Case Report

Extradadular Hematopoiesis Presenting with Thoracic Spinal Cord Compression in a Young Adult with Thalassemia Major: A Case Report

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**ABSTRACT**

**Background:** Extradadular hematopoiesis (EMH) refers to the production of blood cellular components at sites other than the bone marrow, namely liver, spleen, and lymph nodes. The common sites associated with this condition are the liver, spleen, and lymph nodes whereas the common conditions associated with it are myelofibrosis, myelodysplasia, thalassemia, sickle cell anemia, and polycythemia vera. **Case Description:** This report describes a young male with thalassemia major, who presented with symptomatic cord compression due to a thoracic intraspinal lesion. It was surgically excised and diagnosed as a case of EMH. The boy recovered fully and has been asymptomatic for six months now. **Conclusion:** The occurrence of EMH in the thoracic spine is uncommon, whereas symptomatic cord compression as a result of it is even more unusual. Magnetic resonance imaging (MRI) is the diagnostic imaging of choice and treatment options that can be offered are surgical decompression, radiotherapy, hydroxyurea, and transfusion of packed red blood cells (RBCs).

**KEYWORDS:** Cord compression, extradadular, hematopoiesis, spine, thalassemia

**INTRODUCTION**

EMH occurs as a result of reduction in the functional RBCs due to myeloproliferative neoplasms and non-neoplastic hematological pathologies such as thalassemia, sickle cell anemia, and myelofibrosis. The common non-hepatosplenic sites of EMH are the central nervous system (CNS), ovaries, pericardium, pleura, vertebral column, etc.[1] Since the first description of the EMH in 1954 by Gatto et al.[2] in a patient with thalassemia, several treatment options have been proposed in literature, which include surgical excision, focal radiotherapy (RT), hypertransfusion of RBCs, hydroxyurea, etc. However, a consensus on an ideal treatment modality is yet to be reached.[3]

**CASE REPORT**

An 18-year-old boy presented with complaints of back pain and progressive weakness in bilateral lower limbs for one month. He was a known case of thalassemia major since four years, and he had been receiving regular RBC transfusion. On examination, the power in bilateral knee extensors, ankle plantar flexors, and ankle dorsiflexors was 2/5. Knee reflex and ankle reflex was exaggerated in both lower limbs (4+), and the plantar reflex was extensor. There was no sensory abnormality, and bladder function was normal. Liver was palpable and a previous scar over the splenic area was seen, consistent with a past history of splenectomy. A complete blood count (CBC) analysis showed hemoglobin of 9.5 gm/dl, hematocrit of 27.6%, RBC count of 3.32 million/ul, and platelet count of 3,95,000. Peripheral smear analysis showed anisopoikilocytosis, predominantly normocytic normochromic RBCs with the presence of a few macrocytes, polychromasia, the presence of nucleated red cells (68% of total nucleated cells), and target cells. Chest x-ray showed a widening...
of diploic spaces in all the thoracic ribs with a thinning of their inner table along with prominent widening of anterior ends of the ribs.

A MRI of the dorsal spine showed the presence of a large T2 hyperintense paravertebral mass extending from thoracic 4 to thoracic 10 (T4-T10) vertebrae. The lesion was homogenously enhancing, involving the epidural fat and caused a complete obliteration of the corresponding subarachnoid space, leading to marked cord compression [Figure 1]. The differential diagnoses considered were a hematological malignancy or an EMH. An emergency spinal cord decompression was planned. A T5 to T10 laminectomy was performed. The lesion was soft, completely extradural, and grayish white in color. The bony tissue was firm with an abnormal marrow appearance. Complete resection was done to achieve adequate decompression. Intraoperatively, the patient received two pints of packed RBC transfusion. On histopathology, the dorsal lamina of the spine showed fibrocollagenous hyperplastic bone marrow and the mass showed erythroid hyperplasia. By the time of discharge, the patient was able to walk with support [Figure 2] and at a follow-up of 6 months, he showed complete recovery.

**DISCUSSION**

The EMH is a compensatory mechanism occurring in chronic anemia, whereby the body attempts to maintain erythropoiesis. Hence, even though the occurrence of EMH is seen in younger age groups, symptoms tend to develop, if at all, later on in life. A database review of 1933 EMH cases from Mayo clinic found that approximately 83% of cases were associated with myeloproliferative neoplasms. Among the EMH cases with non-myeloproliferative neoplasms (n = 309), the majority occurred concurrently with myelodysplastic syndrome (13%), whereas thalassemia was seen in 7% of cases.
of cases, Paraspinal occurrence of EMH has been reported to vary from an incidence of 11% to 26%, with the thoracic spine being the most common location. The majority of EMH patients are asymptomatic (80%), and the lesion is more often than not incidental in nature. However, the EMH occurring in the thoracic spine has a high propensity to cause severe neurological deficits due to the narrow spinal canal and the relatively limited mobility of the thoracic spine.

Pathologically, the plausible hypothesis suggested for occurrence of paraspinal EMH are:

i) Direct extension of erythrogenesis from adjacent vertebral bone marrow

ii) Common embryonic origin from thoracic hematopoietic tissue masses

iii) Development from branches of the intercostal veins

iv) Arterial embolus

History of myeloproliferative disorders or hematological pathologies should be ruled out in any young patient presenting with lower limb weakness. A MRI is the diagnostic imaging of choice and it shows the presence of a lobulated mass, hyperintense on T1 and T2-weighted images with minimal post-contrast enhancement if the disease is early or in late phase and marked enhancement if the lesion is active. This feature helps to differentiate it from the more commonly occurring epidural lesions, namely abscess, tumors (lymphoma) etc.

Although many treatment options have been described in literature, immediate surgical decompression is imperative to reverse the myelopathy in cases presenting with acute cord compression. Care must be taken to prevent the complication of bleeding from an incompletely resected mass since this might deteriorate

### Table 1: Review of English literature mentioning cases with pediatric thalassemia and with extramedullary hematopoeisis causing cord compression

| Sr no. | Author            | Year | Age (years) | sex  | Presentation | Lesion     | Treatment | Recovery     |
|--------|-------------------|------|-------------|------|--------------|------------|-----------|--------------|
| 1      | Issaragrisil et al. | 1981 | 17/M        |      | Paraparesis  | T4-12      | RT + BT   | Partial      |
| 2      | Ahmed et al.      | 1981 | 17/M        |      | Paraparesis  | N/A        | Surgery   | Partial      |
| 3      | Ibrahim et al.    | 1983 | 17/M        |      | Back pain    | T4-8       | Surgery   | Complete     |
| 4      | Amirjamshidi et al. | 1991 | 14/M 16/F   |      | Paraparesis  | Male T5-T11 | Male given | Complete in both |
|        |                   |      |             |      |              | Female T9-T10 | adjuvant RT 1500 rad + multiple BT | |
| 5      | Khandelwal et al. | 1992 | 14/M        |      | Paraparesis + hesitancy in micturition | D8-11      | BT        | Complete     |
| 6      | Mancuso et al.    | 1993 | 15/M        |      | Foot weakness + urinary retention | L2-4       | BT        | Complete     |
| 7      | Parsa et al.      | 1995 | 16/F        |      | Paraparesis  | T9-10      | BT + 1500 rad RT | Complete |
| 8      | Tan et al.        | 2002 | 17/F        |      | Paraparesis + difficulty in micturition | T5-8       | BT + Surgery + RT 2000 rads RT in 10 fractions | Complete |
| 9      | Tai et al.        | 2006 | 15/F        |      | Paraparesis  | T2-10      | BT        | Complete     |
| 10     | Moncef et al.     | 2008 | 18/M        |      | Back pain    | T12        | Surgery + BT + RT (2000 cGy) | Complete |
| 11     | Ileri et al.      | 2009 | 9/M         |      | Lower limb pain | L4-S1      | RT. 2400 cGy in 10 fractions, + HU 15 mg/kg oral | Complete |
| 12     | Soman et al.      | 2009 | 16/M        |      | Gait disturbance due to sensory symptoms in lower limbs Paraparesis | T5-7       | BT + 1500 cGy RT | Complete |
| 13     | Bukhari et al.    | 2016 | 18/M        |      |              | T6-L3      | Surgery – recurrence after 4 months – surgery + two cycles of RT | Complete |
| 14     | Current case      | 18/M | 18/M        |      | Back pain + paraparesis | T4-10      | Surgery + BT | Complete     |

RT = radiation, BT = blood transfusion, HU = hydroxyurea

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the hematological profile further. Low-dose focal spine RT (200cGy x 10 doses) has a higher risk of recurrence, which is variably reported in literature to be as close to 30%. The RT may be reserved for spinal EMH cases that are associated with myelofibrosis-myelodysplastic syndrome. Medical management with hydroxyurea (stimulates fetal hemoglobin production) and with packed RBCs has also been described in literature in cases where surgical treatment is either not warranted or contraindicated. Asymptomatic, incidental cases can be conservatively managed with regular monitoring. An extensive review of English literature shows that 13 cases with pediatric thalassemia and with EMH causing cord compression have been reported so far [Table 1]. Among them, the majority (9/13) have had combination therapy. Recurrence within four months was seen in a single case in whom only surgical excision was initially performed. Subsequently, redo surgery with adjuvant radiation was then given.

**CONCLUSION**

The EMH is a common condition occurring in patients with ineffective erythropoiesis. It must be considered as a strong differential diagnosis in young adults with a history of hematopoietic disorder, presenting with acute cord compression and an epidural hyperintense lesion enhancing on contrast MRI. Consensus on treatment protocol is not yet defined, with data limited to isolated case reports or small series. Surgery or radiation therapy can be offered to patients with symptomatic spinal cord compression with surgery, offering the possibility of rapid reversal of neurological deficits. Literature favors a multipronged approach in these patients.

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**Conflicts of interest**

There are no conflicts of interest.

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