Scholars Journal of Medical Case Reports

OPEN ACCESS

Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

Para-Vertebral Extramedullary Hematopoiesis (PVEH) A Report of Two Cases and Literature Review

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DOI: 10.36347/sjmcr.2021.v09i10.022 | **Received:** 30.08.2021 | **Accepted:** 04.10.2021 | **Published:** 21.10.2021

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Abstract **Case Report**

Para-vertebral extramedullary hematopoiesis (PVEH) is defined as an ectopic development of hematopoietic tissue outside the bone marrow. It is a very rare condition, secondary to haematological disorders leading to chronic anemia. The present report describes two cases of chronic anemia patients with paraspinal medullary hematopoiesis and analyzes the clinical manifestations, imaging, pathology, diagnosis and treatment of EMH. In addition, a supplementary review of previously published cases is provided along with a review of the related literature. Computed tomography (CT) of the first case revealed paraspinal masses, and the largest was 6.2x8.0 cm in diameter. Likewise, MIRy of the second patient revealed multiple paraspinal masses in the bottom of the left thoracic cavity, and the largest was measured 10.1x10.5 cm. In conclusion, EMH is a compatible and rare disease, and should be distinguished from other neoplasms.

Keywords: Para-vertebral extramedullary hematopoiesis, haematological disorders.

INTRODUCTION

Extramedullary hematopoiesis (EMH) refers to the hematopoiesis that occurs in organs other than bone marrow [1]. It is a compensatory phenomenon that results in the production of blood cell precursors outside the marrow in patients with haematological disorders. It has been reported to occur in 15% of cases of thalassemia, and it occurs in myelofibrosis and in other anemic conditions [2]. The process can involve virtually any organ or tissue, such as the liver, spleen, abdominal viscera, pleura, lymph nodes, adrenal glands, breast, thymus, kidneys, gastrointestinal tract, intracranial structures and paraspinal regions [3–6]. EMH is a rare entity and often asymptomatic but can sometimes lead to symptomatic tumor-like masses. Treatment options are controversial and include hypertransfusion, surgical excision, radiotherapy, and hydroxyurea [7]. Paraspinal involvement is the associated with morbidity secondary to spinal cord compression [8]. Among the various body regions reported, paraspinal involvement deserves special attention due to the debilitating clinical consequences and challenges in its diagnosis and management. The present study described two cases with paraspinal medullary hematopoiesis in two haematological disorders patients. Verbal informed consent was obtained from both patients.

CASE REPORT

Case 1

44-year-old patient, chronic tobacco user with the notion of tuberculosis contagion and a chronic cough for a year with back pain in a context of apyrexia and intense fatigue. He is monitored in the hematology department for chronic idiopathic erythroblastopenia. The CT scan shows multiple posterior para-vertebral mediastinal tissue masses bilateral opposite T11-T12, of regular contours, homogeneous with a fatty component, without calcifications within them, moderately enhanced after injection of contrast product. With associated hepato-splenomegaly. MRI shows the presence at the level of the two costo-vertebral splints opposite T9-T10-T11 of five oval formations, well circumscribed, in heterogeneous T1 and T2 hypo signal with fine partitions, signal void zones and a component fat that is canceled out in FAT SAT within them, evoking foci of extra-medullary hematopoiesis.

Case 2

59-year-old patient, followed in hematology for hemolytic anemia, admitted for pain in the right hypochondrium in a febrile context. With clinical examination painful hepatomegaly and splenomegaly. On ultrasound, hepatomegaly with cystic formation of segment IV and multi-lithiasic VB with a laminated wall is noted, as well as a homogeneous splenomegaly.

CT scan: Appearance in favor of acute cholecystitis complicated by an abscess of segment IV opposite. With the lesional process of the two costo-vertebral splints associated with a homogeneous splenomegaly, first of all suggesting an extra-medullary para-vertebral hematopoiesis.

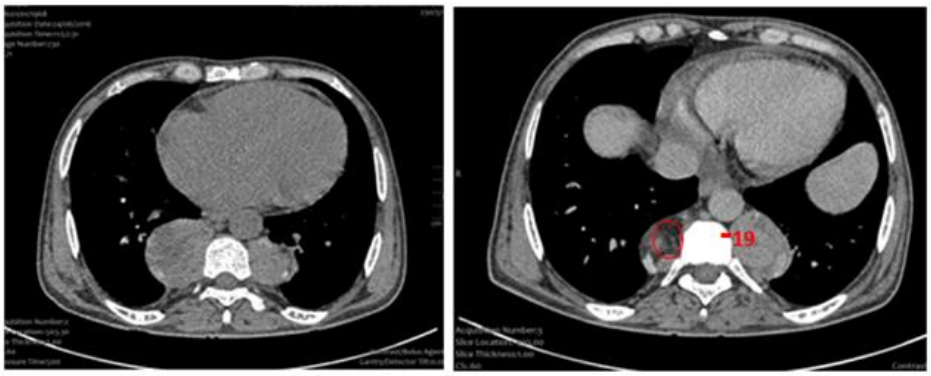

Figure a et b: case 1: Axial sections: showing multiple posterior para-vertebral mediastinal tissue masses bilateral opposite T11-T12, of regular contours, homogeneous with a fatty component, without calcifications within them, moderately enhanced after injection of contrast product

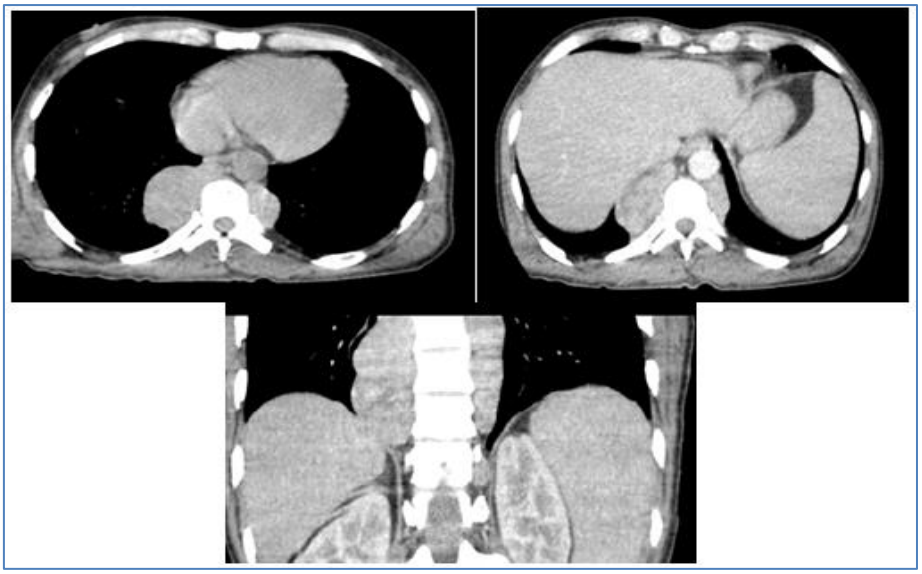

Figure a et b et c: Axial and coronal sections: showing a lesional process of the two costo-vertebral splints, well limited measuring 5x3.8 cm on the right and 3x2 cm on the left, heterogeneously enhanced hypodenses after injection of the PDC, without detectable bone lysis or endoductal extension.

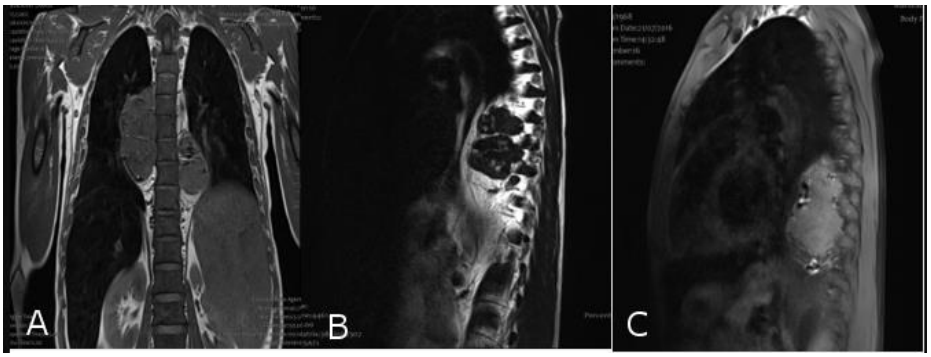

Figure a et b et c: Thoracic MRI: Coronal T1: Bilateral para-vertebral masses in heterogeneous T1 hypointense with T1 hyperintensity component. b: T2 sagital: Fat component in T2 hyperignal. c: T1 FAT SAT sequence: Bilateral para-vertebral masses with fatty component which fades in FAT SAT sequence.

DISCUSSION

EMH is defined as hematopoiesis occurring in organs outside of the bone marrow. It occurs if the bone marrow is no longer functional and is identified in a number of hematological diseases; its occurrence in chronic hemolytic anemias remains highest, particularly in transfusion-independent thalassemia intermedia (2). Ineffective red cell production by the bone marrow forces expansion of the hematopoietic tissue outside the marrow medulla and results in hematopoietic compensatory involvement, particularly in the form of masses, of other areas in the body [3].

Among the various body regions described, paraspinal involvement requires particular attention due to the debilitating clinical consequences and challenges in management and diagnosis. A paraspinal location in hematopoietic tissue occurs in 11–15% of cases with EMH [8]. 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 frequently involved sites. The cause of this predilection is uncertain; however, these sites are understood to normally engage in active hematopoiesis in the fetus during gestation [4]. This pathway typically stops at birth, but the extramedullary hematopoietic vascular connective tissues retain the ability to produce red cells under conditions of long-standing ineffective erythropoiesis.

Paraspinal EMH typically presents as pseudotumors, and can sometimes result in symptomatic tumor-like masses, which may cause a variety of neurological symptoms due to spinal compression. However, it is understood that >80% of cases do not have signs and symptoms related directly to the disorder, and the lesions are typically discovered incidentally by radiologic techniques. The development of neurologic symptoms is suggested to depend 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, Lasègue sign, paraparesis, paraplegia, ankle clonus, spastic gate, urgency of urination and bowel incontinence. The size and location of lesions and the extent of spinal cord involvement determines the severity, acuteness and multiplicity of signs and symptoms.

Although the history and physical examination may help narrow the differential diagnosis, radiographic imaging remains essential to confirm the existence of hematopoietic tissue. Characteristic appearance has been observed primarily using magnetic resonance imaging (MRI) or CT scan. MRI is the diagnostic investigation of choice. Paraspinal EMH appears as unique, multiple iso- or hyperintense masses, with homogeneous enhancement following contrast administration. These masses are usually lobular, well-circumscribed masses of intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted image. The CT appearance is characterized by the heterogeneous soft tissue density mass. The diagnosis is confirmed following surgical removal of the mass. Biopsy remains the gold standard.

Because of its rarity, there is no standard treatment approach in patients with symptomatic EMH, and no evidence-based guidelines for the treatment of EMH. Therapy typically depends on the severity of symptoms, size of the mass, patient's clinical condition and previous treatment. Numerous treatment options have been described, including transfusion therapy, laminectomy, radiotherapy and the use of fetal hemoglobin, inducing agents that decrease the hematopoietic drive. However, the ideal management scheme remains controversial. Until large prospective trials evaluate the efficacy and safety of the available treatment options, both in single and in combination therapy, an individualized approach should be devised.

CONCLUSION

EMH is a rare disease, which is complicated to preoperatively distinguish from other neoplasms. In the present study, two patients with EMH are presented. The two patients underwent surgery and the pathological findings indicated that EMH was the correct diagnosis. These two cases suggest that EMH must be taken into consideration when masses with characteristic radiologic appearance are identified in patients with various hematological diseases.

RÉFÉRENCES

1. Bozzini, C. E., Barrio Rendo, M. E., Devoto, F. C., & Epper, C. E. (1970). Studies on medullary and extramedullary erythropoiesis in the adult mouse. *American Journal of Physiology-Legacy Content*, *219*(3), 724-728.
2. Papavasiliou, C. (1994). Clinical expressions of the expansion of the bone marrow in the chronic anemias: The role of radiotherapy. *International Journal of Radiation Oncology* Biology* Physics*, *28*(3), 605-612.
3. SANEEI, T. M., Birang, S. H., Shahnazi, M., & Hemadi, H. (2005). Large splenic mass of extramedullary hematopoiesis.
4. Miyake, H., Matsuda, M., Iyomasa, S., & Mizuno, K. (2004). Presacral extramedullary hematopoiesis. *Surgery*, *135*(1), 112-113.
5. Aessopos, A., Tassiopoulos, S., Farmakis, D., Moyssakis, I., Kati, M., Polonifi, K., & Tsironi, M. (2006). Extramedullary hematopoiesis-related

pleural effusion: The case of β-thalassemia. *The Annals of thoracic surgery*, *81*(6), 2037-2043.

6. Al-Habib, H., & Hadzikaric, N. (2007). Spinal cord compression due to intraspinal extramedullary hematopoiesis in thalassemia intermedia. *Neurosciences Journal*, *12*(3), 261-264.
7. Eskazan, A. E., Ar, M. C., & Baslar, Z. (2012). Intracranial extramedullary hematopoiesis in patients with thalassemia: a case report and review of the literature. *Transfusion*, *52*(8), 1715-1720.
8. Dore, F., Cianciulli, P., Rovasio, S., Oggiano, L., Bonfigli, S., Murineddu, M., & Papa, G. (1992). Incidence and clinical study of ectopic erythropoiesis in adult patients with thalassemia intermedia. *Annali italiani di medicina interna: organo ufficiale della SocietÃ italiana di medicina interna*, *7*(3), 137-140.
9. Cromwell, L. D., & Kerber, C. (1978). Spinal cord compression by extramedullary hematopoiesis in agnogenic myeloid metaplasia. *Radiology*, *128*(1), 118-118.
10. Spector, N., Lavrado, F. P., Nobre, L. F., de Mattos, J. P., Pulcheri, W., Nucci, M., & de Oliveira, H. P. (1996). Spinal cord compression due to extramedullary hematopoiesis in the proliferative phase of polycythemia vera. *Acta haematologica*, *96*(4), 242-244.