement determine the severity, acuteness, and multiplicity of signs and symptoms. [1)].

Treatment

Surgical excision followed by radiation therapy has been the recommended treatment. Repeated blood transfusions may help reduce EMH and may be useful post-op instead of RTX except for refractory cases.

Surgery on these patients is difficult because of:

1. low platelet count

2. the poor condition of the bone

3. cardiomyopathy: increased anesthetic risk

4. anemia, coupled with the fact that most of these patients are "iron-toxic" from multiple previous transfusions

5. total removal of the mass is not always possible

Case reports

2016

A report illustrates the importance of considering EMH in the differential diagnosis of SCC, even in the absence of signs of its most common etiologies [2].

2013

A 25-year-old male, known to have thalassemia intermedia, who presented with a 1-month history of stiffness and weakness in both lower extremities. On physical examination, he had palpable splenomegaly accompanied by spinal tenderness at the D5 level, weakness in both lower extremities, hyperactive bilateral Patellar and Achilles reflexes with bilateral Babinski responses, and a graded sensory loss to pin appreciation below D5.

The magnetic resonance (MR) study revealed a posterior, isointense and soft tissue epidural mass extending from D2 to D12 on both the T1- and T2-weighted images. These findings were consistent with the diagnosis of "red marrow," and long-segment spinal epidural extramedullary hematopoiesis.

Although extramedullary hematopoiesis is rarely encountered within the spinal canal, it should be considered among the differential diagnoses when a posterior compressive thoracic lesion contributes to myelopathy in a patient with a history of thalassemia intermedia and the accompanying chronic hemolytic anemia [3].

1)
http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2899982/
2)
Wang A, Carberry N, Solli E, Gillick J, Islam H, Hillard V. Spinal Cord Compression Secondary to Extramedullary Hematopoiesis: Case Report and Review of the Literature. Case Rep Oncol. 2016 May

31;9(2):290-7. doi: 10.1159/000446473. eCollection 2016 May-Aug. PubMed PMID: 27462228.
3)
Garg K, Singh PK, Singh M, Chandra PS, Sharma BS. Long segment spinal epidural extramedullary hematopoiesis. Surg Neurol Int. 2013 Dec 26;4:161. doi: 10.4103/2152-7806.123657. PubMed PMID: 24404404.