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Case Report

Synovial osteochondromatosis in a 14-year-old boy with a history of Legg–Calve–Perthes disease

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ABSTRACT

We describe a case of a 14-year-old boy with a history of Legg–Calve–Perthes disease diagnosed at the age of 6 years and development of synovial osteochondromatosis of the same hip joint 7 years later. Synovial osteochondromatosis is very rare in children, and to the best of our knowledge, only a single case of Legg–Calve–Perthes disease and secondary synovial osteochondromatosis was described in the literature in a 35-year-old male, making this the first reported case of Legg–Calve–Perthes disease with development of synovial osteochondromatosis in a pediatric patient.

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Introduction

Synovial (osteo)chondromatosis is a benign condition with synovial membrane proliferation and formation of cartilaginous or osseous bodies. Synovial osteochondromatosis can be either primary or secondary to various conditions that affect the joint. Trauma, osteochondritis dissecans, and inflammatory and noninflammatory arthropathies have commonly been reported as predisposing factors in cases of secondary synovial osteochondromatosis. A possible suggested mechanism is that a cartilaginous or osteocartilaginous fragment becomes attached to a synovial membrane, and like in primary synovial osteochondromatosis, these fragments nourished by synovial fluid grow and proliferate [1].

The two types of synovial osteochondromatosis may be difficult to differentiate based on imaging alone because long-standing primary synovial osteochondromatosis predisposes to degenerative joint disease. Conversely, secondary synovial osteochondromatosis may be caused by degenerate change [2].

A useful differentiating feature is that in primary synovial osteochondromatosis, the mineralized bodies are usually numerous (normally over five) and equal sized, whereas in secondary synovial osteochondromatosis, there are fewer bodies of variable size. Primary synovial osteochondromatosis typically affects adults, predominantly men, in the third to fifth decades of life. Patients with secondary synovial osteochondromatosis are generally older than those with primary

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Fig. 1 – Supine anterior posterior with hips abducted radiograph at 6 years of age shows flattening and fragmentation of the proximal right femoral epiphysis with secondary remodeling of the acetabulum, foreshortened and broadened femoral neck, and demineralized femoral neck related to proliferative synovitis.

disease [3]. Therefore, primary and secondary synovial osteochondromatosis are quite rare in the pediatric age group [4,5].

A single report in the literature describes association between Legg–Calvé–Perthes disease and secondary synovial osteochondromatosis in a 35-year-old male, wherein the synovial osteochondromatosis presented after 27 years of follow-up [6].

We present a case of a 14-year-old male who first presented with Legg–Calvé–Perthes disease at the age of 6 years and developed radiological evidence of synovial osteochondromatosis 7 years later, at the age of 13 years.



Fig. 2 – Standing anterior posterior radiograph at 11 years of age shows mature healed Legg–Calvé–Perthes with coxa magna, advanced maturation compared to the left and mild pelvic obliquity.



Fig. 3 – Frog lateral view of the right hip at 12 years of age shows progressive chondrolysis and new intraarticular loose bodies of similar size and shape.

Case report

A 14-year-old male with right hip changes associated with known history of Legg–Calvé–Perthes disease since the age of 6 years (2008) was referred for follow-up MR imaging (August 2016). He was followed with annual radiographs and had 1 prior MR in April 2013. The radiographs demonstrated progressive changes in the hip over time. Figure 1 shows the initial image from 2008, wherein the right femoral head and neck are significantly decreased in density. The right femoral head appears flattened superiorly, and there is significant widening of the femoral neck. There is widening of the right hip joint in comparison with the left. Figure 2 shows a follow-up image obtained April 2013, wherein severe ongoing deformity of the right femoral head and neck is evident with coxa magna and coxa vara configuration of the hip. The right femoral neck is short and wide with flattening and irregularity of the femoral head and hypertrophy of the greater trochanter. There is mild periarticular osteopenia. Subtle uncovering of the femoral head with mild acetabular dysplasia is also evident. The joint space now is narrowed diffusely suggestive of cartilage loss.

On the subsequent radiograph obtained December 2014 (Fig. 3), there was a new substantial finding of numerous round calcific densities of similar size and shape seen within the right hip joint, most likely intraarticular calcific bodies. At this stage, a diagnosis of synovial osteochondromatosis was made.

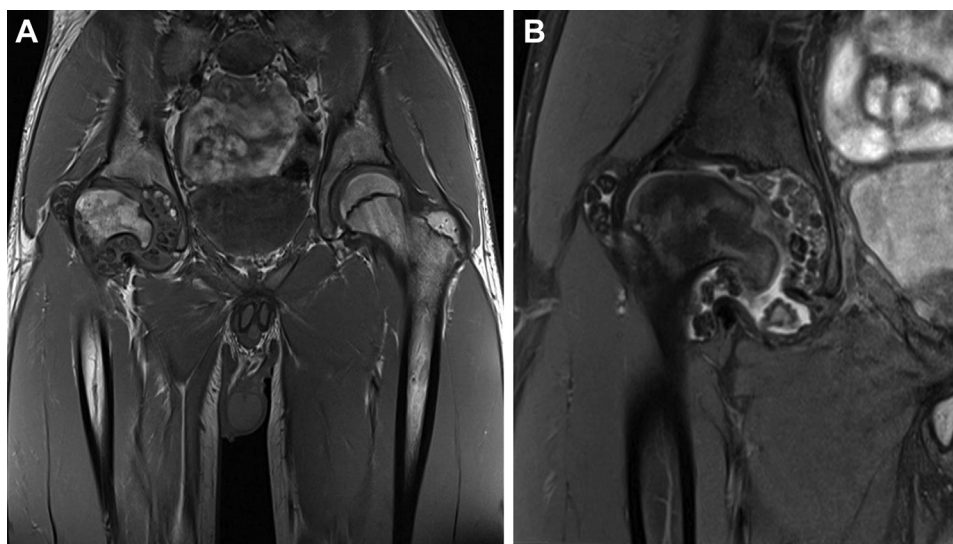


Fig. 4 – Coronal magnetic resonance imaging images on T1 (A) and proton density fat suppression (B) at 11 years of age show multiple intraarticular calcified round bodies of similar size with low signal intensity in all sequences around the head of the right femur.

It is noteworthy to mention that on the magnetic resonance imaging study obtained in April 2013, there was no evidence of synovial chondromatosis, that is, cartilaginous intraarticular bodies which are not seen on plain films were absent.

The magnetic resonance imaging obtained in August 2016 (Fig. 4) revealed multiple calcified round bodies of similar size (approximately 0.6 cm each) and low signal intensity in all sequences seen around the head of the right femur, in keeping with synovial osteochondromatosis as seen in the radiograph from December 2014.

In addition, the study revealed interval closure of the physis of the right femur, flattening of the right femoral head, decreased acetabular coverage due to lateral extrusion of the femoral head, and evidence of degenerative changes with chondromalacia. The left hip joint was unremarkable.

Although the findings on both the radiograph and the magnetic resonance imaging were more in keeping with primary synovial osteochondromatosis (ie, numerous bodies of equal size), the clinical history of slowly progressive degenerative joint disease was more suggestive of secondary synovial osteochondromatosis.

Discussion

The only previous reported case of Legg–Calve–Perthes and synovial osteochondromatosis described findings of typical secondary synovial osteochondromatosis that presented 27 years after the initial diagnosis at the age of 35 years [6].

Our case also presents a case of Legg–Calve–Perthes and synovial osteochondromatosis; however, it differs in several ways, which render it even more unique. The age of presentation of synovial osteochondromatosis in our case was much younger; synovial osteochondromatosis in the pediatric age group is quite rare as it usually presents in the 3rd–5th

decade. Additionally, the radiological appearance of the synovial osteochondromatosis was very much in keeping with the classical appearance of primary synovial osteochondromatosis, that is, multiple loose bodies of similar size. As Legg–Calve–Perthes disease has been described in association with osteochondritis dissecans [7] and with chronic synovitis [8] and since these conditions have been associated with secondary synovial osteochondromatosis [1], we assume that the changes associated with Legg–Calve–Perthes in our case render secondary synovial osteochondromatosis the more likely diagnosis. Nonetheless, secondary synovial chondromatosis can only be distinguished histologically from primary synovial chondromatosis [3]; however, a histological specimen was not obtained in this patient.

Conclusion

We hereby presented a case of Legg–Calve–Perthes disease with development of synovial osteochondromatosis occurring in the pediatric age population. Although a direct link between the two pathologies cannot be established based on this single case report, we do suspect that possibly a loose body generation from osteonecrosis or chronic synovitis in Legg–Calve–Perthes disease could be a predisposing factor for secondary synovial osteochondromatosis.

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