



Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

# Bilateral iliopsoas haemophilic “soft tissue pseudotumours”: A case report



Achmad Fauzi Kamal\*, Ananto Satya Pradana, Yogi Prabowo

Department of Orthopaedic and Traumatology Ciptomangunkusumo National Central Hospital/Faculty of Medicine Universitas Indonesia, Jakarta, Indonesia

## ARTICLE INFO

### Article history:

Received 22 December 2014  
Received in revised form 7 May 2015  
Accepted 8 May 2015  
Available online 6 June 2015

### Keywords:

Haemophilic pseudotumour  
Iliopsoas  
Haemophilia

## ABSTRACT

Haemophilic soft tissue pseudotumour is one of the rarest complications of haemophilia that caused by repetitive bleeding resulting in an encapsulated mass of clotted blood and necrotic tissue. Soft tissue pseudotumour may not only cause flexion contracture but also chronic pain and femoral nerve compression that cause severe disability. Thus, surgical excision is the treatment of choice. It should only be carried out in a major haemophilic center by an integrated multidisciplinary surgical team.

© 2015 The Authors. Published by Elsevier Ltd. on behalf of Surgical Associates Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Muscle haematoma (“soft tissue pseudotumour”) represent 10–25% of all bleeds in patients with severe haemophilia. Iliopsoas is the commonest site (55%) which could develop into a pseudotumour [1]. If it is not treated properly, the pseudotumour may reach a enormous size, causing pressure on the adjacent neurovascular structures and result in severe disability [2]. This paper reports one case of haemophilic pseudotumour that occurred in iliopsoas muscle.

## 2. Case illustration

A 22 year old–haemophilic man came to our hospital with chief complain of painful mass in his right lower abdomen since one month prior to admission. The pain was sharp, continuous, worsened with activity, and was not relieved with analgesics. This condition appear after the patient fell down in sitting position. Subsequently, he also felt progressing paraesthesia and weakness on both lower extremities which caused him to wheelchair bound. The patient had had recurrent haemorrhage from his gum and also a haematoma at knee region previously. He already been diagnosed with severe haemophilia since 3 month of age with the level of haemophilic factor 0.85%.

Physical examination showed the mass in right lower quadrant of abdominal region. It was firm, with smooth overlying skin, marked tenderness, and ill-defined margin. There were bilateral hip flexion deformities because of pain (Fig. 1). Anaesthesia was found from upper medial thigh to his medial right lower leg. Hypoaesthesia and paraesthesia were also found in the left side. Motoric functions of both lower extremities were decrease, varying from grade 2/5 to 4/5 (weakness of the right hip flexion with motoric strength score 4 and knee extension and ankle dorso flexion with motoric strength score 2).

Laboratory finding showed haemoglobin level of 6.5 g/dL (12–15 g/dL), haematocrit 20.8% volume (36–46% volume), white blood cell  $9510/\text{mm}^3$  ( $5000\text{--}10000/\text{mm}^3$ ), platelet  $307 \times 10^3/\text{mm}^3$  ( $150\text{--}400/\text{mm}^3$ ), prothrombin time 11.7 s (control: 11.3 s), activated partial thromboplastin time 48 s (control: 17 s) and levels of factor VIII 45.7% (50–150%).

Computed tomography (CT) scan showed heterogenic density masses at right and left retroperitoneum. Magnetic resonance imaging (MRI) showed heterogenic intensity lesions with clear border, multiple septation, sized  $12 \times 12.8 \times 19.4$  cm in right psoas major muscle at the level of L2–L3 vertebral body that shifted the quadratus lumborum muscle to posterolateral, and slightly shift the kidney and bowel to the superior. There was also mass with the same intensity in left pelvic region, sized  $4.2 \times 4.7 \times 5.9$  cm that shifted left psoas muscle slight anteriorly (Fig. 2).

The patient was diagnosed as bilateral haemophilic pseudotumour of bilateral iliopsoas muscle and treated with factor VIII concentrate 1500 units b.i.d for 2 weeks, followed by 1000 units two times a week as maintenance dose. After series of discussions at the meeting of haemophilia integrated service in our hospital, it

\* Corresponding author at: Department of Orthopaedic and Traumatology Cipto Mangunkusumo Hospital-Faculty of Medicine Universitas Indonesia, Jl. Diponegoro No. 71, Jakarta Pusat, Jakarta, Indonesia. Fax: +62 21 3905894.

E-mail address: [fauzikamal@yahoo.com](mailto:fauzikamal@yahoo.com) (A.F. Kamal).



Fig. 1. Clinical picture showing mass in the right lower abdomen.

was decided to perform surgery for evacuation of the haematoma (excision) of the pseudotumour.

One hour before surgery patient was given factor VIII 3500 unit. The first incision was performed at right pseudotumour with the ilioinguinal approach, dissection was performed layer by layer. After the mass was seen, we opened the capsule and then we evacuated the haematoma (Fig. 3). The haematoma was dark red with soft and springy consistency. The volume of the haematoma was about 1.5 l, we excised the capsule until we met healthy tissue, and preserved the iliopsoas muscle as much as we could. Then fibrin glue (bioglu) was applied in the space that left and the wound was closed layer by layer leaving a drain. After that, the same procedure was done to the second pseudotumour at left iliopsoas muscle (Fig. 3). Postoperative histopathology showed haemorrhage, reactive osteoid, and fibrous tissue that confirmed the clinical diagnosis of haemophilic pseudotumour (Fig. 4). Postoperative rehabilitation was done by active–passive muscle strengthening to increase the muscle power of quadriceps femoris and decrease flexion contracture.

At three months follow up after surgery there was no recurrence and other significant complaint. The muscle power of his lower limb especially the left side improved to score 4/5 and he was able to stand up assisted for about 10–20 s (Fig. 5).

### 3. Discussion

The severity of bleeding in haemophilia is generally correlated with clotting factor level. In severe haemophilia, characterized by clotting factor level < 1 IU/dl (< 0.01 IU/ml) or < 1% of normal, spontaneous bleeding episode of joint or muscle is common [3]. Our patient had already been diagnosed with severe haemophilia. Thus, he has a tendency of spontaneous bleeding in joint or muscle which made an increase suspicion of muscle haematoma. Moreover, he did not routinely come to outpatient clinic so he did not routinely get transfusion of antihaemophilic factor.

A hemophilic pseudotumor is the result of repeated episodes of bleeding into a soft tissue, bleeding at a bone fracture site or as a result of sub-periosteal hemorrhage. Inadequate resorption of extravasated blood results in an encapsulated area of clotted blood and necrotic tissue. With successive hemorrhagic episodes, these lesions expand over time, eventually causing symptoms by mass effect [4].

Haemophilic haematoma or “soft tissue pseudotumour” is categorized into intramuscular and extramuscular (interfascial, subcutaneous) [5,6]. Intramuscular bleeding in haemophilia are generally associated with direct minor trauma or may also rise de novo [7]. The diagnosis of fresh intramuscular bleeding is usually very obvious due to the presence of pain, swelling, local warmth, and frequently an overlying bruise [2,7]. Recurrent and non-resolving soft-tissue bleeding may get organized and cause joint contractures (deformities) and soft tissue pseudotumours. The latter are most common in the thigh, gluteal region, and iliopsoas [5,8].

The most frequently affected muscle is the iliopsoas which classically presents with a painful flexion deformity of the hip. Any attempt to extend the hip result in an increase in the level of pain and compensatory lumbar lordosis. Therefore, it may also be associated with abdominal pain mimicking acute appendicitis with mass (peri-appendicular mass) or soft tissue tumor in the pelvic region. A bleeding into iliopsoas muscle (haematoma) has a similar clinical finding to an intraarticular bleeding into the hip joint [2]. The extent and nature of the haematoma (pseudotumour) should be assessed carefully by sonography, CT scan or MRI.

Sites of the muscle haematoma that are associated with neurovascular compromise (due to mass effect) include: iliopsoas muscle with the risk of femorocutaneous and femoral nerve palsy [7,8], gastrocnemius muscle with the risk of tibial nerve injury and muscle contracture leading to equinus deformity, and flexor mus-

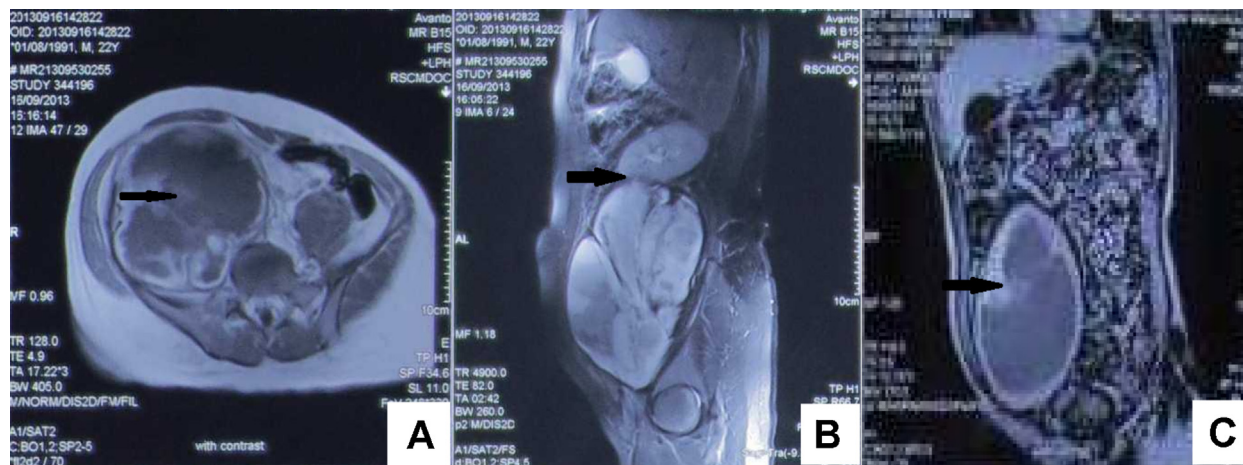
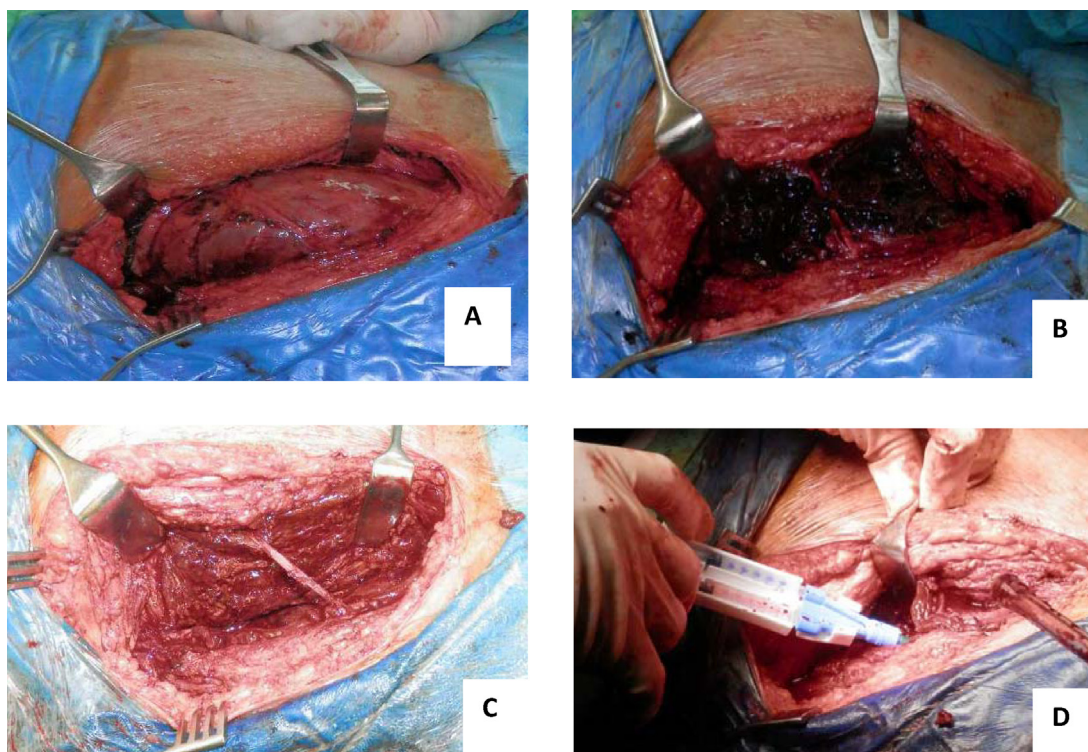
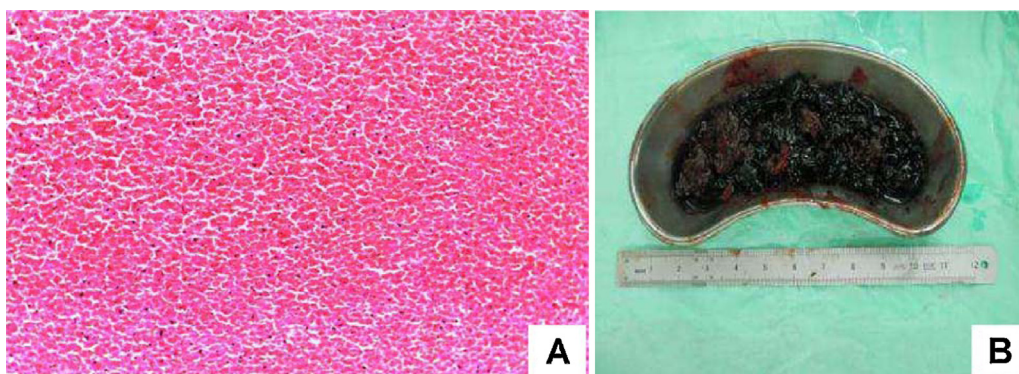


Fig. 2. (A) MRI T1-weighted image axial plane with contrast showed heterogenic intensity lesion with clear border and septation (arrow). (B) MRI T1-weighted image sagittal plane showed hyperintense heterogenic lesion with hypointense rim encircling the mass and slight cranial shift of right kidney and bowel (arrow). (C) MRI T1-weighted image coronal plane showing mass on right iliopsoas muscle.



**Fig. 3.** Intraoperative finding of the right mass (A, B) and left mass (C, D). (A) Exposed mass with right lateral femoral cutaneous nerve crossing the tumor. (B) Incision of the capsule showed dark red haematoma with soft and springy consistency. (C) After the evacuation of haematoma and excision of capsule, preservation of right lateral femoral cutaneous nerve and preservation of iliopsoas muscle. (D) Application of fibrin glue. (For interpretation of the references to color in the text as well as in the figure legend, the reader is referred to the web version of this article.)



**Fig. 4.** (A) Postoperative histopathology (haematoxylin eosin, 100× magnification) showing haemorrhage at the central of pseudotumour, no tumour cell was found. (B) Gross pathology of the mass excised.

cles of forearm with the risk of Volkmann’s ischemic contracture [9].

Bleeding into deeply situated muscles (pseudotumour) may be somewhat difficult to diagnose. Therefore, imaging procedures is needed to point toward certain diagnosis and role out the other ones. MRI and CT scan, which are obviously more precise than ultrasound, will demonstrate not only the size and distribution of soft tissue pseudotumour, but may also determine whether the contents are in liquid or in solid form. These procedures are preferred in providing accurate diagnosis of musculoskeletal manifestations of haemophilia including haemophilic arthropathy, pseudotumour, and other differential diagnoses compared to invasive procedures [2,6]. MRI is superior in demonstrating different stages of haemorrhage, haemosiderin/sclerotic rim, and in displaying the soft tissues condition. It is also able to accurately asses neurovascular involvement as well as monitoring the therapeutic response [4,6]. In this

patient, MRI showed different stages of haemorrhage and describe location of the lesions exactly.

Based on world federation of haemophilia guideline, the principle treatment of fresh intramuscular bleed is generally conservative and relies upon restoration of normal clotting by administration of factor replacement. The limb is rested in a position of function and adequate analgesia is provided. Subsequently, following the initial acute phase, physiotherapy should begin early in the rehabilitation in an attempt to restore full function [9].

However, there are a number of therapeutic modalities for haemophilic pseudotumour, such are conservative treatment, surgery, embolisation, and radiotherapy [10,11]. By conservative treatment [9,12], it may fail to prevent progression of the pseudotumour. Although regression is occasionally seen, there are no true cures and recurrence and/or progression is very common [2]. Surgery is indicated when the pseudotumour continues to



**Fig. 5.** After three months follow up, patient was able to stand up on his left foot with being assisted.

grow eventhough anti-haemophilic factor is replaced. Other indications include severe joint degeneration with disability, severe pain, haemophilic chronic synovitis, severe soft-tissue contractures, deformity requiring bone osteotomy, and non-functioning extremity or the presence of chronic infection [10].

An iliopsoas bleeding usually does not respond promptly to the administration of factor concentrate and a flexion deformity of the hip may persist for several weeks [2], like in our patient. Soft tissue pseudotumour in this case not only cause hip flexion deformities but also severe pain and signs of femoral nerve compression that result in disability. Thus, surgery is indicated.

Percutaneous aspiration as an easy and relatively non-invasive procedure is not suitable in our case because of the location of pseudotumours. It is indicated for pseudotumours which are located more peripherally, for small pseudotumours measuring <3 cm in diameter and for the pseudotumours with liquid content [13]. So, surgical excision is the treatment of choice and should only be carried out in a major haemophilic centre by an integrated multidisciplinary team [14,15].

#### 4. Conclusion

Iliopsoas pseudotumour of haemophilia is a rare complication of haemophilia. It may cause painful mass, deformities and disability of the patient. Without adequate management, iliopsoas pseudotumour may cause pressure on surrounding tissue

including neurovascular structures and bone. There are a number of therapeutic modalities for haemophilic pseudotumour, such as conservative and surgical treatment. Surgical management is indicated in progressive enlargement of the haematoma, haemodynamic deterioration, or occurrence of a complication. However, further studies with larger subjects are needed. Adequate rehabilitation and routine exercise is important to get the best result patient rehabilitation.

#### Conflict of interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

#### Sources of funding

Private funding by the authors.

#### Ethical approval

This is a case report, patient' written consent has been obtained and will we available on request.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

AFK and YP write the paper, performed the operation and scientific analysis. ASP and KH performed the documentation of the patient and follow up care, assisted in operation and draft writing.

#### Guarantor

AFK is the Guarantor.

#### References

- [1] R. Beyer, J. Ingerslev, B. Sorensen, Current practice in management of muscle haematoma in patients with severe haemophilia, *Haemophilia* 16 (6) (2010) 926–931.
- [2] E.C. Rodriguez-Merchan, N.J. Goddard, Muscular bleeding, haematomas, and pseudotumours, in: E.C. Rodriguez-merchan, N.J. Goddard, C.A. Lee (Eds.), *Musculoskeletal Aspects of Haemophilia*, Blackwell Science, Oxford, 2000, pp. 85–90.
- [3] M. Gilbert, Pseudotumours (haemophilic blood cysts), in: C.D. Forbes, L.M. Aledort, R. Madhok (Eds.), *Haemophilia*, Chapman & Hall, London, 1997, pp. 123–130.
- [4] A. Pakala, J. Thomas, P. Comp, Haemophilic pseudotumour: a case report and review of literature, *Inter. J. Clin. Med.* 3 (3) (2012) 229–233.
- [5] S. Jaovisidha, K.N. Ryu, J. Hodler, M.E. Schweitzer, D.J. Sartoris, D. Resnick, Haemophilic pseudotumour: spectrum of MR findings, *Skeletal Radiol.* 26 (8) (1997) 468–474.
- [6] S. Jaganathan, S. Gamanagatti, A. Goyal, Musculoskeletal manifestations of haemophilia: imaging features, *Curr. Probl. Diagn. Radiol.* 40 (5) (2011) 191–197.
- [7] A.K. Ahlberg, On the natural history of haemophilic pseudotumour, *J. Bone Joint Surg. Am.* 57 (8) (1975) 1133–1136.
- [8] J.S. Park, K.N. Ryu, Haemophilic pseudotumour involving the musculoskeletal system: spectrum of radiologic findings, *Am. J. Roentgenol.* 183 (1) (2004) 55–61.
- [9] A. Srivastava, A.K. Brewer, E.P. Mauser-Bunschoten, N.S. Key, S. Kitchen, A. Llinas, C.A. Ludlam, J.N. Mahlangu, K. Mulder, M.C. Poon, A. Street, Guidelines for the management of haemophilia, *Haemophilia* 19 (1) (2013) e1–47.
- [10] E.C. Rodriguez-Merchan, Musculoskeletal complications of hemophilia, *HSS J.* 6 (2010) 37–42.
- [11] R. Garcia-Perez, G. Torres-Salmeron, F. Sanchez-Bueno, A. Garcia-Lopez, P. Parrilla-Paricio, Intraabdominal hemophilic pseudotumor: case report, *Rev. Esp. Enferm. Dig.* 102 (2010) 275–280.

- [12] M.S. Gilbert, Surgical management of the adult haemophilic blood cyst (pseudotumours), in: E.C. Rodriguez-Merchan, N.J. Goddard, C.A. Lee (Eds.), *Musculoskeletal Aspects of Hemophilia*, Blackwell Science, Oxford, 2000, pp. 92–96.
- [13] S. Shaheen, E. Alasha, Haemophilic pseudotumour of the distal parts of the radius and ulna. A case report, *J. Bone Joint Surg. Am.* 87 (11) (2005) 2546–2549.
- [14] A.F. Kamal, L. Sukrisman, I.H. Dilogo, H. Priyamurti, M.N. Qomaruzzaman, Pelvic haemophilic pseudotumour: a case report, *J. Orthop. Surg.* 22 (2) (2014) 263–268.
- [15] D.V. Poenaru, J.M. Patrascu, B.C. Andor, I. Popa, Orthopaedic and surgical features in the management of patients with haemophilia, *Eur. J. Orthop. Surg. Traumatol.* 24 (5) (2014) 685–692.

#### Open Access

This article is published Open Access at [scimedirect.com](http://scimedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.