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Progressive bilateral lipoma arborescens of the knee complicated by juvenile spondyloarthropathy: A case report and review of the literature

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ABSTRACT

Objectives: To report an unusual case of lipoma aborescens (LA) presented in a patient with treatment-responsive juvenile spondyloarthropathy (JSPA) and to summarize the clinical manifestations, therapy and prognosis of LA by literature review.
Methods: We report an atypical case of a 17-year-old patient with an initial presentation of juvenile spondyloarthropathy, whose inflammatory condition was improved successfully by traditional anti-rheumatic drugs and an anti-TNF alpha agent but developed progressive swelling of bilateral knees. Lipoma arborescens were diagnosed in each knee by synovial biopsy obtained by arthroscopic surgery. Fifty-one cases of LA have been reported and are reviewed in detail.
Results: Clinically, LA could present as monoarthritis or oligoarthritis. The lateral compartment of the knee is the most common site of involvement. Several cases were reported as a comorbidity of inflammatory diseases, but were not improved by anti-inflammatory therapy. Most patients were diagnosed by classic MRI and biopsy findings. The lesions can be managed by open or arthroscopic surgery, but a minority of the cases may have reoccurrence in the same or opposite joint.
Conclusions: LA is a very rare lesion of the synovial and bursal tissue with an unknown etiology. It is considered to be a benign proliferation of the synovial fat associated with trauma, degenerative or inflammatory conditions. LA should be considered as a secondary or comorbid condition in inflammatory arthropathies if other joints respond well to intensive therapy and one or more do not.

Lipoma arborescens (LA) is reported as a rare intra- or extra-articular lesion occurring in various joints, with the knee being the most commonly involved one. Most cases have been described in elderly patients with degenerative or post-traumatic joint disease, but in several case reports it has been considered to be related to inflammatory joint diseases. Here we describe an atypical case in a patient with an initial presentation of juvenile spondyloarthropathy (JSPA), also considered as a subtype of juvenile idiopathic arthritis (JIA), followed by the development of synovial lipoma arborescens in bilateral knees during anti-TNF alpha therapy.

Case report

A Chinese girl presented with recurrent joint pain and swelling in both knees, left ankle and left hip, and was diagnosed with JSPA at the age of 12. She was eventually diagnosed with LA at the age of 16. There was no history of trauma or other illness. At the initial presentation, the sacroiliac CT scan showed left iliac bone erosions (Fig. 1A and B) with serum HLA-B27 positivity, elevated levels of ESR (44 mm/h) and increased levels of C-reactive protein (38.6 mg/dl). Juvenile spondyloarthropathy was diagnosed based on the sacroiliac CT scan and a positive HLA-B27 allele result. Diclofenac acid and sulfasalazine (SSZ) were given for 3 months without much improvement. Then the TNF alpha antagonist (Recombinant Human Tumor Necrosis Factor-a Receptor II:IgG Fc Fusion Protein for Injection) was instituted after the initial diagnosis. She received 25 mg weekly for about two years. The pain and swelling in the left hip and ankle improved and has subsequently been stable. However, progressive swelling with mild pain in both knees began to develop. Methotrexate (MTX) at 10 mg/wk was also administered during the past 2 years. There was still no improvement, nor was there improvement in bilateral pain and swelling despite treatment with the combinations of diclofenac, SSZ, MTX and anti-TNF alpha agent. On physical examination, there was a large-sized bilateral knee joint effusion without bony tenderness, as well as a mild limitation in range of motion. All other joints were normal. The muscle strength of both lower extremities was limited at four out of five.

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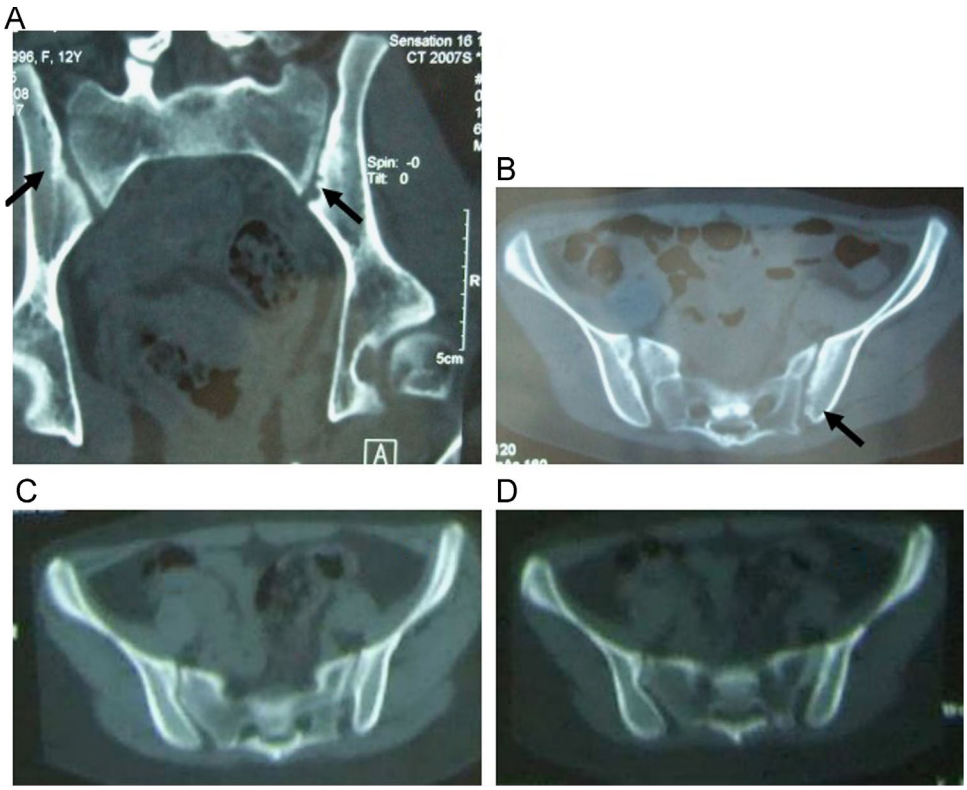

Fig. 1. CT scan of sacroiliac joints in 2008 (A and B) showing bone erosions in the left iliac edges (arrow); repeated scan showed the erosions were repaired in sacroiliac joints in 2012 (C and D).

An MRI examination of both the knees demonstrated synovial hypertrophy with large-size joint effusions, which showed different signal intensity on Tl-weighted, T2-weighted, STIR, and contrast images (Fig. 2A–D). The bones, ligaments, and articular cartilage appeared normal. The MRI image findings were not definitely diagnostic for LA at that time. However, a sacroiliac joint CT scan revealed repair of the prior bone erosion (Fig. 1C and D).

An arthroscopic synovectomy of both the knees was performed 6 months prior to this report. Each knee contained a large number or finger-shaped synovial proliferations with yellow glistening villous masses in the suprapatellar bursa, patellofemoral joints and intercondylar fossa (Fig. 3A and B). Moderate amounts of villous synovial masses covered the superficial tibial plateau and femoral cartilage. Fibrillation of the articular cartilage was noted in the intercondylar fossa. Histopathological examination of the villous masses demonstrated that they were composed predominantly of adipose tissue with constituent blood vessels and mixed chronic inflammation. The surface was lined in places by synovial cells (Fig. 3C and D). LA was then confirmed by the diagnostic histopathology findings.

Six months after the therapeutic surgery, the patient had continued resolution of bilateral knee swelling and improvement in range of motion. The TNF alpha blocker was tapered and was discontinued.

Literature review

A systematic literature search was performed using subject term ‘‘Lipoma Arborescens’’ in PubMed to identify articles from 1950 through May 2012. The language was restricted to English. Review articles, articles with important missing information and those articles not found in full text were excluded.

In total, 114 articles were identified by the search criteria, of which 16 non-English articles, as well as 17 review articles and 28 articles without full text were excluded from the review. Forty-seven articles [1–47] with comprehensive clinical and laboratory data from 51 patients (including our patient) were analyzed in detail including race, age of onset, gender, disease duration, symptoms, pattern of arthropathy, comorbidities, MRI images, pathologic lesions and prognosis (Table 1).

The racial distribution of the 51 patients in the reviewed articles was Asian (45%), Caucasian (45%), African (6%) and Oceanian (4%). The age range of disease onset was from 10 to 80 years, and the mean age was 37.8 $\pm$ 18.9 years. There were 10 patients whose onset age was below 18 years (20%). The ratio of females to males was 22:29 and disease duration ranged from 1 to 240 months, the average duration was 39.2 $\pm$ 47.6 months. Thirty-six (71%) of the 51 patients reviewed in the literature had follow-up information. The average length of follow-up duration was 21.5 $\pm$ 23.9 months. Joint fluid was studied in 12 patients (24%); findings were nonspecific, including predominantly yellow fluid, increased white cell count (200–11,360 mm^{-3}), elevated lymphocyte counts (3/12), and elevated neutrophil granulocyte counts (4/12).

The pattern and different areas of the involved joints have been summarized for the 51 cases. Knee joint involvement with severe swelling was the most common clinical manifestation of LA, and in 62.2% of the patients knee involvement was unilateral. Other areas that were involved included the elbow [5,13], shoulder [27], ankle, hip, wrist, hand or foot. The majority of LA patients had no joint pain or only mild pain with motion. For all the patients reviewed, the main pattern of the joint involvement was monoarthritis (66.7%) and oligoarthritis (25.5%). Only 4 patients (8%) were reported with polyarthritis complicated by inflammatory arthritis [3,11,16,24]. LA in the bicipital bursa has also been identified in other patients [36,38].