Case Report

# Extramedullary hematopoiesis causing spinal cord compression with excellent durable response after radiation therapy: Case report and review of the literature

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## Abstract

Extramedullary hematopoiesis (EMH) is a rare occurrence in the setting of spinal cord compression. We report on a 72-year-old who was initially diagnosed with polycythemia vera (PV) which after approximately 15 years converted to myelofibrosis which confirmed on bone marrow biopsy. In 2016, he presented to our ED with clinical symptoms suggested of spinal cord compression at the T3–8 region. This was confirmed by MRI imaging. After a review of existing literature, it was elected to treat the affected area with radiation consisting of 15 fractions of 200 cGy. Within 10 days, the patient had begun to regain strength in the affected regions both motor and sensory. At the 2 month follow-up, he was symptom-free and imaging also showed a complete response. In January 2019, the patient again presented with clinical symptoms of spinal cord compression in the T10–12 area. Again, this was confirmed by MRI imaging. The same fractionation scheme was used and again the patient had a complete resolution of all symptoms both motor and sensory at the 1-month follow-up. Of interest is that during both the courses of treatment there was not a significant in any blood indices from baseline presentation. In the setting of EMH-causing cord compression, the use of radiation is warranted with excellent early response that appears durable. In addition, we present a review of the literature on this topic.

**Keywords:** Extramedullary hematopoiesis, radiation therapy, spinal cord compression

## Introduction

Extramedullary hematopoiesis (EMH) is defined as the finding of hematopoietic elements outside the traditional location of the bone marrow. It has been seen in various locations of the body including the spleen, liver, and lymph nodes which seem to be favorite sites;[1,2] however, the finding of EMH in the spinal cord is a rare condition. It has been reported in connection with thalassemia,[3,4] sickle cell anemia,[5] polycythemia vera (PV),[6,7] and myelofibrosis[8,9] among hematologic conditions.

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**Received:** 03-01-2020 **Revised:** 11-03-2020
**Accepted:** 14-03-2020 **Published:** 30-07-2020

Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/jfmpc.jfmpc_22_20

In a review of the literature, we can find very few cases of PV converting to myelofibrosis and associated with EMH that result in spinal cord compression (SCC). A recent abstract by Yassin *et al.*[10] reported only 16 cases since 1956. There was a 7:1 male to female ration with the age group ranging from 30 to 75 years old. Mattei *et al.*[11] reported a 5:1 male to female ratio in their case report. We could find only two previous cases of a patient initially diagnosed with PV converting to myelofibrosis and then developing a spinal cord due to EMH. Our case is even more unique in that our patient has had two episodes of EMH presenting as SCC in two separate/close regions and both obtaining radiographic and clinical complete response.



**How to cite this article:** Fontanesi J, Margolis H, Fontanesi GR. Extramedullary hematopoiesis causing spinal cord compression with excellent durable response after radiation therapy: Case report and review of the literature. J Family Med Prim Care 2020;9:3741-4.

## Case Presentation

A 72-year-old male who was initially diagnosed with PV approximately 25 years ago converted to myelofibrosis which was confirmed on bone marrow biopsy. In 2016, he presented to our ED with clinical symptoms suggested of SCC at the T8–12 region. This was confirmed by MRI [Figures 1 and 2]. After a review of existing literature, it was elected to treat the affected area with radiation consisting of 15 fractions of 200c Gy. Within 10 days, the patient had begun to regain strength in the affected regions both motor and sensory. At 2-month follow-up, he was symptom-free and imaging also showed a complete response. In January 2019, the patient again presented with symptoms of SCC in the area immediately above the previously treated area. Again this was confirmed by MRI [Figures 3 and 4]. The same fractionation scheme was used and again the patient had a complete resolution of all symptoms both motor and sensory at a 1-month follow-up. Of interest is that during both courses of treatment there was no significant change in any blood indices from baseline presentation.

## Discussion

EMH is a relatively common occurrence in many chronic hematologic diseases such as B thalassemia, sickle cell anemia, and PV It has been reported in the spleen, liver, kidney, adrenal glands, heart, and lymph nodes among many sites,[1,2] however, finding of EMH causing an SCC is even rarer. The cause of EMH occurring in the spinal cord has been speculated but no definitive answer is known although various hypothesis has been suggested such as bone marrow transport through bony erosions/fracture, development from preexisting primitive tissues.[12] Whenever an SCC is suspected in a patient with clinical findings supporting this diagnosis, time is of the essence. From an imaging standpoint, MRI is best suited to allow identification of the extends of the location. It will usually be well-defined as an isointense mass with rim enhancement on Tl signal and hyperintense mass on T2.[4]

The treatment of SCC in the setting of EMH varies in the literature. Surgical decompression alone or in conjunction with postoperative radiation has been reported as has the use of radiation alone and various systemic therapies such as chemotherapy and transfusion therapy.[13]

We do know that the components that are involved in EMH are extremely sensitive to radiation yet various radiation total doses (ranging from 7.5 to 35 Gy), the daily dose (1–5.5 Gy). This is interesting in that seems to be a good consensus regarding the treatment of "chloma's" that are associated with all patients and the components of EMH are similar.[14,15]

The reports of EMH presenting as an SCC in patients that convert from PV to MF are even more unique. Reale *et al.* reported 15 cases from the literature on patients with myelofibrosis who had developed SCC of which two had converted from an initial diagnosis of PV.[16] In their series, PV/MF patients had surgery and progressed to AML and died. The second received steroids plus 22 Gy/4 fractions with reported rapid improvement.

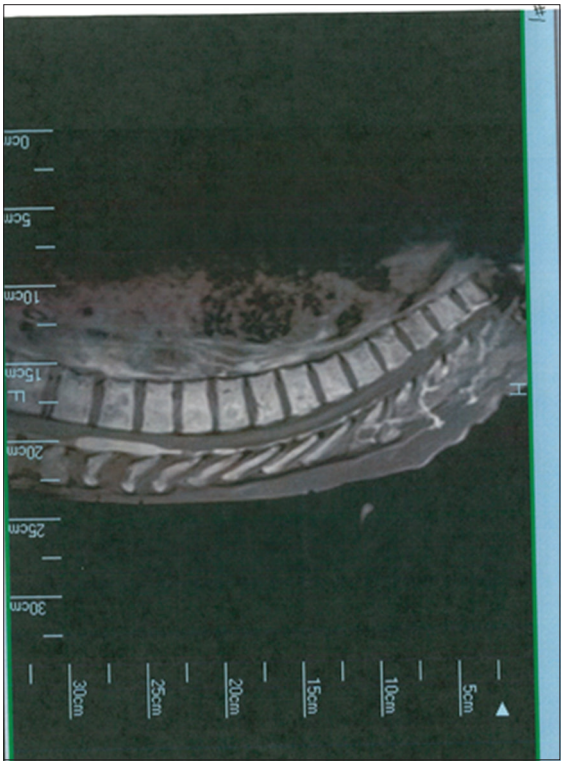


**Figure 1:** MRI 1

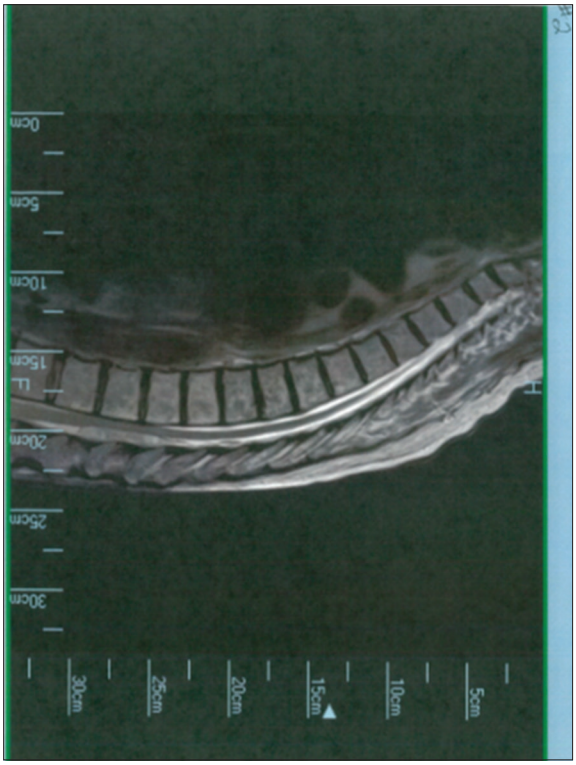


**Figure 2:** MRI 2

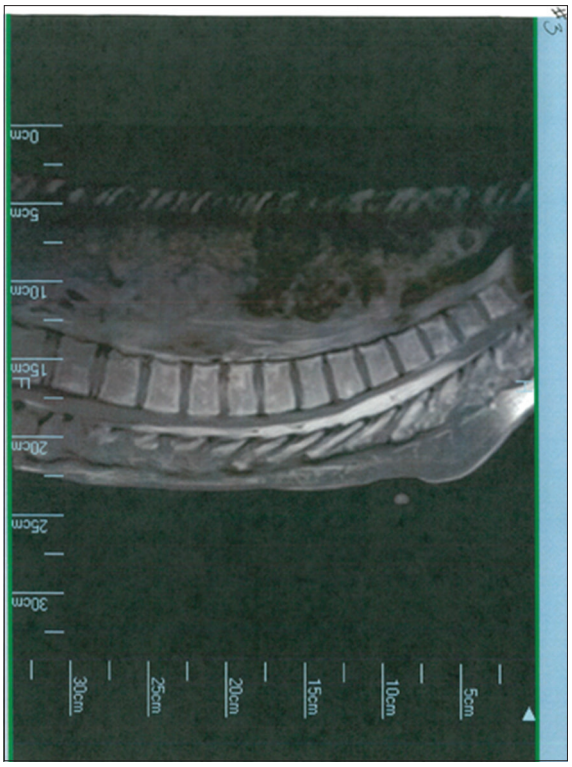


**Figure 3:** MRI 3

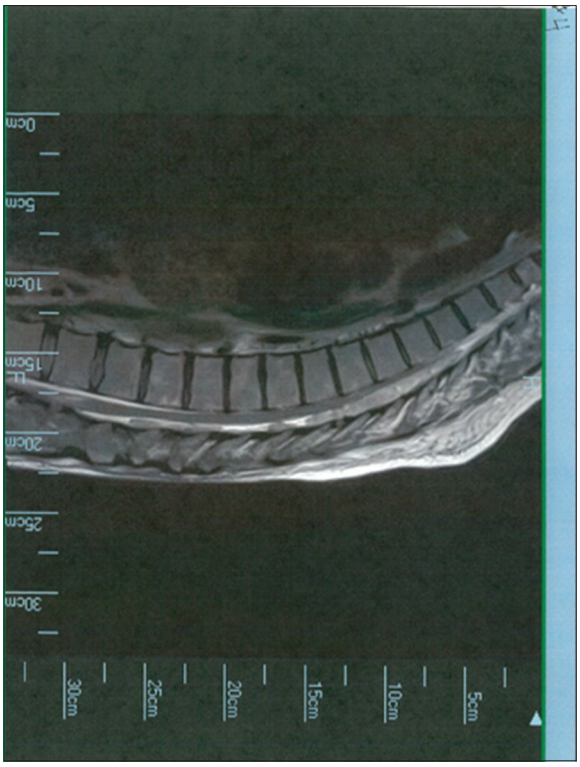


**Figure 4:** MRI 4

However, there have been reports of up to 19% recurrence rates after decompressive surgery +/radiation therapy in all cases that have been treated for EHM.[17,18] Unfortunately, there are not enough that allow us to suggest any dose-response curve for this entity.

Our case is unusual in that the patient had a complete response to our first course of therapy both clinically and from an imaging standpoint until he presented with the second SCC in the area above the previous treatment area. For our first treatment site, we used the MRI-described finding and added one vertebral body above and below that imaging anomaly. It is interesting that the second SCC developed at the vertebral above the superior border of our first field.

Based on our limited experience and review of literature the finding of SCC in patients that convert from PV to MF is extremely rare. We believe that our dose fractionation schedule is effective and has shown durability. In select cases, surgery might be considered but early diagnosis, we believe, is the key to the resolution of clinical symptoms, as it for most SCC regardless of cell type/disease entity.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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