Clinical case

Synovial Chondromatosis of the Hip in a Child with Down Syndrome*

Gómez Palacio V.E[1], Gil Albarova J.[2], Bregante Baquero J.[2].
(1) Servicio de Cirugía Ortopédica y Traumatológica, Hospital San Pedro de Logroño. C/ Piqueras nº 98, 26006, Logroño. Spain
(2) Sección de Cirugía Ortopédica y Traumatología Infantil, Hospital Universitario Miguel Servet de Zaragoza. Paseo de Isabel La Católica 1-3, 50009, Zaragoza. Spain

Correspondence:
Victoria Gómez Palacio
e-mail: vgpalacio@riojasalud.es

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Abstract

Synovial chondromatosis of the hip is a rare benign condition characterized by multiple intraarticular loose bodies. We report the case of a fourteen-year-old child with Down syndrome who suffered symptomatic synovial chondromatosis of the hip, treated by means of arthrotomy and intraoperative radiological assessment to confirm the removal of all loose bodies.

The literature about treatment options is reviewed, and controversial points of this condition are discussed.

Keywords. Synovial chondromatosis. Hip. Children. Down syndrome.

Introduction

Synovial chondromatosis (SC), also known as synovial osteochondromatosis or chondrometaplasia, is a generally benign and rare condition that gives rise to multiple cartilaginous nodules inside connective tissue, tendon sheaths, synovial membranes and joint bursae (1-4). It predominantly occurs in males (2:1) (1, 3-5) in the third and fifth decade of life, and rarely occurs in children.

This condition of unknown etiology is predominantly monoarticular, but in 10% of cases affects multiple joints, particularly large joints such as the knee, shoulder and hip, but also, to a lesser extent, smaller joints. Clinical manifestations range from asymptomatic forms to the full-blown clinical picture with mechanical pain, impaired mobility, swelling, blockage, and joint effusion (4, 5, 7).

Primary or idiopathic SC should not be confused with secondary SC, which develops where there is prior impairment or trauma.

Pathophysiologically, this condition involves the formation of sessile or pediculated cartilaginous nodules which may detach and become intraarticular loose bodies of varying size that may subsequently grow in size. They are not always calcified, and in 30% of cases are not radiologically visible (5). They can be found using arthrography, computed tomography (CT), or magnetic resonance imaging (MRI) (7).

Treatment consists of the removal of the loose bodies using open surgery or arthroscopy in association with synovectomy, particularly in repeat cases (4, 8). True recurrence after incomplete removal and repeat recurrence rates are low, and malignant transformation to chondrosarcoma is exceptional (5%) (5).

Clinical findings

This is the case of a 14-year-old boy with Down syndrome, atrioventricular patency, progressive mitral insufficiency, hypothyroidism, high blood pressure, and bilateral hip dysplasia associated with previous episodes of voluntary hip dislocation. His presenting complaint was a change in gait pattern and an inability to perform sports activities for two and a half months prior to the first visit.

Physical inspection showed comparatively limited

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mobility of the left hip, both for flexion/extension and rotations. X-rays (Figure 1) showed multiple calcifications in the left hip, with a subluxation of the head of the femur that was compatible with a diagnosis of SC in the left hip. MRI (Figure 2) confirmed the suspected diagnosis and enabled accurate determination of where the foreign bodies were located.

After presurgical workup, the left hip was approached anteriorly, multiple bodies were extracted and a total synovectomy was performed with the help of a controlled anterior luxation o f the femoral head (Figure 3); a plicature of the capsule was also performed, as well as acetabuloplasty to center the head of the femur in the hip socket.

Intraoperative scans confirmed extraction of all free bodies prior to closure of the wound with vacuum dressings. Finally, the patient was immobilized with a single hip spica cast.

Samples were sent to the Pathology Service, which confirmed the diagnosis.

Five weeks after surgery, the cast was removed and the patient was allowed to walk on crutches and gradually increase weight bearing in the Rehabilitation Service. Eight weeks after surgery the patient was able to walk on his own with bilateral symmetrical mobility of the hips.

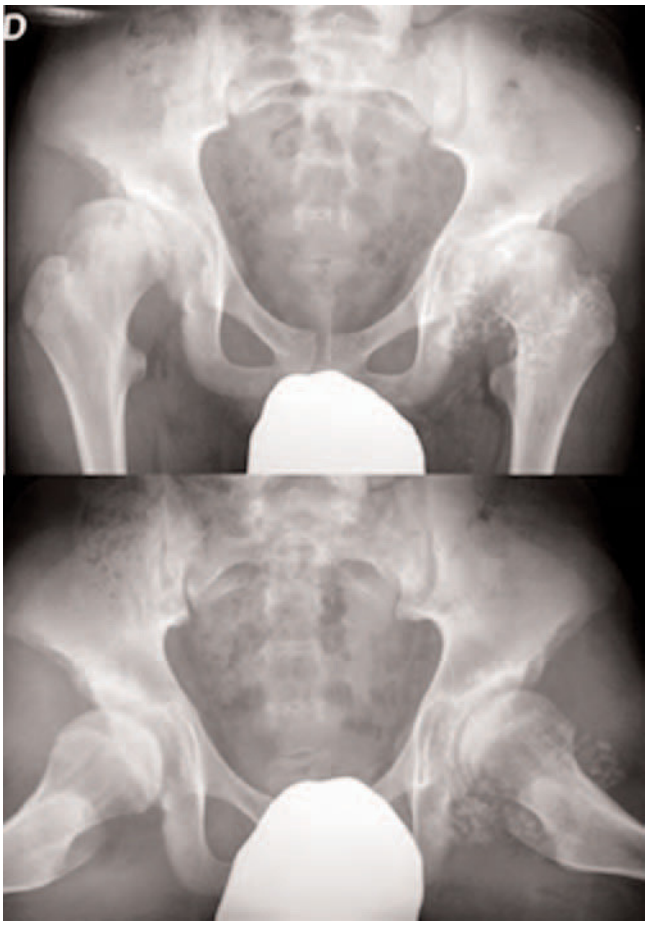

Figure 1: Presurgical x-ray
Note the many loose bodies and the lateral subluxation of the femoral head.

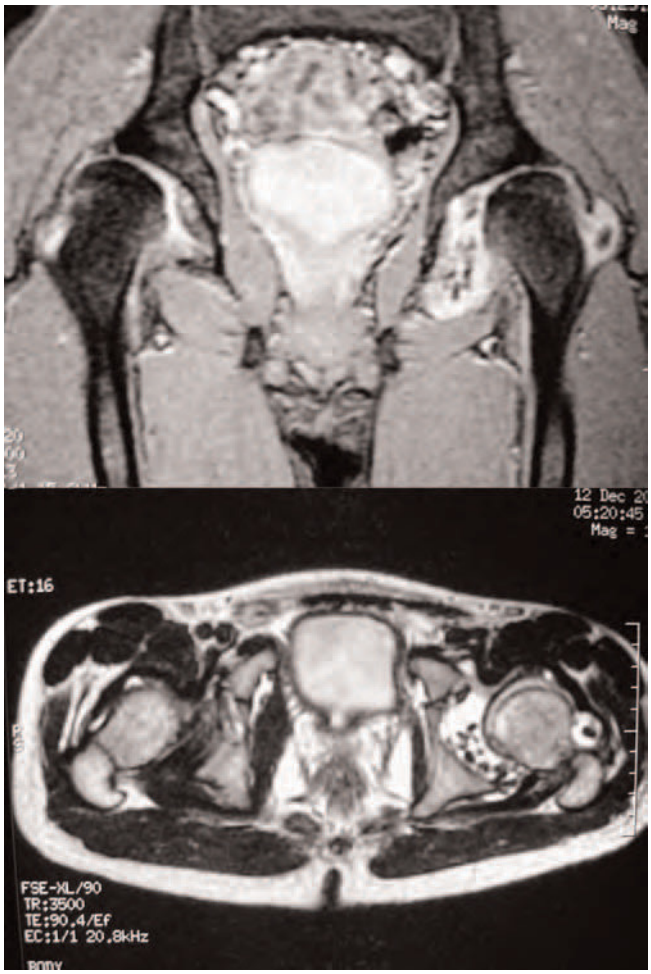

Figure 2: Presurgical MRI
Detailed view of loose body distribution.

At 6 months' followup the patient remains asymptomatic, with bilateral symmetrical mobility of the hips and no radiological signs (Figure 4) of further free bodies. Sports activity has been resumed.

Discussion

The first well-documented description of SC was published by Jones in 1924, although some authors prefer to cite Jaffe, who in 1949 stated that the presence of intrasynovial metaplastic cartilage and of loose bodies were both essential to this diagnosis (4, 7). Milgram (2, 6) considers a diagnosis of SC even if no active synovial membrane is discerned, and defines three phases: Phase I with active intrasynovial disease only, with no loose bodies; Phase II with both active intrasynovial proliferation and free loose bodies; and Phase III with multiple loose bodies but no activity in the synovial membrane. There is unanimous agreement (1, 4) that this condition generally affects a single joint, usually

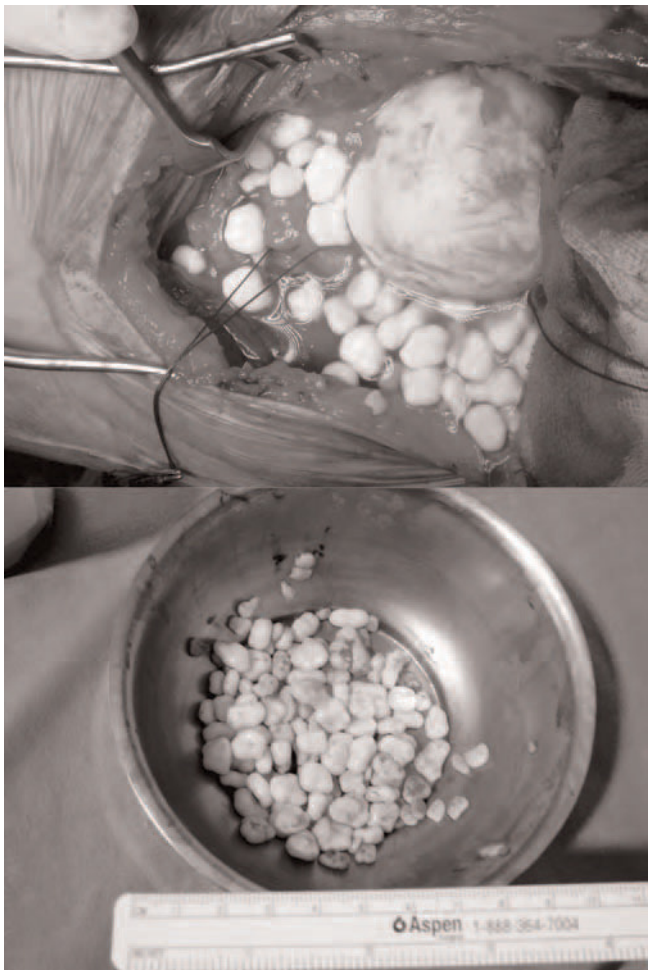

Figure 3: Anterior luxation of the femoral head. Detailed view of extracted loose bodies.

the knee, and that this is an uncommon finding in children (3-7).

Our literature search found no case report of synovial chondromatosis of the hip in a child with Down syndrome.

Because of the patient's age, we feel this case to be a primary form of the condition, although the history of voluntary hip luxation of both hips might allow for the possibility that it is a secondary form. Two cases of secondary SC were found in our literature search: one secondary to progressive ossifying dysplasia, the other secondary to Perthes disease (5).

Treatment, which varies according to the affected joint (4, 5, 8, 9), is arthroscopic surgery for the shoulder and knee, and arthrotomy for the hip joint (4).

Hip luxation increases the risk of avascular necrosis of the head of the femur, epyphysiolysis, and fracture of the femoral head, lesser trochanter or bone shaft, as well as femoral nerve neuroapraxia (8, 9). Some authors advocate controlled luxation to prevent hip fracture, and state that metaphyseal and retinacular vessels may supplement vascularization of the femoral head (3).

Complications of hip arthroscopy include neurovascular damage or damage to the labrum or the joint cartilage, impairment of the femorocutaneous nerve, and breaking of osteophytes (8, 9), so this procedure is not advised for the present indication.

Figure 4: X-ray, 6 months post-op.

More repeat cases have been published for arthroscopic surgery (up to 33%) than for open surgery, although this is a rare condition and it must be borne in mind that the overall numbers are low (7). A distinction must be drawn between true recurrences with incomplete removal of all loose bodies and repeat cases; they are kept distinct in the published series (22-50%) (4). We agree with other authors (4) in recommending intraoperative scanning to verify extraction of all loose bodies and prevent a recurrence. As for repeat cases, their rate is spectacularly reduced if the surgery is performed together with a partial or total synovectomy (4, 8). It has been suggested (2) that if chondromatosis is in phase III or even phase II there is no need for synovectomy. Other authors (10) feel that synovectomy is not curative if the location is intraarticular, but conversely is curative for an extraarticular location.

Malignant conversion is a very infrequent occurrence (5) that mainly involves the knee. This is highly controversial topic in the case of primary synovial chondromatosis, as it is very hard to distinguish between primary synovial chondromatosis and low-grade chondrosarcoma (1). Some authors (5) suggest that primary synovial chondromatosis is more likely to become malignant than secondary SC.

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New

Your child with Down syndrome. An A to Z. A practical guide for parents on the medical aspects of Down syndrome.

Josep Maria Corretger, Agustí Serés, Jaume Casaldàliga, Ernesto Quiñones, Katy Trias.

The prognosis for people with Down syndrome was bleak 25 years ago. Today, their life expectancy is around 60 years old. Medical progress has been definitive.

In the past, many clichés and misconceptions have been circulated regarding the issues and illnesses that can affect children with Down syndrome. The information on offer has been inaccurate, at times linked to confusion with the constitutional aspects of the syndrome itself. In fact, these children are subject to the same conditions as any other child of the same age and, whilst they are more prone to some, they are less prone to others. The treatment is the same and can even work better and more efficiently.
This book is aimed at parents of children with Down syndrome, providing an objective and practical introduction to the health, behavioural and developmental issues that their children may experience. It is broken down into short chapters and begins with a review of genetics and Down syndrome.
The main body of the book is set out as an alphabetical index of the medical aspects, and a final appendix contains the Catalan Down Syndrome Foundation Health Programme and Down Medical Centre (CMD) growth charts. Over thirty specialists from the CMD and renowned University Hospitals in Catalonia and Ecuador have contributed to the book.

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Tel. (+34) 93 215 74 23 or
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