Long-term outcome of an unusual haemophilic pseudotumour

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Summary
Haemophilia, a lifelong congenital bleeding disease, is a highly demanding disorder, due to the costs of its replacement therapy. In the absence of this pivotal treatment, life expectancy and quality of life are deleteriously affected.

As illustration, we present a 14 years long follow-up of a patient with severe haemophilia A, treated sporadically with fresh plasma, cryoprecipitate and factor concentrates, who developed a giant iliopsoas pseudotumour. Since he was an infant, under on demand therapy with cryoprecipitate and factor VIII, he presented many spontaneous bleedings, developing multiple disabling arthropathies. At the age of 14 years, an iliopsoas hematoma occurred, which relapsed several times, developing an iliopsoas pseudotumour. After 5 years, sepsis with Klebsiella was diagnosed. A CT scan revealed fistula between the pseudotumor and the gut. Under antibiotics, the evolution of sepsis improved, but over a period of 10 months 5 episodes of haematemesis and melena, followed by one episode of macroscopic haematuria occurred; two months later he developed an inguino-crural mass, which fistulized through the abdominal wall. A mixt german-romanian team solved the clinical concern. After 108 hospitalization days and consumption of 104 840 IU factor VIII he left the clinic in good condition. One year later, the temporary colostomy with anus praeter was closed. The follow-up reveals now, after almost 10 years with favourable outcome, that the patient is well, active within his family and profession.

Schlüsselwörter
Hämophilie, iliopsoas-Pseudotumor, Fistula

Zusammenfassung

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Case report

We present a case with a history of an iliopsoas pseudotumour (IPT), followed-up for 14 years. MA is a man (age 38 years) with severe haemophilia A without inhibitors, sporadically treated with fresh frozen plasma, cryoprecipitate and low doses of factor VIII (FVIII) concentrates. Since infancy, he had many spontaneous bleedings, developing multiple disabling arthropathies. At the age of 24 (July 1997), his first iliopsoas haematoma (IPH) occurred. In April 1998 it relapsed in the form of a massive left IPH with secondary haemorrhagic shock. Surgical removal and drainage was attempted. Under the scarce replacement therapy, it relapsed several times, with development of a left IPT, requiring several hospitalizations and blood transfusions.

Since March 2002, he presented a worsening of the clinical condition with prolonged
fever, 13 kg weight loss in about 4 weeks, diarrhea and pain at the left iliac fossa. Sepsis with Klebsiella was diagnosed. Under prolonged intermittently adapted antibiotherapy the sepsis improved, but five episodes of haematemesis and melena occurred over the following five months (April-August 2002). Abdominal computed tomography (CT) revealed in November 2002 a fistula between the haematoma and bowel (Fig. 1).

In December 2002, he also presented an episode of macroscopic haematuria. In February 2003, he developed a left inguino-crural mass, which opened at the abdominal wall (Fig. 2). This was a threatening condition, suggesting the need of a life-saving intervention under an adequate replacement with FVIII. In March 2003 a diversion colostomy and anus praeter were performed; retroperitoneal and thigh long-term drainages were inserted, with initially > 1 l bloody pus discharge. The drainage was intermittently repositioned until it was discontinued in June 2003. After 108 days of hospitalization and consumption of 104840 IU FVIII, the patient left the clinic in good health. One year later, the colostomy was closed.

The follow-up over almost 10 years revealed a favourable outcome: without relapse of IPT, sepsis, digestive or urinary bleeding. A recent CT scan revealed no residual mass in the region of the original PT. The patient is active, employed, but with multiple chronic arthropathies.

Discussion

HPT is a well-organized, encapsulated blood mass, progressively expanding, compressing or eroding local tissues (3). First described in 1918 by Starker (4) it represents a severe complication of haemophilia, with a prevalence highly depending on the quality of replacement therapy (5, 6). Proximal intraabdominal PTs are mainly localized within the iliopsoas muscle. Their fistulation into the bowel and externalization on the abdominal wall are exceptionally reported (4, 5). Percutaneous aspiration or drainage attempts may predispose to this type of complications. The left colon, as it was in our case, is an atypical place for fistula formation (5). Fecal contamination leads to intraabdominal abscess development, with bleeding tendency and risk of neural palsy or hydronephrosis/haematuria. Sepsis is life-threatening in the situation of a persisting communication between PT and bowel.

Management of such a complicated condition is most challenging, with a mortality exceeding 20%. Early diagnosis with conservative approach consisting of generous substitution with factor concentrates is the most simple and safe treatment. Nevertheless, a residual PT represents a permanent risk for bleeding, infection or necrosis (6–8).

Surgical removal remains the treatment of choice (7). It must be undertaken by a multidisciplinary team in a specialized center. But, complete resection of a haemophilic PT may often not be possible (8). A local condition as described in our patient, characterized by coexistence of encapsulated cystic PT, abscess, extensive intra-abdominal adherences, fibrous fistula tract, entero-cutaneous fistula, may hinder a radical surgical excision. Fortunately, our patient is witnessing that even in desperate situations, minimal invasive surgery associated to replacement therapy can assure a long-term favourable outcome.

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Conflict of interest

The authors declare, that there is no conflict of interest.

References