Pelvi-Femoral Complete Bone Bridge in a Patient with Hemophilia

Wei Zhu[1], Lin-Jie Zhang[2], Chao Jiang[1], Xi-Sheng Weng[1]

[1]Department of Orthopedics, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100730, China

[2]School of Clinical Medicine, Peking Union Medical College, Beijing 100730, China

Wei Zhu and Lin-Jie Zhang contributed equally to this study.

To the Editor: Heterotopic ossification (HO) is a formation of bone outside of the skeletal system. It usually occurs in the following five broad clinical settings: genetic, posttraumatic, neurogenic, postsurgical and as distinctive reactive lesions most often seen in the hands and feet.[1] Although there is an increased tendency to bleed in soft tissue, HO has rarely been reported in hemophilia. At present, a few cases reported the location relationship between ectopic osteogenesis and the pelvis, and only two cases presented pelvi-femoral bone bridge-like progression of this phenomenon.[2,3] More precisely, it was situated in the posterior area of the hip joint. Here, we describe a rare case of HO bridging from the anterior superior spine to the lesser trochanter of the femur in hemophilia.

The orthopedic clinic was presented with an 18-year-old male with left hip pain, flexion deformity, and restricted walking. These symptoms had developed during the previous 4 years after falling off of a horse. He was diagnosed hemophilia A when he was 2 years old and almost never treated by factor VIII (FVIII).

At the physical examination, there was no obvious swelling in the left hip. The left hip could only buckle 60°–70° with limited adduction, internal rotation, and abduction. Further, it could touch about an anterior 10 cm × 1 cm long rigid fixed mass that had gradually increased in the 4 years following the trauma. He had no neurological impairment.

For the radiological examination, an X-ray revealed a clearly distinguished heterotopic bone bridging from the anterior superior spine to the lesser trochanter of the femur [Figure 1a]. A three-dimensional reconstruction of the computed tomography scan was carried out to ascertain its location, which was just like a rib restricting hip joint function [Figure 1d]. As demonstrated in the figure, there are no clear gaps between the two ends of the heterotopic bone with that of the anterior superior iliac spine and femur.

Case characteristics were as follows: (1) main problems of this patient were abnormal gait and limited activities; (2) the atopic bone was most likely derived from posttraumatic ossifying myositis rather than fibrodysplasia ossificans progressiva; and (3) the operation was necessary to solve this condition.

Access this article online

Quick Response Code:

Website: www.cmj.org

DOI: 10.4103/0366-6999.244110

In the treatment, we consulted the hematology department and conducted a pharmacokinetic experiment of coagulation factor (preexperiment) infusion of FVIII 40–50 U/kg. Blood was taken before infusion, as well as 0.5 h, 3 h, 6 h, 8 h, 12 h, and 24 h after the infusion to detect FVIII and activated partial thromboplastin time. In the meantime, FVIII inhibitor concentrations were measured before and 24 h after infusion. Perioperative FVIII replacement therapy was conducted based on the guidelines of the World Federation of Hemophilia.[4] The specific quantity of infusion was 2600 U q8h in the surgery day to the next day, 1800 U q12h between day 2 and day 3, and 800 U qd from day 4 to day 7 after operation. The hip mass [Figure 1c] was resected from the patient. Besides loxoprofen sodium tablets for 2 weeks, no additional postoperative medicine was prescribed to prevent the re-occurrence. Eventually, the flexion of his left hip was significantly improved and recovered to a normal walking position [Figure 1b]. After 7 months of follow-up, his hip joint activity returned to normal, with no difference from the healthy one. Even now, he has no re-occurrence.

HO is described as a bone formation in soft tissues. This pathologic process – demonstrated in different diseases and circumstances – may occur in sites such as the skin, subcutaneous tissue, skeletal muscle, and fibrous tissue adjacent to joints. Nevertheless, the location of ectopic bone in a hemophiliac as seen in our patient is uncommon and rarely reported.

It is a known fact that intramuscular hematoma is the second most common bleeding type after intra-articular bleeding in hemophilia. However, the most common locations are the muscles of the thigh rather than where our patient presented. At present, peripelvic new bone formation has been previously described in some rare case reports, but their natural history and specific treatment options were

Address for correspondence: Dr. Xi-Sheng Weng, Department of Orthopaedics, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100730, China
E-Mail: xshweng@medmail.com.cn

Received: 12-07-2018 **Edited by:** Yuan-Yuan Ji

How to cite this article: Zhu W, Zhang LJ, Jiang C, Weng XS. Pelvi-Femoral Complete Bone Bridge in a Patient with Hemophilia. Chin Med J 2018;131:2618-9.

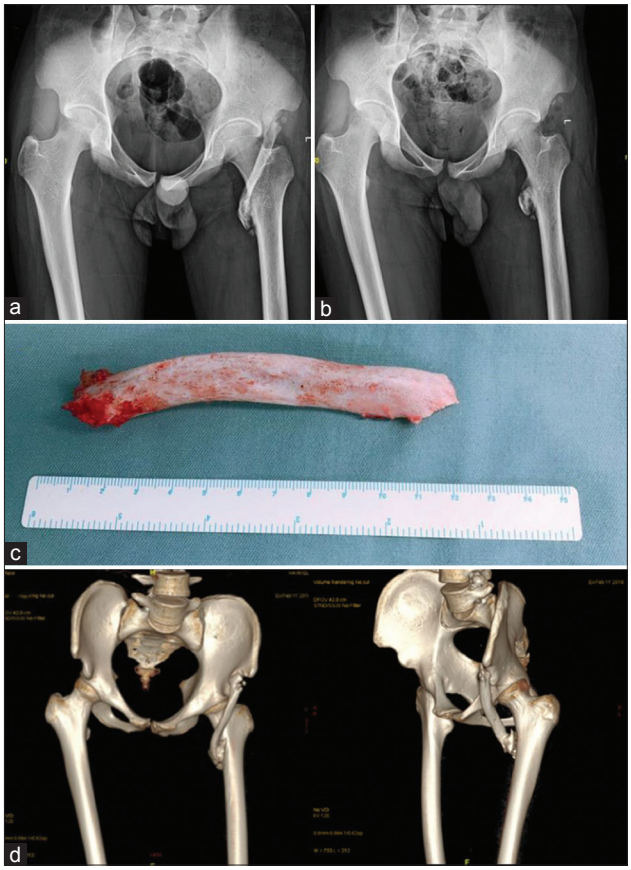

Figure 1: X-ray findings before (a) and after (b) operation; (c) macroscopic findings of the resected heterotopic bone; (d) three-dimensional reconstruction of computed tomography scans in the anterior view (left) and left anterior oblique view (right).

not adequately described. Aydogdu *et al*.[2] reported a case with HO near the lesser trochanter extending to the pelvic rim. They decided to take conservative treatment since their patient did not exhibit intractable pain or neurological involvement. Kalenderer *et al*.[3] presented a case with HO in the quadratus femoris muscle reclining from the femur to the pelvis. Similar to the previous one, the bridge was incomplete. This was owing to the fact that the maturation of the HO was completed reaching to its endpoint. Their treatment consisted of postural exercises and clinical follow-up for the patient. As for our patient, the location of HO had produced definite symptoms for this young boy, significantly affecting daily activities. Although there was no apparent neurological involvement, the surgical indication was specific. The patient was diagnosed with hemophilia at the age of 2 years and did not receive adequate treatment, undoubtedly increasing the difficulty and risk of surgery. Adequate FVIII was infused in the perioperative period along with rehabilitation exercise. After that, his left hip recovered to normal walking position.

Compared with the two former cases: (1) their pelvi-femoral bridges were completed and the heterotopic new bone formation, which connected the trochanter minor and the ischio-pubic region, was located in the posterior area of the hip joint. However, the new bone of this boy connected the trochanter minor and the superior spine, which was located at the anterior of the hip joint, and the bridge was completed. (2) The first two patients had no surgical indications, so only clinical observation was taken. On the other hand, this patient had clear surgical indications, and the prescribed treatment was detailed in this article.

According to the previous reports and experience, conservative management should be the first choice for the patients in hemophilia who have persistent pain and restriction of range of motion in any joint without neurological involvement. Otherwise, surgical intervention must be performed, especially when patients have strong demand.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. Although anonymity cannot be guaranteed, the patient understands that his name and initials will not be published and that due efforts will be made to conceal his identity.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. McCarthy EF, Sundaram M. Heterotopic ossification: A review. Skeletal Radiol 2005;34:609-19. doi: 10.1007/s00256-005-0958-z.
2. Aydogdu S, Memis A, Kavakli K, Balkan C. The pelvi-femoral incomplete bone bridge in a patient with mild haemophilia. Haemophilia 2001;7:224-6. doi: 10.1046/j.1365-2516.2001.00496.x.
3. Kalenderer O, Bozoglan M, Agus H. Heterotopic ossification in quadratus femoris muscle in a haemophilic patient. Haemophilia 2012;18:e13-4. doi: 10.1111/j.1365-2516.2011.02637.x.
4. Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, Llinas A, *et al*. Guidelines for the management of hemophilia. Haemophilia 2012;19:e1-47. doi: 10.1111/j.1365-2516.2012.02909.x.