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Case Report

# Primary Synovial Osteochondromatosis of the Hip Joint

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## Abstract

Primary synovial osteochondromatosis (PSO) is a benign disease characterized by the production of small bodies of hyaline cartilage released in the joint following metaplasia of the synovial membrane. Imaging techniques plays an essential role in the diagnosis of this rare entity. We report a case of PSO of the right hip in its primary form in a 23-year-old male patient.

***Keywords:*** *Primary Synovial Chondromatosis; Osteochondromatosis; Synovial; Imaging*

## Introduction

Primary synovial osteochondromatosis (PSO) is a rare benign disease characterized by the production of small bodies of hyaline cartilage released in the joint following metaplasia of the synovial membrane [1].

Imaging plays an essential role in the diagnosis of this entity and helps to differentiate it from other etiologies.

We report the case of synovial osteochondromatosis of the right hip in its primary form in a 23-year-old male patient.

## Case Report

A 23-year-old male, with no medical or traumatic history, presented with pain in his right hip and complaints of difficulty in walking since almost 4 years associated with swelling in the same area, that gradually progressed in its severity through the years. The physical examination of the right hip joint showed a limitation of the abduction, adduction and rotation movement. The patient was given symptomatic treatment but has not been relieved of his pain.

X-ray of the hip showed multiple rounded and well limited same-size calcified opacities, distributed evenly within the hip joint, with no sign of erosion of the cortical bone or osteoarthritis.

CT scan was preformed, and showed free bodies with peripheral calcifications and hypodense focal center resulting the appearance of the target sign, associated with an erosion of the acetabulum in its inferior part (Figure 1A-1C). The CT scan confirmed the peri-articular and intra-articular involvement of these calcified lesions and helped identify the ring and arc sign (Figure 1B).

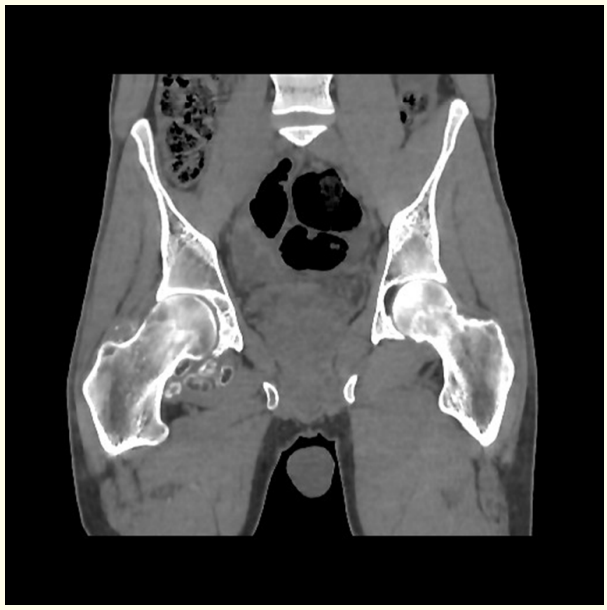

***Figure 1A:*** *Frontal CT shows multiples rounded well defined calcified lesions with a hypodense focal center resulting the appearance of the "target sign" located around the femoral neck which suggest their intraarticular localization.*

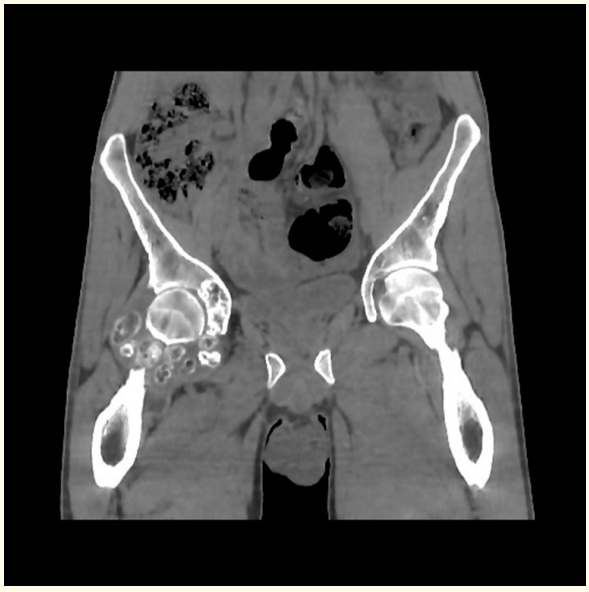

***Figure 1B:*** *Frontal CT shows the ring and arc signs associated with the erosion of the acetabulum in its inferior part resulting a "scalloping" appearance.*

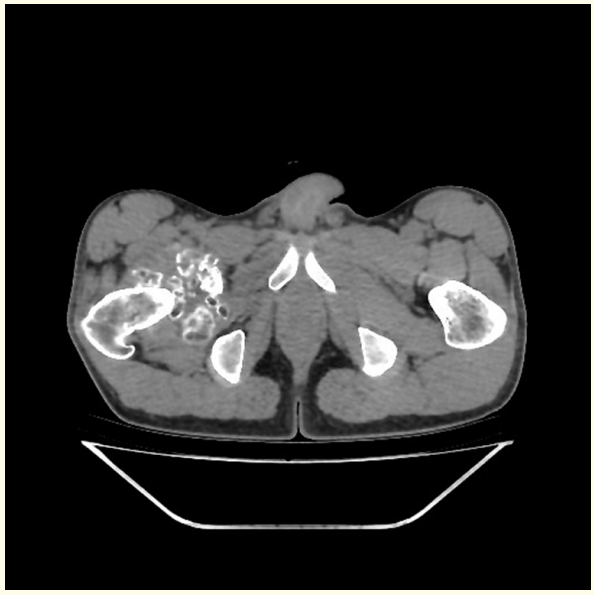

***Figure 1C:*** *Axial CT shows the localization of these lesions around the femoral neck (intraarticular and periarticular), and a slight atrophy of the gluteal muscle in comparison with the left side.*

The patient underwent a hip arthrotomy with arthroscopy and the procedure constitutes ablation of free bodies associated with a synovectomy (Figure 2). The histologic study confirmed the diagnosis of the primary synovial osteochondromatosis stage II. No complications occurred during the post-operative follow-up.

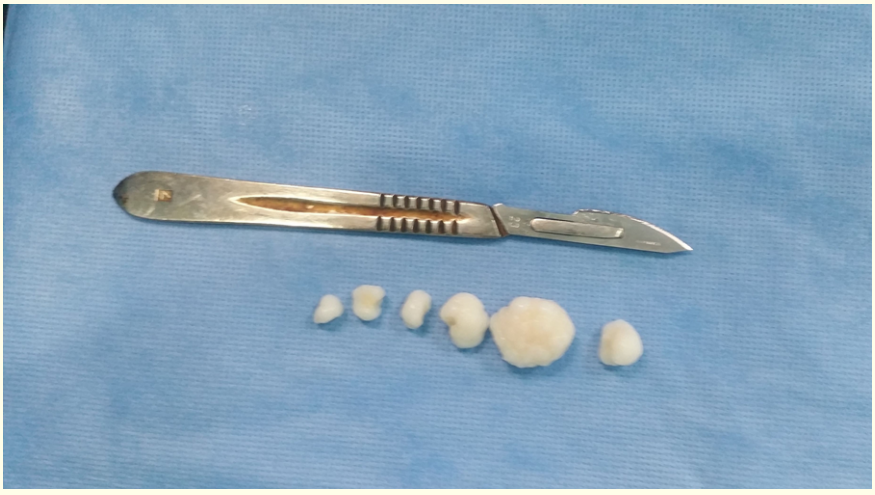

***Figure 2:*** *Samples of osteochondromas excised after surgery.*

## Discussion and Conclusion

PSO is a rare disease with an incidence of 1/100,000, studies reports a male predominance and the most affected age group is 30 - 50 years [2].

Classically, PSO is a mono-articular disease but multi-articular forms have been described in the literature [2,3]. The knee is the most commonly affected joint (50%), and involvement of the hip joint is relatively rare [2].

Clinically, PSO may manifests as chronic mechanical pain in the hip, joint swelling or even restriction of joint mobility [1]. However, there is no specific symptoms of this disease and the diagnosis is often missed or delayed due to its rarity.

Milgram describes three evolutionary phases of PSO [4] (Figure 3):

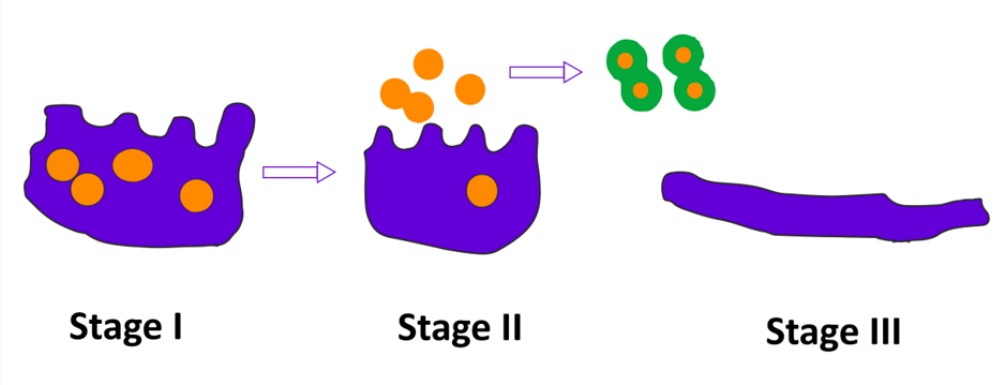


***Figure 3:*** *Stage I: Metaplasia of the synovium (purple) with internal production of chondromas (orange). Stage II: Active synovial proliferation with free foreign bodies. Stage III: Quiescent synovium and peripheral ossification (yellow) of chondromas.*

- Phase 1: It is an active phase characterized by hypertrophy of the synovium and production of chondromas which remain attached to the synovium.
- Phase 2: It is a transitional phase with active intrasynovial proliferation and free loose bodies.
- Phase 3: It is an inactive phase characterized by the presence of multiple calcified intra-articular free bodies with quiescent or discreetly inflammatory synovium.

Conventional radiography is sensitive for calcified chondromas in 70% of cases, which appears as free bodies of the same size, rounded and well limited, distributed evenly around the joint [2]. At the hip joint (which has a tight capsule) osteochondromas can be responsible for erosion of the cortical bone "scalloping" in 30% of cases [2,5].

CT is the imaging modality of choice for diagnosing calcifications. It helps identify the ring and arc sign which is very characteristic of chondroid calcifications. Free bodies with peripheral calcifications and a hypodense focal center realize the target sign which is also

characteristic and may also be seen at CT [6]. Possible bone erosions during PSO are well-visualized on CT than conventional radiography [6]. However, CT does not show marrow invasion and "stage I" chondromas as well as MRI [6].

In magnetic resonance imaging, the appearance of PSO is variable depending on the stage of the disease. In general, loose bodies are of low signal on T1- and high signal on T2-weighted images corresponding to Milgram phase 2 lesions [3,6,7]. Injection of contrast agent causes an enhancement of the inflammatory synovial signal which may reveal chondromas [3].

The treatment of PSO is surgical, based on extraction of free bodies associated with synovectomy. Incomplete synovial resection is correlated with an estimated recurrence rate of 3% to 23% depending on the case series [3,6].

Transformation into synovial chondrosarcoma is possible but rarely described in the literature. PSO of large joints (knee and hip), long-standing disease and multiple local recurrences may increase this risk [3,6].

## Authors' Statements

The authors declare that they have no conflict of interest in relation to this article.

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