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Case report

Hemarthrosis subtalar, a rare diagnosis☆

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ABSTRACT

Type B hemophilia usually affects patients with a family history of this disease and has a typical clinical picture. However, in the present case it appeared in a patient outside the typical age with no family history of hematologic malignancies and with an unusual clinical picture.

Hemartrose subtalar, um diagnóstico raro

Palavras-chave:
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RESUMO

A hemofilia do tipo B afeta normalmente pacientes com história familiar positiva para a doença e se apresenta com quadro clínico típico. No presente caso, no entanto, o diagnóstico se deu em um paciente fora da idade típica, sem histórico familiar de doenças hematológicas e quadro clínico diferente do habitual.

Introduction

Hemophilias are hematologic diseases that results in changes in the coagulation process, due to deficiency of a clotting factor. The disease has a genetic cause and the most common hemophilias are type A (factor VIII deficiency) and type B (factor IX deficiency).

Type B (or Christmas disease) accounts for 14% of cases, equivalent to one case per 30,000 births of male children.[1,2] The disease is highly associated with males due to the fact

☆ Study conducted at the Santa Casa de Misericórdia de São Paulo, Departamento de Ortopedia e Traumatologia, Grupo de Ortopedia e Traumatologia Pediátrica, São Paulo, SP, Brazil.

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that the altered gene is located in the long arm of the X chromosome. The most common signs are bleeding, often in the mucous membranes, joints, and subcutaneous tissue. Symptoms include hemarthrosis, bruisings, hematuria, and gastrointestinal bleeding.[3] The diagnosis is made by tests that assess the coagulation cascade: prothrombin time (PT), international normalized ratio (INR), and activated partial thromboplastin time (aPTT). If the intrinsic pathway of coagulation is affected, the PT will be normal, but the aPTT will be higher. To confirm factor IXdeficiency, the factor itself should be measured.[3,4]

Hemarthrosis is a bleeding in the joints and can be of traumatic, hematological or neurological origin, among others.[5] The most common cause is trauma to articular regions. In cases where it courses with neurological problems, they are usually associated with the Charcot joint, an alteration in which there is no articular proprioceptive (neurological) perception. The hematological etiology may be induced by drugs or acquired hematological diseases (myeloproliferative diseases and thrombocytopenia, among others), or be hereditary (hemophilia). The most common bleeding sites are the knee, elbow, and tibio-talar joints.[6,7] The initial symptoms are redness and swelling, sometimes associated with paresthesia, and the process may progress to severe pain.[8] If the hemarthrosis is not treated, the inflammatory process will become chronic and may lead to functional impairment. The diagnosis can be made using imaging tests such as ultrasonography, CT scan, or magnetic resonance imaging (MRI); however, the gold standard is the aspiration of the joint fluid.[7,9,10]

Hemarthroses in patients with hemophilia B are usually indicators of moderate or severe factor IX deficiency. When the ankles are affected, it is difficult to exclusively use the symptoms to distinguish which specific joint was affected. According to Rodriguez-Merchan[11] and Lofqvist et al.,[12] the subtalar joint may be associated in approximately 50% of the cases. However, it is rarely involved and reported in isolation. The clinical picture in these patients may be unclear, since many do not experience severe pain or significant functional gait limitation.

This study aimed to describe the difficulty in the diagnosis of hemophilia B in an extremely rare event of isolated hemarthrosis of the subtalar joint.

Case report

Male patient, aged 1 year and 8 months, born and raised in São Paulo, Brazil, white, and of no declared religion. His mother reported he had been having difficulty to walk for eight days, due to problems in weight bearing and dorsiflexion of the left foot. The mother also reported an abrupt and progressive swelling in the lateral and posterior region of his ankle. He remained afebrile at all times and was prescribed Predsim® (sodium prednisolone phosphate) and Hixizine® (hydroxyzine hydrochloride) for suspected insect bite (Fig. 1).

The mother denied changes in other systems, routine use of medications, and history of chronic diseases in the family. The vaccine immunizations pertinent to his age range were up-to-date. The child had a history of bronchiolitis at 5 months; coxsackievirus infection at 8 months, and an episode of transient hip synovitis at age 1 year and 2 months, which resolved itself in a short period of time.

At the general physical examination, there were no cardiovascular, pulmonary, abdominal, or neurological changes.

The orthopedic physical examination showed increased volume throughout the ankle, but without local temperature increase or redness. The gait was altered, with left side limping and impaired weight bearing.

The results of the initial tests were as follows: C-reactive protein (CRP), 8.22 mg/L; capillary glucose, 87 mg/L; and erythrocyte sedimentation rate (ESR), 15 mm. The complete blood

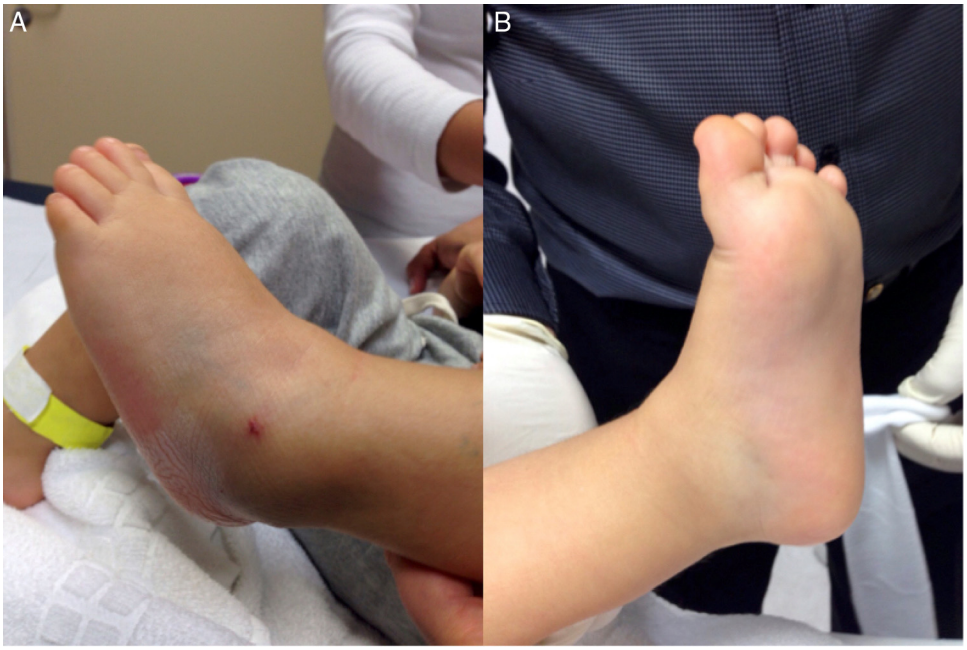

Fig. 1 – (A) Lateral view of the patient's foot at admission; (B) medial view of the foot at admission.