Case Report

A 20 year old female was referred to the Shahid Baghaei hospital for the investigation of paraplegia. The patient had back and leg pain, and leg weakness . She was unable to climb stairs and had limitation of movement. No history of blood transfusion was found.

Cont……

Physical examination revealed a mild pallor and jaundice liver and spleen was palpable 3 cm below the costal margin.

Neurological examination showed hypoesthesia to touch and pinprick with thoracic sensory level on both sides, motor strength was decreased in the lower extremities, symmetrically. Tendon reflexes were hypoactive and symmetrical. Chest examination was normal.

Conti.....

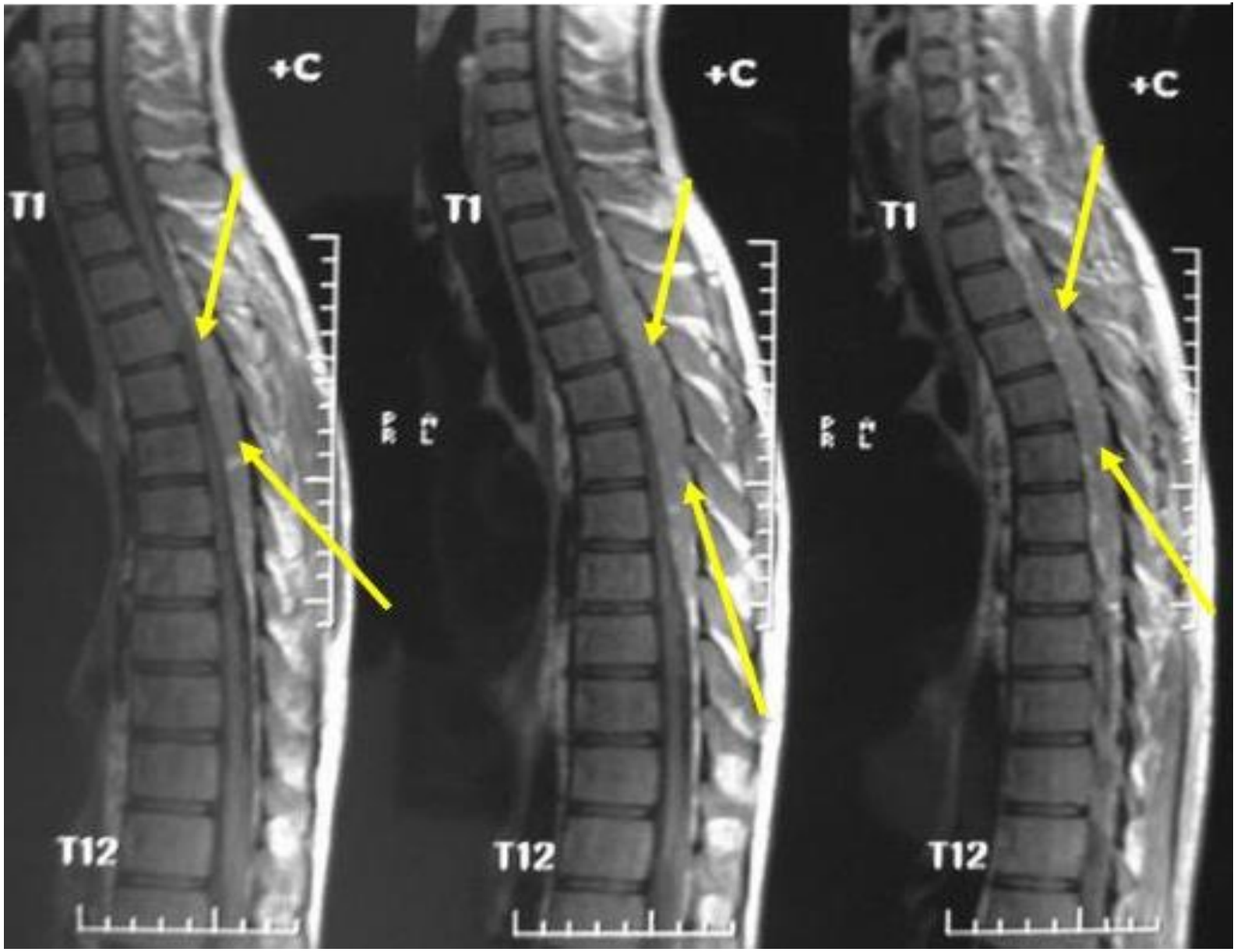
Hematological examination showed:

Hb: 9.5 g/dL, HCT:32.2%, reticulocyte count : 6%, MCV: 68 fL, MCH:19.8 pg, MCHC:30.6 g/dL, RBC count : WBC:6500, and platelet count : 320000.

Serum biochemical findings were as follows:

total bilirubin: 4.2 mmol/dL, ferritin:850 ng/mL.

Hemoglobin electrophoresis showed an HbA1 : 0%, HbF : 96% and HbA2 : 4%.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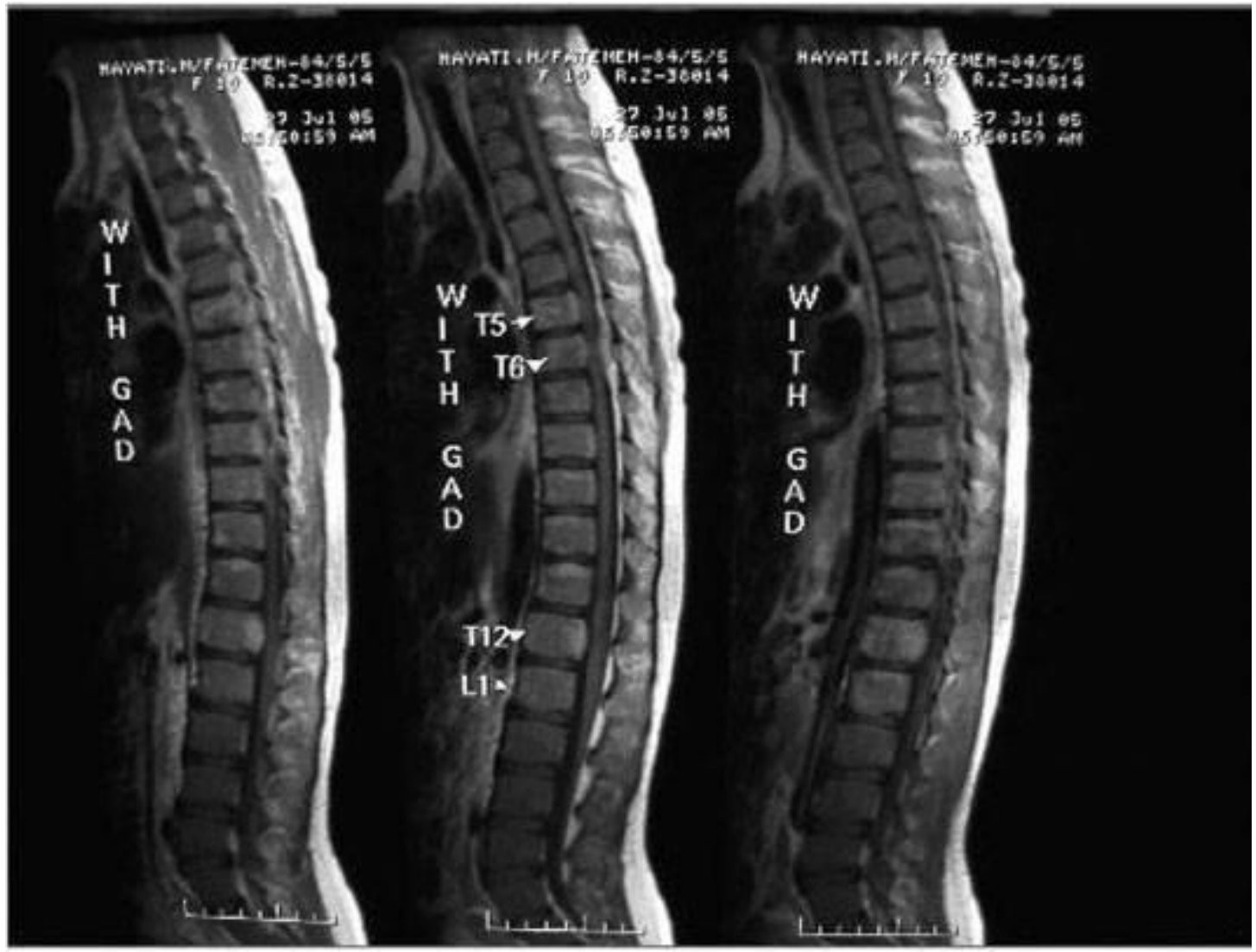
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Questions

You have experience of EMH in other organs?

You have experience of EMH in other diseases?

Treatment

Surgical decompression

Radiation

Transfusion

HU

Case Report

A 26-year-old female with Beta-thalassemia Intermediate group O, Rh positive was admitted to the hospital with symptoms of back pain and dark urine. She was on occasional blood transfusion regimen.

She had History of blood transfusion in 5 days ago, when she was transfused with two units of red blood cells and her Hb increased from 6 to 8.5g/dl.

Conti……

At admission ,the patient experienced fever, back pain, On examination, she was severely pale and had moderate splenomegaly. Lab findings were as follow:

Hb:4.5g/dl,MCV:70 FL ,MCH:20pg
total bilirubin, 12.6 mg/dL, Direct billi:0.5. 150 nucleated RBCs per 100 WBC.
The urine showed: 4+hemoglobin
Direct & indirect coombs test were positive..

Anti K & antic was positive.

Conti…….

Blood bank reported difficulty in cross matching of blood.

She was treated with methylprednisolone and IVIG and was discharged with a hemoglobin of 6.5g/dl. One week later, she came with Hemoglobin of 5g/dl. she received another dose of methylprednisolone, and IVIG and discharged with oral prednisolone.

Conti…….

Alloimmunization to red cell antigens is a known complication that occurs in patients with hemoglobinopathies who receive regular transfusions.

The risk of alloimmunization ranges from 5 % to 32%.

Anti-E, anti-C, and anti-Kell alloantibodies account for majority of cases. About 5%–10% of patients can present with alloantibodies against rare red cell antigens or unidentified antigens.

Conti.....

The risk of alloimmunization is much higher in patients with SCD and TI.

The risk of alloimmunization is higher in patients who received their first blood transfusion after the age of three.

Extended red cell antigen typing for D,d, C, c, E, e, and Kell antigens before the first blood transfusion is essential.

prophylactic extended antigen matching policy is cost benefit for blood Banking.

Case Presentation

A 34-year-old female patient known to have thalassemia Intermediate had been followed up in our clinic since 5 years old.

The last lab findings was as follow:

Hb:8-11g/dl, RBC :4.5-5.1,HCT;31-39,Platelet:165000-640000 (splenectomy was done at 12 yrs old),Total Billirubin:4.5,Direct:0.5,AlT:35, AST:40, FBS:140(the other endocrine tests were normal) ,BUN:20,Cr:0.9, Ferritin:650-1200ng/ml

MRI: Heat: normal, Liver: moderate Iron Overload

The case was a naïvely TI patient.

Conti…….

What is the best method to assess the Iron overload in this patient.

What is the cutoff value for serum ferritin to start treatment.

If serum ferritin is below the cutoff value for starting treatment, MRI shows a greater increase in iron load which is preferable to start treatment.

How can we evaluate and follow the treatment?

Which treatment policy do we support continues (non stop) or intermittent?

Should be paid more attention to the side effects of the iron chelator drugs compared to treatment in patients with thalassemia major?

Case Presentation

CASE PRESENTATION •

A 25-year-old female was diagnosed as a case of thalassemia intermediate since childhood on occasional blood transfusion and iron chelation therapy admitted to the thalassemia ward of Shahid Baghaei Hospital with severe abdominal pain.

Past history was significant for splenectomy two weeks ago for massive splenomegaly causing abdominal discomfort and hypersplenism.

On examination, the patient was pale, Jaundiced, not tachypneic, cardiorespiratory examination was unremarkable.

Abdominal examination revealed midline scar, no ascites, no tenderness, and hepatomegaly 3 cm below costal margin.

Conti…….

Lab findings was as follow:

RBC :4.5

Hb:8.5g/dl

HCT;26

WBC:15000

Platelet:1100000

Total Billirubin:4,Direct:0.5,AlT:55, AST:40, FBS:100,BUN:20,Cr:1, Ferritin:2000ng/ml

Uric acid:9

Ca:8,p:4

PT/PTT/INR –Normal

Conti.....

U/S and Doppler study, :Hepatomegaly, spleen not seen; enlarged portal vein contain thrombus extending to the superior mesenteric vein and splenic vein, mild ascites.

Upper GI endoscopy showed no esophageal varices

Questions

What are the risk factors for thrombosis in this patient?

What is the preventive treatment for thrombosis in this patient?

What is the appropriate treatment for the patient?