

ARTICULAR BLEEDING (HEMARTHROSIS) IN HEMOPHILIA

AN ORTHOPAEDIST'S POINT OF VIEW

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INTRODUCTION

The most typical manifestation of hemophilia is articular bleeding (hemarthrosis). When hemarthroses become frequent and/or intense, the synovium may not be able to reabsorb the blood. To compensate for such reabsorptive deficiency the synovium will hypertrophy, resulting in what is called chronic hemophilic synovitis (1)(2)(3). Thus, it is very important not only to avoid acute hemarthrosis, but also to manage it as efficiently as possible, with the aim of avoiding the development of synovitis.

Hematological prophylactic treatment from the age of two to the end of skeletal maturity is the best way to avoid articular bleeds, or at least to diminish their intensity. However, one should remember that problems may be caused by the permanent intravenous infusion needed in such circumstances. Most hemophilia centres in developed countries have on-demand treatment, which consists of the administration of the deficient coagulation factor when hemarthrosis occurs.

TYPES OF HEMARTHROSES

It is important to differentiate acute bleeding and subacute bleeding. Subacute hemarthrosis is generally associated with previous synovitis or arthropathy, while acute hemarthrosis commonly occurs in a previously healthy joint (4). Acute bleeding is usually felt by the patient as a burning sensation in the joint. Hemarthrosis develops within a few hours; the joint becomes inflamed, tense, warm, and painful, and the skin becomes bright red. The affected joint is held in an antalgic flexion position, with painful and limited mobility. After administration of the appropriate doses of factor concentrates, pain will rapidly diminish, although inflammation and limitation of

articular mobility commonly disappear more slowly. The degree of inflammation and limitation of motion are always related to the amount of blood in the joint.

Subacute hemarthroses commonly occur after two or three articular bleeding episodes and persist despite adequate hematological treatment. Pain can be tolerable and is commonly associated with hypertrophic synovium on palpation and a slight lack of joint mobility. When subacute hemarthroses recur for months and years, they will result in a state of hemophilic arthropathy. This usually occurs in young adults, who complain of persistent pain in the affected joint, not only with movement but also at rest. They may also have intermittent episodes of acute pain and inflammation related to synovitis or articular bleeding.

TREATMENT OF HEMARTHROSES

Acute Hemarthroses

Optimal treatment of acute hemarthroses involves a combination of factor replacement, joint aspiration, rest (with or without splinting), ice, appropriate analgesia, and supervised rehabilitation once the acute phase has been controlled and the risk of bleeding reduced (5). The objectives of treatment are to avoid muscular atrophy, maintain an adequate degree of articular mobility, control the recurrence of hemarthroses, and recover joint function if possible.

Joint Aspiration

Joint aspiration is not commonly performed, but in cases of severe bleeding it may relieve the patient's pain and speed up rehabilitation. There is a great deal of controversy on the role of arthrocentesis in hemophilia. The author's view is that minor bleeding episodes can be treated by other means. However, major bleeds may benefit from joint aspiration, providing it is done within the first 12 hours. Before deciding on joint

aspiration, the presence of a circulating inhibitor must be investigated to determine the best treatment. In any case, three to four days of joint rest are recommended. When hemarthrosis does not respond to hematological treatment, septic arthritis must be suspected, especially if the patient is immunodepressed; joint aspiration and culture will allow us to reach a diagnosis (6).

If hemarthrosis does not respond to hematological treatment, one must suspect hemophilic synovitis, which can be detected by clinical examination. Ultrasonography and Magnetic Resonance Imaging will help confirm the occurrence of synovitis. In such cases only aggressive treatment of synovitis will allow us to control articular bleeding, which is secondary to synovial hypertrophy. Synovitis can be controlled with early prophylactic treatment or by synovectomy (radionuclide synovectomy or surgical synovectomy). Diagnostic imaging is paramount to assess the response to any type of treatment. In 1998, Heim et al. (7) reported an interesting case of a person with hemophilia who had a fixed flexed hip and intractable pain. This clinical picture was suggestive of hemorrhage in that area. Ultrasonography confirmed the diagnosis of acute hip hemarthrosis. Narcotic drugs failed to alleviate the severe pain. Joint aspiration produced dramatic pain relief and early joint rehabilitation. However, Heim et al did not suggest that every coxhemarthrosis should be aspirated. It should be remembered that raised intra-articular pressure may contribute to femoral head necrosis in adults, or to Perthes' disease in children. It is important to emphasize that while arthrocentesis of the elbow, knee, and ankle are quite simple procedures that can be performed at the outpatient clinic, both shoulder and hip joint aspirations require sedation and radiographic control by an image intensifier; that is to say, they are surgical procedures done in an operating room, with an anesthetic and by an orthopaedic surgeon.

Rest and Splinting

Rest for lower limb bleeding episodes should include bed rest (one day) followed by avoidance of weight-bearing and the use of crutches when ambulating, and elevation when sitting (three to four days). For the knee a compressive bandage is adequate, although in very painful cases the bandage should be supplemented with a long-leg posterior plaster splint. For the ankle, a short-leg posterior plaster splint is recommended. For the upper limb,

usually a sling (for the shoulder) or a long-arm posterior plaster splint (for the elbow) will provide sufficient rest, support, and protection. Lifting and carrying heavy items should be avoided until the bleeding has resolved (four to five days).

Ice

Ice therapy relieves pain and reduces the extent of bleeding by promoting vasoconstriction. Ice therapy can be applied to the affected joint in various ways: cold packs, wet towels, crushed ice and Cryocuff. Applications of ice over a 24- to 48-hour period can help control volume of blood and pain. Ice should not be applied directly to the skin, but be wrapped in a thick towel, because it burns and prolonged application can cause skin damage. The effectiveness of ice as a treatment for acute bleeds lies not only in its physical effects but also in its ease and simplicity of application.

Analgesia

Depending on the degree of pain, paracetamol or a combination of paracetamol and dextropropoxyphene should be administered. Usually they provide adequate relief. Aspirin-containing products and nonsteroidal anti-inflammatory drugs must be avoided.

Subacute Hemarthroses

It is advisable to treat subacute hemarthroses with hematological substitutive therapy, with two to three weeks of immobilisation by means of a semi-flexible splint. Some studies recommend six to eight weeks of prophylaxis with physiotherapy. It is recommended to administer enough of the deficient factor, three times a week, to obtain 20% to 30% of the normal level. After each transfusion the patient should complete an exercise program focusing on active joint mobility, under the surveillance of an expert physiotherapist. If such mobility exercises are painful, only isometric exercises should be done.

When a flexion contracture does appear, it should be treated early and aggressively by conservative means to avoid its becoming irreversible. Conservative measures include Oxford's inverted dynamic splints, extension-desubluxation hinged casts, dynamic splints, and traction followed by a polypropylene orthosis. Oxford's technique was specially designed for the knee joint and requires admitting the patient to hospital. The lower limb is put in balanced traction on a semicircular Thomas splint which has a knee-flexion Pearson's device. Then soft traction is

put on the calf with the heel free; a posterior force is applied on the thigh by means of a cushioned spring located on the distal part of the thigh, which is connected with a string to a 3 kg weight. Such a posterior force counteracts the anterior force produced by the springs located on the posterior part of the calf. Both the longitudinal traction and the thigh weight are progressively increased. When the knee becomes fully extended, or if the technique does not work after one week of treatment, the patient is mobilised with a Böhler cast which is open in its anterior part. According to the Oxford authors, hematological therapy is not required (8).

The hinged extension-desubluxation cast can be made of plaster of Paris or of a thermoplastic material; it should be open in its anterior part. The hinge is adjusted once or twice a day to correct the deformity. When the contracture is less than 20°, the cast can be removed and replaced with a plaster splint. Hematological substitutive therapy is necessary during the procedure. The dynamic splint is adjustable and allows a low intensity but long duration force through the knee joint. A gain of 5° to 10° of knee extension can be expected in six to nine months with this procedure. However, many patients may have hemarthrosis during the follow-up. Traction followed by orthosis is another alternative.

Flexion contracture has a different treatment and prognosis, depending on its chronicity and other associated deformities (9). A flexion contracture of a few days' duration can be corrected by means of traction followed by rehabilitation and orthosis. A flexion contracture with a duration ranging from weeks to months may require surgery: hamstring release and/or supracondylar extension osteotomy. A flexion contracture associated with osseous or fibrous ankylosis may also require a patello-femoral osteotomy.

CONCLUSION

Prompt treatment with factor replacement and rest of the affected limb should allow rapid resolution of the bleeding episode with minimal risk of long-term problems. The affected joint should remain at rest for a short period of time (four to five days): bed rest for the hip, a sling for the shoulder, and a compressive bandage and plaster splint for the elbow, knee, and ankle. Ice therapy helps to relieve pain and reduce the extent of bleeding. Analgesics

(paracetamol) may also be required, depending on the degree of pain. An early and progressive physical therapy is then required to recover the full range of movement and the strength of periarticular muscles. While some authors recommend joint aspiration to remove the blood as an important therapeutic measure, others do not routinely perform this procedure in hemophilic patients. Currently, arthrocentesis is one of the most controversial issues regarding treatment of hemarthrosis in hemophilia. My view is that arthrocentesis should always be performed in major hemarthrosis (voluminous, very tense and painful joint). Minor hemarthrosis commonly responds to hematological treatment and rest. Arthrocentesis of the hip and shoulder should be done under radiographic control in an operating room by an orthopaedic surgeon. Aspiration of the elbow, knee, and ankle are quite simple procedures that can be done in the outpatient clinic, not necessarily by an orthopaedic surgeon. Joint aspiration should always be done under factor coverage and in aseptic conditions, to avoid recurrence of bleeding or septic arthritis.

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