



Case report

Resection and total femoral prosthesis reconstruction treatment of massive femoral hemophilic pseudotumor: A case report

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ABSTRACT

Introduction and importance: Hemophilic pseudotumor is a rare and serious complication of hemophilia, often occurring in the long bones and muscle tissue of the extremities, with an incidence of about 1–2%. However, there is no effective surgical treatment for massive hemophilia pseudotumors of the extremities. Therefore, we would like to report this case to provide new ideas and methods for the treatment of massive hemophilic pseudotumors of the extremities through resection and total femoral prosthesis reconstruction.

Case presentation: After admission, the patient first underwent a month-long coagulation factor replacement therapy to maintain the patient's factor IX level at approximately 100%. Then, extensive resection of hemophilic pseudotumor and total femoral prosthesis reconstruction was performed in collaboration with several departments, and we collected the patient's preoperative, intraoperative and postoperative data for analysis.

Clinical discussion: After coagulation factor replacement therapy the patient's factor IX level was significantly increased and approached 100%. The surgical procedure was uneventful, and postoperative X-ray suggested a well-positioned prosthesis with postoperative pathological features consistent with a hemophilic pseudotumor. The patient was able to move around with the assistance of a walker 14 days after surgery, and there was no recurrence after one year of follow-up.

Conclusion: For treat the massive hemophilic pseudotumors of the extremities, extensive resection and prosthetic replacement with coagulation factor replacement therapy is a safe and effective treatment that can significantly improve the prognosis and quality of life of patients.

1. Introduction

Hemophilic pseudotumor is also known as hemophilic cyst which is a collection of encapsulated blood caused by recurrent extra-articular hemorrhage in soft tissues. The incidence of hemophilia pseudotumor in hemophilia patients is about 1–2%, while it increases to 10% in hemophilia patients with coagulation inhibitors [1–3]. Pseudotumors most often occurs in the long bones (especially the femur) and the pelvis in adults hemophilic patients, while in children with hemophilia pseudotumors usually affects the immature bones of limbs [4,5]. As the pseudotumor grows, it will cause progressive compression of the major nerve and vascular compression, fatal massive hemorrhage, pathological fractures and intestinal obstruction [6–9]. Although there has been extensive research on the treatment of hemophilia. However, due to the rarity of hemophilic pseudotumor, there is no standardized treatment guidelines for hemophilic pseudotumor. It has been reported that surgical resection and drainage [3,10,11], coagulation factor replacement

therapy [12,13], radiotherapy [12,14] and other non-surgical treatment [15] of hemophilic pseudotumor all have varying deficiencies. To our knowledge, the case we report is the first case of hemophilia pseudotumor treated with extensive pseudotumor resection and total femoral prosthesis reconstruction with better outcome.

2. Case presentation

A 48-year-old man with a history of mild hemophilia B for 20 years. 7 days before admission, the patient suffered severe left thigh pain and his left lower limb could not bearing weight after an accidental fall. At the age of 24, he was diagnosed with hemophilia B due to recurrent bleeding gums (Factor IX level 10%) but did not received any treatment. After the injury, he suffered severe pain and progressive swelling in his left thigh. When admission, his left thigh circumference was observed to be 22 m greater than the right thigh (Fig. 1a). X-ray examination confirmed a pathological fracture of the left femoral shaft accompanied

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by radiological lesions, cortical thinning, and bone destruction (Fig. 1b). Computer Tomography (CT) and Magnetic Resonance Imaging (MRI) scan revealed a large multicystic soft tissue mass in the left thigh with involvement of the medullary cavity and destruction of the bone cortex (Fig. 1c,d). Coagulation factor tests suggest that Factor IX is significantly lower than normal (2.6%). In summary, we diagnosed the patient with hemophilic pseudotumor.

After admission, the patient need absolute bed rest and traction therapy. Meanwhile, the coagulation factor replacement therapy was maintained for a month to keep Factor IX levels at about 100%. This level needs to be maintained especially in the preoperative and postoperative periods. After a month treatment, the pain was significantly relieved, but his left lower limb still could not bearing weight, and MRI showed that the soft tissue mass in the left thigh had no significant change. Thus, due to severe bone destruction, involvement of the bone marrow cavity, the large size of the pseudotumor and the patient's wishes the radical resection of pseudotumor, followed by reconstruction with a total femoral prosthesis was planned.

Prior to surgery, we ensure that the patient's Factor IX remained above 95%. Then During the surgery, the patient was placed in the right lateral decubitus position, a long, direct lateral incision was made, and fasciocutaneous flaps were created. It can be seen that the pseudotumor encompassed the entire vastus lateralis and intermedius (Fig. 2a). Subsequently, we dissected the fibrous capsule of the pseudotumor and cleared it of a large amount of blood clots and necrotic tissue (Fig. 2b).

Then, the rectus femoris and vastus medialis were separated from the pseudotumor, and the femoral neck and shaft are dissected above the supracondylar crest to facilitate resection of the femur (Fig. 2c). After en bloc resection of the pseudotumor, the proximal and distal femurs were removed subperiosteally. The proximal tibia was prepared to receive the rotating hinge prosthesis and fixed with antibiotic cement. Then, total femoral component was assembled and placed (Fig. 2d).

The postoperative course of the patient was stable without incision infection and pulmonary embolism. The coagulation factor replacement therapy was continued to keep the levels of Factor IX at approximately 80%. He was kept at complete bed rest for 14 days. Fourteen days after operation, the incision healed well and he then began weight-bearing as tolerated with abduction brace (Fig. 3a, b). Postoperative X-ray showed resection of the hemophilic pseudotumor and femur, and reconstruction with a custom total femoral replacement (Fig. 3c, d). The results of pathological examination showed fibroblasts, histiocytes, and hemosiderin pigment consistent with hemophilic pseudotumor (Fig. 3e, f).

The patient was discharged home from the hospital stable and in good medical condition two weeks after the surgery. About 1 year after the operation, the patient had no recurrence of pseudotumor and could walk without crutches. This case report is compliant with the SCARE 2020 guidelines and criteria [28].

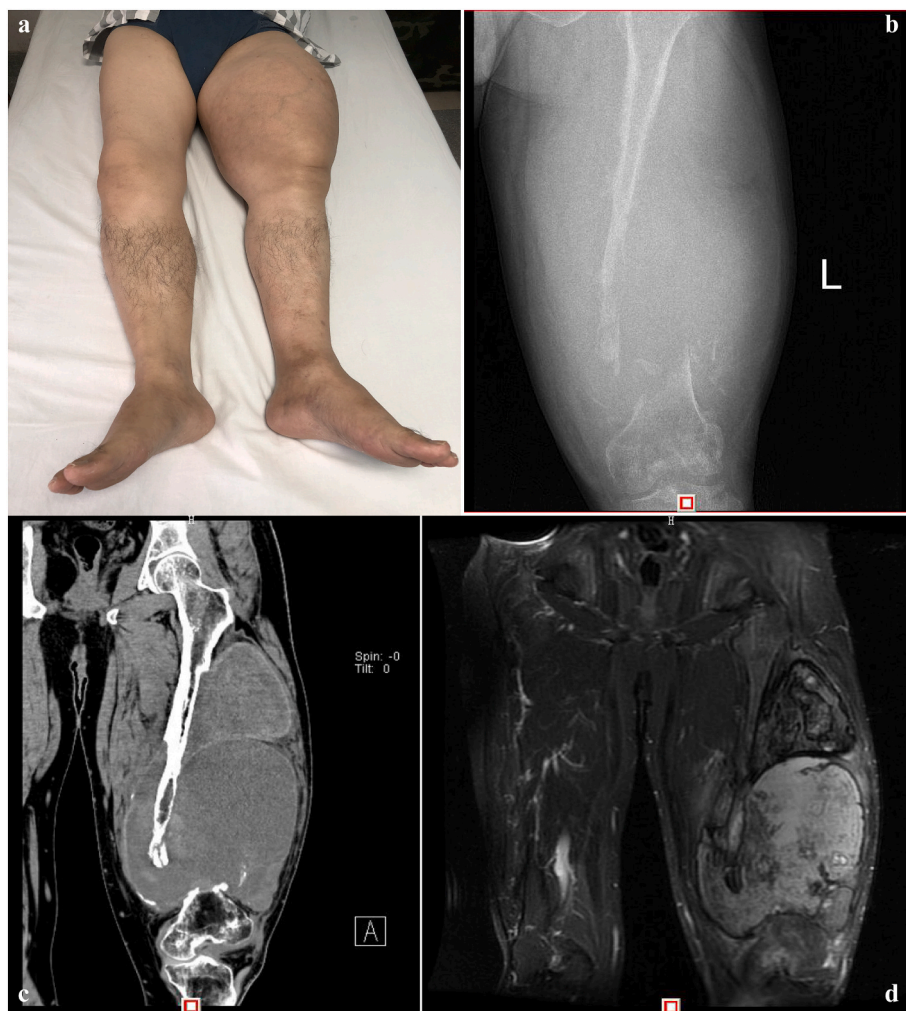


Fig. 1. (a) Significant swelling of the left thigh. (b) X-rays showed pathological fracture of the left thigh. (c) CT scan of left thigh lesion and fracture. (d) MRI scan of soft tissue mass in left thigh.

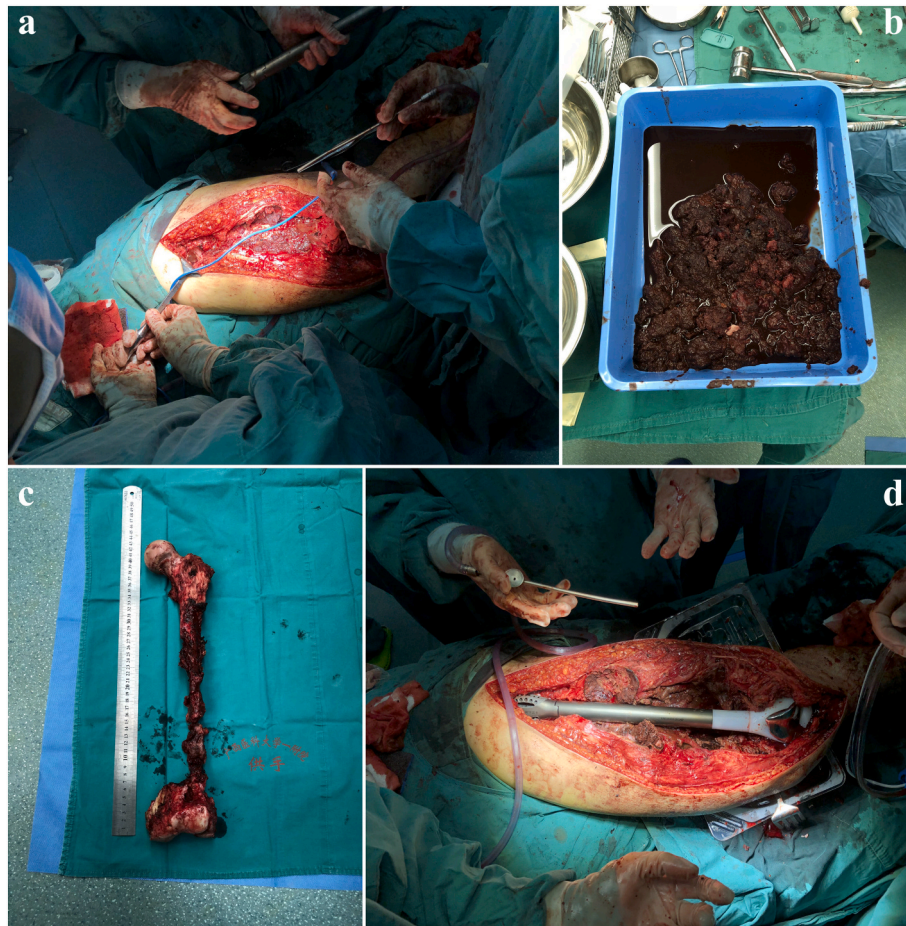


Fig. 2. (a) Intraoperative visualization of a cystic mass in the left thigh with a large amount of blood clot and necrotic tissue. (b) blood clot and necrotic tissue. (c) Left femur destroyed by pseudotumor. (d) Installation of total femoral prosthesis.

3. Discussion

Hemophilia is a coagulation disorder caused by X-linked recessive mutations in the coagulation factor VIII (A) or IX (B) genes [16]. In 1918, Starker first described the hemophilic pseudotumor as a slow progressive subperiosteal hemorrhage [17]. Hemophilia patients are prone to develop hemophilic pseudotumor after recurrent spontaneous or traumatic bleeding in bone or soft tissue [18]. Previous studies have shown that about 1–2% of hemophilia patients (especially those patients whose coagulation factor level < 1%.) develop to hemophilic pseudotumor [1–3]. In essence, hemophilia pseudotumor is a chronic encapsulated collection of blood caused by recurrent intramuscular, intraosseous and subperiosteal bleeding. Pseudotumors most often occur in or close to the large bones of the proximal skeleton [19], but the small bones of the hands or feet also can be involved, especially in children [1,2,15,19]. The initial hemorrhage may be intraosseous or subperiosteal, and bleeding frequently occurs in the muscle. The clinical manifestations are gradually enlarged painless mass (severe pain if pathological fracture occurs), and the mass is usually firm non-tender, and adherent to the deep structures [19]. Hemophilic pseudotumors can remain asymptomatic for decades, and then a sudden rupture of the pseudotumor can lead to uncontrollable bleeding [1], or severe pain when a pathological fracture occurs [19]. About 50% of the patients with pseudotumors and fractures had a history of previous trauma [2,15,19].

The radiographic findings are characteristic. Typically, a large soft tissue mass can be seen, accompanied by the adjacent area bone destruction, and calcification and ossification can often be seen in the

mass [19]. When the initial hemorrhage occurs in the muscle, secondary changes in the adjacent periosteum and bone occur, leading to periosteal elevation with new bone formation at the edges of the lesion [19,20]. Destruction of medullary bone may produce smaller subperiosteal cysts, but occasionally larger expansive lytic lesions [20]. In terms of diagnosis, computed tomography is useful in showing the size and location of the pseudotumor, the bone destruction, the adjacent relationship with neurovascular structures, and the presence of fibrous capsule [20–22]. Magnetic resonance imaging is very useful in the evaluation of hemophilia pseudotumor, not only in defining the anatomical features, but also in monitoring treatment response. A hypo-intense capsule around the hematoma often can be seen and it is observed better on T2-weighted images than on T1-weighted images [23]. The variations of signal intensity can be seen in the hematoma, which reflects the development of pseudotumor is characterized by the existence of different degradation stages blood products secondary to the chronic recurrent hemorrhage [23]. The signal intensity of fresh hemorrhage in the pseudotumor was similar to that of skeletal muscle on T1-weighted images, and markedly hypointense on T2-weighted images. On the contrary, due to the presence of methemoglobin, the part of chronic hemorrhage is hyperintense on T1-weighted images and has multiple manifestations on T2-weighted images [23]. Blood products at different stages can distinguish hemophilic pseudotumor from solitary bone cysts (containing homogeneous fluid), giant cell tumors and aneurysmal bone cysts (space filled with blood) [20]. On histologic evaluation, a hemophilic pseudotumor is characterized by a multicystic hematoma in various states of organization surrounded by a fibrous capsule. Calcification, ossification, and hemosiderin deposition can be seen in the hematoma; fibroblasts,

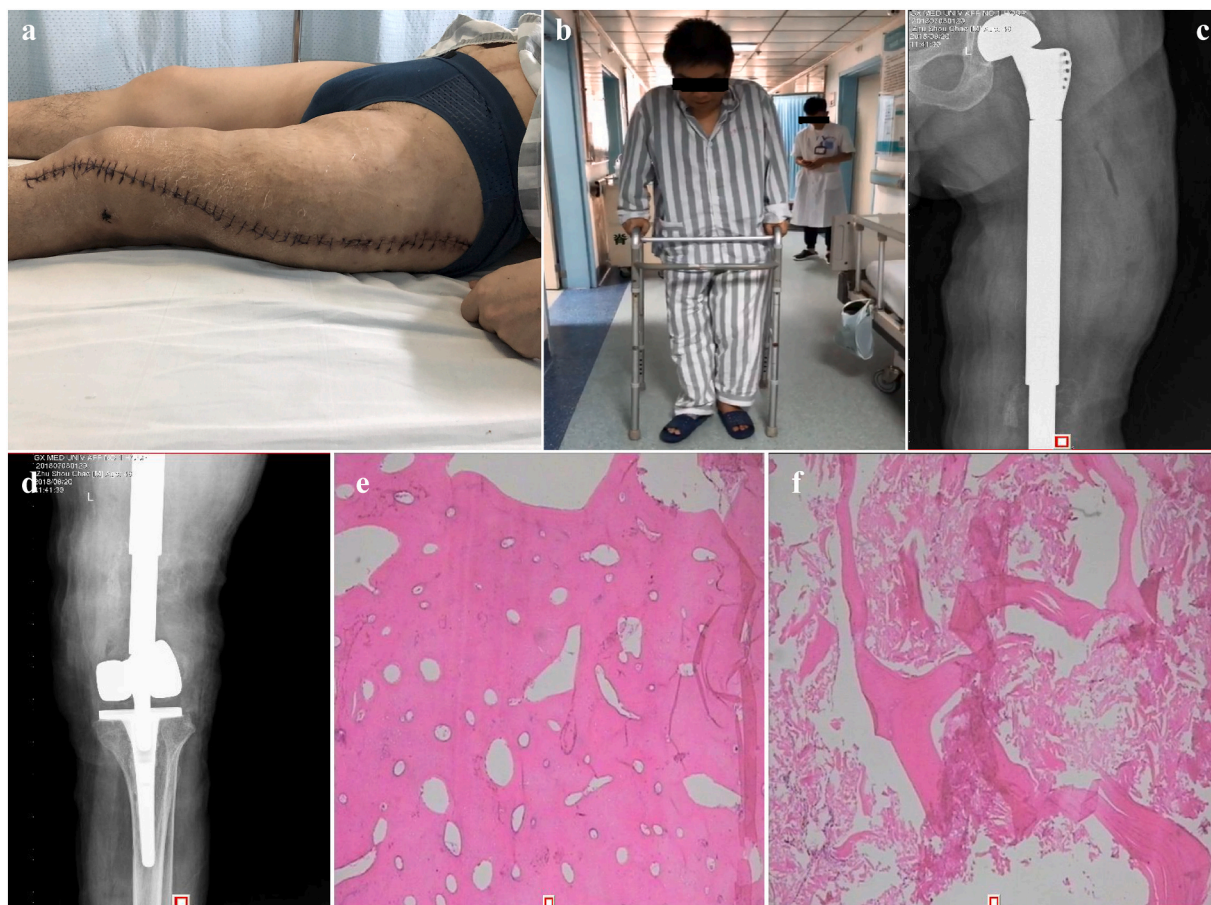


Fig. 3. (a) Status of incision healing 14 days after surgery. (b) Walking with abduction brace 14 days after surgery. (c) and (d) Post-operative radiographs show a well-positioned prosthesis with no displacement or fracture. (e) and (f) The results of pathological examination showed fibroblasts, histiocytes, and hemosiderin pigment consistent with hemophilic pseudotumor.

hemosiderin-loaded histiocytes, giant multinucleated cells, evidence of neovascularization, and hemosiderin stores also can be observed in the capsule [19,24].

Various treatments for hemophilic pseudotumor have been proposed; however, because of its rarity, there are no standard guidelines for treatment so far. Factor VIII replacement therapy [12,13], local irradiation [12,14], and surgery [10,11] have been proposed in previous studies. Before the advent of factor replacement therapy, the treatment of hemophilic pseudotumors was largely nonoperative; However, for pathological fractures caused by pseudotumor, non-operative treatment cannot stop the progression of these lesions [1,19]. With the advent of factor concentrates, chronic factor replacement and immobilization have been applied and more positive results have been achieved, especially in children with distal lesions (distal ankle or wrist), factor replacement and immobilization can reduce immobilization and induce bone remodeling [1,19]. Our patients received coagulation factor replacement therapy, immobilization and skin traction before operation, but these treatments did not achieve a cure.

Radiotherapy has achieved varying degrees of success in the treatment of hemophilia pseudotumor. Because local irradiation destroys the nutrient vessels of the pseudotumor, it is thought to interrupt the circulation of chronic recurrent bleeding that promotes the growth of these lesions [12,14,24–26]; However, no large-scale trials have been done. Several authors have questioned the efficacy of radiation in the treatment of hemophilic pseudotumor. Although radiotherapy has achieved some success in the treatment of hemophilic pseudotumor, several authors have questioned the efficacy of radiotherapy. Jensen and Putman [15] report that their own experience shows that radiotherapy is not

effective in treatment of pseudotumors with long-term bone changes, so they conclude that the effect of radiotherapy in treatment of pseudotumors was difficult to assess. Gilbert [19] stated that because of the high recurrence rate, he does not recommend radiotherapy except for patients for whom surgical intervention was not a possibility and replacement therapy is ineffective.

Surgical intervention is more difficult and risky than non-operative treatment, but it seems to have better outcomes. There are many patient reports of successful surgical intervention [10,11,27]. In a multi-center study, Magallon et al [24] stated that surgery was more successful than nonoperative treatment and 8 of 14 patients who received surgical treatment were cured (compared with 2 out of 15 patients were successful cured through nonoperative measures). Gilbert [19] stated that surgical resection for hemophilic pseudotumor is the first choice, but he also stated that although it is a very effective treatment, it will be safer if done in a major hemophilia centers. Jensen and Putman [15] concluded that surgery seems to be the first choice when pseudotumor has been exist for many years and nonoperative treatment is ineffective. However, pseudotumor puncture should not be performed before surgery, as it usually leads to bleeding, pseudotumor recurrence, fistula formation, infection or sepsis, or both [15,19,24].

As far as we known, resection of the whole femur and prosthesis reconstruction as an alternative treatment to amputation have not been reported in patients with a massive pseudotumor of the extremities. Although there were no postoperative complications in our patients, extensive resection still had many potential complications including infection, uncontrollable bleeding, delayed wound healing, and systemic problems. Therefore, patients should be consulted about surgical

risks before the surgery.

4. Conclusion

In this paper, we reported the case of a 48-year-old man with a pathologic fracture of the left femoral shaft due to a massive hemophilic pseudotumor treated with radical resection and total femoral prosthesis reconstruction. Although there have been many reports of hemophilic pseudotumor now, the case we reported is the largest pseudotumor ever reported. More importantly, this is the first report of hemophilic pseudotumor treated with radical resection and reconstruction total femoral prosthesis. The radical resection and prosthesis reconstruction could be a new method for the treatment of these destructive and challenging soft tissue masses in the extremities.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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.

Ethical approval

This report does not contain any personal information that could lead to the identification of the patient. Therefore, it is exempt from ethical approval.

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Guarantor

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Registration of research studies

Not applicable.

CRedit authorship contribution statement

All of the authors contributed to the case study, research, and writing of the manuscript.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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