Paraplegia due to extramedullary hematopoiesis in thalassemia treated successfully with radiation therapy

Spinal cord compression due to extramedullary hematopoiesis (EMH) is a rare complication of thalassemia and generally presents as paraparesis with sensory impairment. Complete paraplegia is extremely rare in EMH due to thalassemia although it is known to occur in polycythemia vera and sickle cell anemia. Treatment options mostly include surgery and/or radiotherapy. Whereas cases presenting with paraparesis have been treated with either surgery or radiotherapy with equal frequency and efficacy, almost all reported cases with paraplegia have been treated with surgery with or without radiation therapy. We hereby report a case of thalassemia intermedia with paraplegia treated successfully with radiotherapy.

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Introduction

Thalassemia intermedia includes a wide spectrum of disorders with defective synthesis of the globin moiety of hemoglobin. Extramedullary hematopoiesis (EMH) is a compensatory phenomenon and commonly involves the liver, spleen and lymph nodes. Spinal cord compression due to EMH is an extremely rare complication. Only isolated reports have described patients with complete paraplegia, none of whom have been treated with radiotherapy alone. We present a case of a patient of thalassemia intermedia with paraplegia treated successfully with radiotherapy alone.

Case report

A 27-year-old male presented with a 12 month history of parasthesias and weakness of both lower limbs, progressive difficulty in ambulation and sphincter disturbances. He was diagnosed as case of thalassemia at the age of 2 years when he presented with a hemoglobin level of 6.8 g/dL, following a febrile episode. Through the first decade of life, he maintained a hemoglobin level of 9-10 g/dL without blood transfusions which was attempted only once at the age of seven years and abandoned due to acute transfusion related reactions, details of which were not available. In the second decade of life his hemoglobin level was maintained at 6-7 g/dL. He also had a history of recurrent fractures, either spontaneous or following trivial trauma, involving long bones of the extremities from the age of three years. Sites of fracture were the right tibia thrice (at the ages of 3, 25 and 26 years), right ulna twice (at age of 10 and 18 years), left radius at the age of 7 years and right tibia and fibula simultaneously at the age of 27 years.

Physical examination revealed a young male with a height of 158 cm and weighing 38 kgs with poor muscular development. He had characteristic facies with frontal bossing, prominent malar prominences due to maxillary hypertrophy and depression of nasal bridge. He was pale and icteric. Liver and spleen were enlarged, 5 and 6 cm below costal margin respectively. Higher mental functions and cranial nerve examinations were normal. Motor system examination revealed increased tone in lower limb muscles with complete loss of power (grade 0/5) in all muscle groups of the right and proximal muscles of the left lower extremity. Power in the distal muscle groups of the left lower limb was 1/5. Deep tendon reflexes were exaggerated in both lower limbs with bilater-
Currently, it can be recommended only in cases of paraplegia due to EMH. Of the 56 cases, 42 (34 males and 8 females) were patients of thalassemia none of whom had paraplegia with complete motor and sensory loss. There were 5 cases with complete paraplegia, 4 associated with polycythemia vera and 1 with sickle cell anemia. 4 of these cases were treated with surgical decompression with or without radiotherapy and transfusions but none of them showed any recovery. A 52-year-old male with polycythemia vera was treated with external radiation alone and showed good partial recovery. In cases where radiation therapy as a sole modality fails to control the symptoms, surgical decompression with post-interventional radiation therapy can achieve good results.

EMH in thalassemia has been treated with transfusion therapy with the rationale that correction of anemia would downregulate erythropoietin and lead to reversal of EMH. However, improvement is usually incomplete and short lived. Currently, it can be recommended only in cases of mild spinal cord compression or in special cases like pregnant patients where it may obviate the need for surgery or RT. It is useful as an adjunct to surgery and RT. A few authors have reported good results with the use of hydroxyurea along with hypertransfusion. The drug, in addition to its cytostatic effects, has a favorable effect on foetal hemoglobin production. Gamberini et al. treated a 24 year old patient of thalassemia intermedia with paraplegia due to EMH, with hydroxyurea 1000 mg/day for 5 months followed by 500 mg/day for up to 25 months. Neurological improvement occurred in 6 weeks but symptoms recurred in 5 months and were managed with radiotherapy. Since our patient showed 97.4% foetal hemoglobin on electrophoresis, hydroxyurea was not considered.

Our patient showed rapid and near complete recovery with radiation therapy alone despite having long standing paraparesis and paraplegia. Therefore, we conclude that radiation therapy may be the optimal therapeutic approach in such cases.

Conclusions
In cases of paraplegia due to EMH, there seems to be a bias towards surgical decompression with the aim of causing rapid decompression. Surgery may be associated with various complications including bleeding, hemodynamic instability, spinal instability etc. Radiation therapy is a simple, safe and effective approach for the treatment of spastic paraplegia consequent to spinal cord compression due to EMH. Fears of neurological deterioration due to radiation induced edema remain unfounded. Recovery of neurological function is rapid.
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References