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Case report

Spontaneous iliopsoas hematoma in a trasfusion dependent β-thalassemia patient with hypersplenism: a case report

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Summary. A 27-year-old married man with transfusion dependent β-thalassemia (TDT) complaining low back pain due to a spontaneous iliopsoas hematoma is reported. A magnetic resonance imaging (MRI) confirmed the diagnosis.The patient was managed conservatively. The mechanism of spontaneous iliopsoas hematoma was unclear, although tearing of muscle fibers, unrecognized minor trauma, low platelet count, secondary to hypersplenism, and severe liver iron overload, associated to abnormalities of clotting factors synthesis, were the suspected etiologies. He showed a good response to treatment and was discharged home 11 days later. A new MRI, performed 7 months later, showed a complete resolution of hematoma. Although iliopsoas haematoma is an uncommon complication in patients with TDT, it should be considered in the differential diagnosis of a patient with back pain. (www.actabiomedica.it)

Key words: iliopsoas hematoma, trasfusion dependent β-thalassemia, diagnosis, treatment. hypersplenism

Introduction

β-thalassemia is an inherited hemoglobinopathy caused by β-globin gene mutations that impair the production of one or both β-globin chains. Distinction between the various phenotypes of β-thalassemia relies primarily on the clinical severity of the disease, which should be assessed both at initial presentation and over a period of close follow-up (1). Transfusion-dependent β-thalassemia (TDT) patients commonly present to our clinics in early childhood with severe anemia that requires life-long regular transfusion therapy for survival. On the other hand, non-transfusion-dependent β-thalassemia (NTDT) patients, usually present later in childhood or even in adulthood with mild/moderate anemia which only requires occasional or short-course regular transfusions in certain clinical settings (2).

The physical findings of TDT are related to severe anemia, ineffective erythropoiesis, extramedullary hematopoiesis, and iron overload resulting from transfusion and increased iron absorption. Manifestations generally include anemia, jaundice, pigment stones due to lifelong hemolytic state, skeletal changes secondary to erythroid hyperplasia with intramedullary expansion and cortical bone thinning, hepatosplenomegaly, heart failure and arrhythmia related to either severe anemia or iron overload. Iron overload causes clinical problems like those observed with primary hemochromatosis (eg, endocrine dysfunction, liver dysfunction, cardiac dysfunction) (3).

The iliopsoas muscle compartment can be involved by many different disease processes, including infection, tumor, and hemorrhage. Patients may present with a wide variety of symptoms that are often nonspecific, resulting in a delay in diagnosis. Spontaneous haematomas of the iliac psoas muscle are rare lesions that occur, most often, in patients receiving anticoagulant agents or in patients with either inherited or acquired clotting disorders (4). Furthermore, the iliopsoas compartment may become injured during trauma, percutaneous instrumentation, laparoscopic or open surgical procedures and extension from adjacent bleeding organs and vessels (5). Liver cirrhosis as a cause of spontaneous iliopsoas haematoma has also been reported (6). There is one case report of a haematoma of the iliopsoas muscle from thrombocytopenia resulting from the administration of a third generation cephalosporin (7).

We report a rare case of spontaneous iliopsoas haematoma in a male patient with TDT, diagnosed by magnetic resonance imaging (MRI). The patient responded well to a conservative treatment.

Case report

A 27-year-old married man with TDT, while travelling abroad complained low back pain requiring a hospital treatment with subcutaneous morphine and blood transfusions (2 units of packed red blood cells) because of a severe anemia. After hospital discharge, the patient came back immediately home, but due to the back pain recurrence he was referred to our hospital. He did not have a history of a similar complaint in the past and no personal or family history of bleeding disorders. There was no history of fall or trauma, fever or abdominal pain, motor or sensory complaints. He referred an intact bowel and bladder functions.

The clinical examination, two days after the symptoms appearance, was remarkable only for positive straight leg raising test on right side and hepatosplenomegaly. He was discharged after pain management with a plan to do MRI of spine in our outpatient department. Due to worsening of his back pain, which radiated to right groin, and the "aspect" of right thigh, he returned to the hospital after a few hours. On admission, his weight was 56 kg, height 161 cm, vital signs were stable, but he was in severe pain, lying in bed with the right leg flexed. Physical examination revealed that he had tender fullness over the right iliac crest region compared to the left side. Tenderness was also noted on right upper gluteal area. Passive motion of the hip aggravated his pain, but there were no focal neurological deficits.

Initial laboratory tests showed were significant for white blood cells (WBC) count: 2.4x10^3/uL (reference range: 4-10^3/μL), platelets count (Plt): 110x10^3/uL (reference range: 150-400x10^3/μL), hemoglonin (Hb): 6.5 g/dL (reference range: 13-17 g/dL), International Normalised Ratio (INR): 1.5 (normal range for a healthy person 0.8-1.2), activated partial thromboplastin time (aPTT): 41 seconds (reference range: 9.4-12.5 sec), D Dimer: 0.98 mg/L fibrinogen-equivalent units (FEU) (reference range: 0-0.49 mg/L), fibrinogen: 5.3 g/L (reference range: 2-4.1g/L). Clotting factor assay showed a reduced level of factor VII: 25% (reference range: 50 to 150), and biochemistry was significant for indirect hyperbilirubinemia and mildly increase of aspartate aminotransferase (AST): 45 U/L (reference range: 0-34 U/L). No endocrine complications were documented.

He was regularly transfused with red blood cell concentrate and on treatment with deferasirox (35 mg/kg per day). His last serum ferritin level was 3.247 ng/mL (normal levels: 23-175 ng/mL) An extremely high liver iron content (LIC) measured, five months before, by FerriScan® (8) was found: 39.9 mg/g dry liver. Four classes of LIC have been reported in thalassemic patients: Class 1=normal LIC <3 mg Fe/g dry liver, Class 2=mild overload LIC 3-7 mg Fe/g dry liver, Class 3=moderate LIC overload 7-15 mg Fe/g dry liver, and Class 4=severe LIC overload ≥15 mg Fe/g dry liver (3).

Magnetic resonance imaging (MRI) showed a hematoma in the right iliopsoas muscle extending to the upper thigh. The calculated size of the lesion in all imaging modalities was approximately 12x5x2.5 cm (Figure 1).

He was managed conservatively with transfusion of red blood cell concentrate, analgesics, vitamin K, steroids, bed rest and physiotherapy. He showed a good response to treatment and was discharged home 11 days later. A new MRI, performed 7 months later, showed a complete resolution of the iliopsoas hematoma (Figure 2).

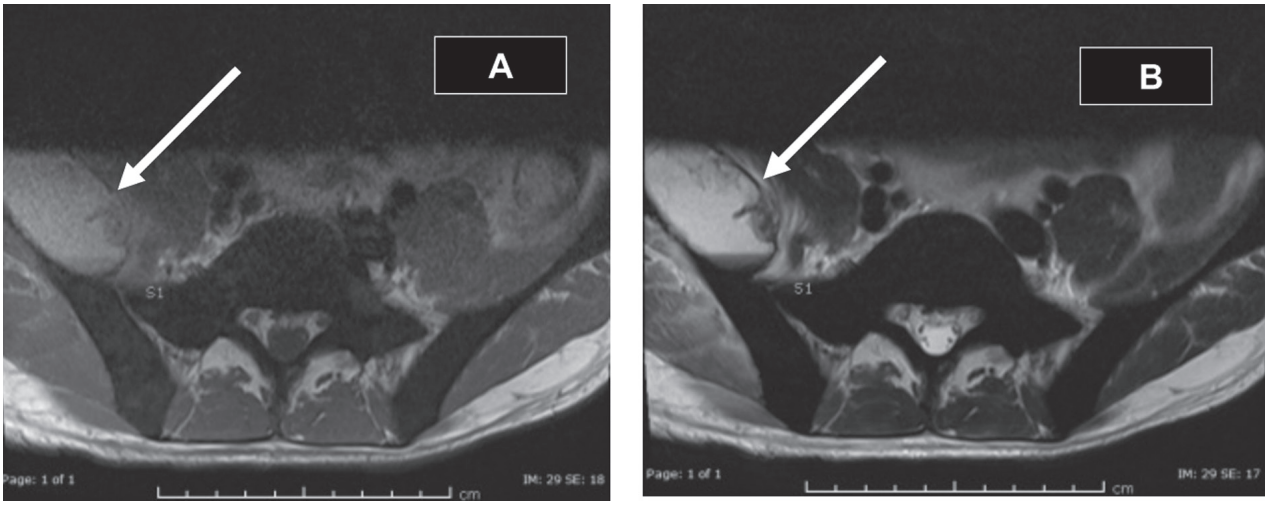

Figure 1 A-B. Axial T1 and T2 WI of the spine at S1 level. Images show a large diffuse hematoma in the right iliopsoas muscle (white arrows)

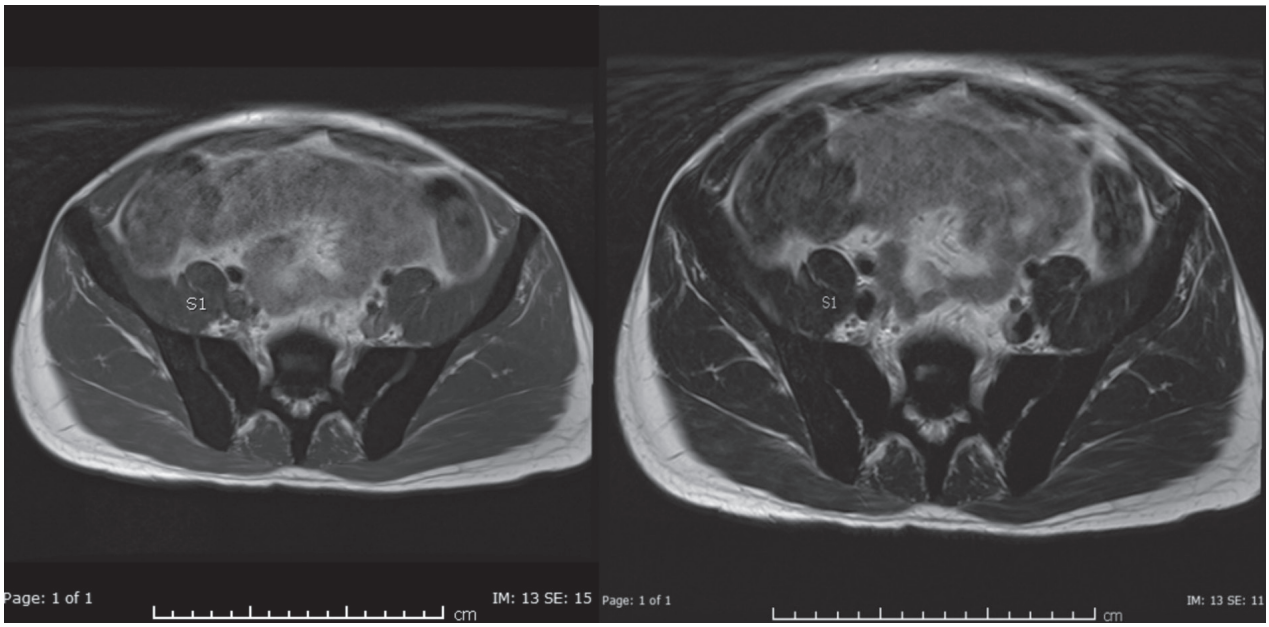

Figure 2. Axial T1 and T2 WI of the spine at S1 level, after 7 months, showing complete resolution of the previous right iliopsoas hematoma

Discussion

Even though iliopsoas hematomas are well-described in the literature, their incidence remains uncommon. Iliopsoas hematomas are typically caused by trauma in patients on anticoagulation/ antiplatelet therapy or in those with hemophilia.

To the best of our knowledge, this is the first reported case of spontaneous iliopsoas hematoma occurring in a patient with TDT. The mechanism of spontaneous iliopsoas hematoma was unclear, although tearing of muscle fibers, unrecognized minor trauma, low platelet count, secondary to hypersplenism, and severe liver iron overload, associated to abnormalities of clotting factors synthesis, were the suspected etiologies.

A review of literature for spontaneous bleeding in patients with TDT revealed three cases of intracranial hemorrhage. Svahn et al. (9) reported a case of

subarachnoid hemorrhage observed during the postpartum period in a 27-year-old woman suffering from TDT. Brain MRI revealed a complex vascular abnormality. Lee (10) reported two cases of fatal intracranial hemorrhage in children with TDT, aged 7and 12 years. According to the author, the bleeding was likely multifactorial due to recent blood transfusion, prolonged prothrombin time, partial thromboplastin time and reduced platelet count.

The iliopsoas muscle is formed by 2 sections (the psoas and the iliacus) and runs from the retroperitoneum through the pelvis into the thigh (11). When a hematoma of the iliopsoas occurs, the main symptoms are pain and functional impairment. However, owing to the anatomical proximity of these muscles to the lumbar plexus and the femoral nerve, this can be a source of neurological dysfunction, through the compression of the former or, more frequently, the latter (12,13). The clinical presentation of iliopsoas hematomas is often that of sudden-onset low back pain or severe persistent pain in the lower abdominal quadrants, as well as inguinal region, and radiate to the anterior, medial, or lateral aspects of the lower extremities. Patients also describe using leg flexion to try to relieve pain on the involved side (psoas sign). The differential diagnosis for back pain is extensive and includes pancreatitis, ureteric colic, lumbar spondylosis, aortic dissection, and musculoskeletal pain (14). A careful physical examination is important but often not specific. Besides clinical examination, there are several imaging modalities that can aid in establishing the diagnosis. Ultrasonography has been used to diagnose iliopsoas hematoma but its sensitivity and specificity are user-dependent, rendering the results potentially less reliable. Computed tomography (CT) has a high degree of sensitivity and is the most commonly utilized test for the diagnosis. However, MRI remains the investigation modality of choice due to its high sensitivity and specificity to identify the site and the extent of bleed (14).

The optimal treatment of iliopsoas haematomas remains controversial, as current evidence favours neither the conservative management nor surgical or percutaneous drainage. All these therapeutic measures had good outcomes in previously published case reports. Generally, a conservative approach is chosen in smaller haematomas in haemodynamically stable patients with slight-to-moderate neurological impairment, and a drainage procedure is the option in larger haematomas with severe neurological impairment and/or haemodynamic instability (15).

Conclusions

Iliopsoas hematoma has a variety of clinical manifestations. The most common symptom is sudden onset of lower back and flank pain. Besides clinical examination, there are several imaging modalities that can aid in establishing the diagnosis. Delay in diagnosis can lead to inappropriate initial treatment and, in some cases, serious complications. There is no clear strategy for treatment of iliopsoas hematoma yet, and a variable treatment plan may be used according to the patient's condition. Physicians should be aware of such potential complication. A high clinical suspicion should be designated to a patients with a sudden onset of back pain. Close monitoring of patient is important for an early diagnosis and for avoiding an inappropriate treatment.

Conflict of interest: None to declare

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