Radiology Case Reports 15 (2020) 218–221

Available online at www.sciencedirect.com

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Tumefactive synovial thickening mimicking synovial chondromatosis in the setting of oligoarticular juvenile idiopathic arthritis in a toddler

Lee K. Rousslang, MD[a,*], *Elizabeth Rooks*[b], *Caleb Hixson, DO*[a], *Priscilla West, MD*[a]

[a] *Tripler Army Medical Center, 1 Jarrett White Road Tripler AMC, Hawaii 96859*
[b] *Duke University, 2080 Duke University Road, Durham, NC 27708*

ARTICLE INFO

Article history:
Received 10 September 2019
Revised 22 November 2019
Accepted 24 November 2019

Keywords:
Juvenile idiopathic arthritis
Synovial chondromatosis
Tumefactive synovium

ABSTRACT

Juvenile idiopathic arthritis (JIA) is the most common cause of arthritis in children. It is characterized by inflammatory cell infiltration of synovial membranes leading to synovitis and synovial membrane thickening. Synovial chondromatosis is rare sequela of synovitis in which foci of cartilage develop within the synovial membrane of a joint capsule. We report a case of a 35-month old boy who developed tumefactive synovial hypertrophy and hyperplasia that mimicked synovial chondromatosis on MRI. The reactive synovium was arthroscopically excised and the patient's arthritis improved. Tumefactive synovial hypertrophy and hyperplasia mimicking synovial chondromatosis on MRI in the setting of JIA, has not been reported in this young of a patient in the literature to date. A discussion on imaging in oligoarticular JIA and synovial chondromatosis is presented herein.

Introduction

JIA is the most common arthritis in children, of which, oligoarticular JIA is the most common subtype. It is associated with a variety of joint findings, with inflammation and hypertrophy of the synovium being the principle component of the disease process. One such finding is synovial chondromatosis, a benign neoplasm of synovium that forms foci cartilaginous nodules, and can be primary, or secondary to trauma, neuropathic osteoarthropathy, osteoarthritis, inflammatory arthritis, or infection, and appears as loose intra-articular cartilaginous bodies with distinct imaging characteristics. This case describes a toddler with oligoarticular JIA whose MRI demonstrated multiple separate ovoid filling defects in the synovial capsule of the knee, with signal characteristics consistent with synovial

Acknowledgements: The views expressed in this manuscript are those of the authors, and do not reflect the official policy or position of the Department of the Army, Department of Defense, or the United States Government. The authors have no financial, personal, or other vested interests in the information contained within this document.

* Corresponding author.
E-mail address: lee.k.rousslang.civ@mail.mil (L.K. Rousslang).

https://doi.org/10.1016/j.radcr.2019.11.016

chondromatosis. The patient was treated with a partial synovectomy with improvement of symptoms. Pathology of the resected specimen demonstrated synovial hypertrophy and hyperplasia, accompanied by inflammatory cells, making it a mimicker of the presumptive presurgical MRI impression of synovial chondromatosis. We describe a case of tumefactive synovial thickening mimicking synovial chondromatosis on MRI resulting in prolonged arthroscopy for presumed synovial chondromatosis.

Case report

A 35-month-old boy presented to the emergency department with 3 weeks of pain and limping exacerbated by running. The father reported no preceding illness, trauma, and that the boy was otherwise well. On examination, there was a mobile mass just superior to the right patella, along with a joint effusion. The joint was not tender to palpation, but range of motion of the right knee was limited from 10-130 degrees, and the boy walked with an antalgic gait. Nonsteroidal anti-inflammatory medication was prescribed for pain with minor relief.

Radiographs of the knee revealed a joint effusion with soft tissue swelling, but no osseous abnormality or evidence of osteomyelitis. The patient was then referred to orthopedics for further evaluation. Complete blood count revealed a normal white blood cell count. The erythrocyte sedimentation rate was elevated at 57 mm/h (0-15 mm/h), and C-reactive protein returned within normal limits.

Due to persistent pain and limp MRI pre and post gadolinium contrast were performed, and demonstrated multiple separate ovoid filling defects within the knee joint effusion, the largest of which measured 1.5 cm. The ovoid intra-articular presumed loose bodies did not enhance post gadolinium, and were isointense on T1 and isointense to slightly hypointense on proton density and T2 weighted sequencing, following signal intensity of cartilage. The expected synovial hypertrophy with wavy enhancement and synovial thickening with mixed intermediate to high T2 signal and heterogeneous mixed enhancement with central fibrin and hemosiderin deposition in the synovium of synovial proliferation was not evident on the MRI. Given these findings, synovial chondromatosis was the main concern. Interestingly, there was also a horizontal course of the anterior cruciate ligament, confirmed at arthroscopy, presumed to be due to the abundance of synovial thickening distending the joint.

Arthroscopy demonstrated pearly white ovoid structures that were easily grasped and not chondroid in texture with an inflamed, hypertrophic synovium, but no chondroid intra-articular loose bodies. A synovectomy was performed and the patient was discharged after 48 hours. Upon follow-up 2 weeks later, the patient demonstrated increased ROM, resolution of his antalgic gait, and his knee effusion improved and swelling decreased.

Pathologic analysis of the samples revealed hypertrophic and hyperplastic synovium with reactive change, and scattered histiocytes and lymphocytes on immunohistochemical staining. There was no evidence of synovial chondromatosis, and the patient was referred to pediatric rheumatology.

Rheumatology laboratory result demonstrated an elevated anti-nuclear antibody of 1:640 (≤1:40), with a normal rheumatoid factor level. Rheumatology also performed a non-dilated eye exam revealing possible cataracts, and the patient was referred to ophthalmology where bilateral cataracts was confirmed, as well as synechiae, and patient scheduled for cataract extraction with synechiaelysis.

Discussion

Juvenile Idiopathic arthritis is the most common type of arthritis in children, and oligoarticular JIA is the most common subtype, comprising approximately half of all JIA cases, and is defined as JIA involving fewer than 5 joints. Oligoarticular JIA is more common, and in fact has a peak incidence in the second and third years of life, but is a diagnosis of exclusion [1]. Although JIA is considered idiopathic, it is increasingly thought that autoimmunity may be a factor [2]. In oligoarticular JIA, systemic symptoms are characteristically absent, with the exception of uveitis. On ophthalmologic exam, our patient was found to have significant bilateral cataracts, a sequela of recurrent uveitis, consistent with oligoarticular JIA. Additionally, the only lab abnormality typically found in oligoarticular JIA is an elevated ANA, which our patient also had [3]. An elevated erythrocyte sedimentation rate (ESR) was also present in our patient, and is also a common finding, and confers increased risk of disease progression to polyarticular disease [3,4].

The knee joint is commonly affected in oligoarticular JIA, with early radiographs showing soft tissue swelling, joint effusion, and osteopenia. Late stage radiographs can demonstrate subchondral sclerosis, ankyloses, joint space narrowing, and erosions [5]. In contrast to radiographs, MRI has been shown to be more sensitive for detecting synovitis, a potentially important prognostic indicator, [6,7] as well as bone marrow edema, a potential indicator of future erosions, and an indication to initiate therapy [8]. MRI is also useful for detecting cartilage lesions, and synovial hypertrophy [9]. MRI findings of SC most commonly demonstrate lesions that are hypointense or isointense to muscle on T1 and have high signal intensity on T2, with or without focal areas of low signal intensity [10,11]. Areas of signal void correspond to calcifications that frequently occur in SC were not seen in this case [10]. In this patient, the lesions were nonenhancing on MRI. This may be because the tumefactive synovium was in a fibro-inflammatory state, involving synoviocyte proliferation and metaplasia, on its way to forming synovial chondromas, which have a limited blood supply and are characteristically nonenhancing [11].

The present case demonstrates an unusual form of tumefactive synovial hypertrophy resembling ovoid loose bodies normally seen in synovial chondromatosis. The characteristic intra-articular bodies on MRI were thought to be consistent with SC criteria. There was enhancement of the synovium but no enhancement of the ovoid bodies. (Figs. 1 and 2)

During arthroscopy, the entire synovial capsule was visualized, and multiple pearly separate white ovoid structures were seen, which upon forceps grasp, were soft and easily removed, most consistent with synovial thickening. (Fig. 3) Pathology of

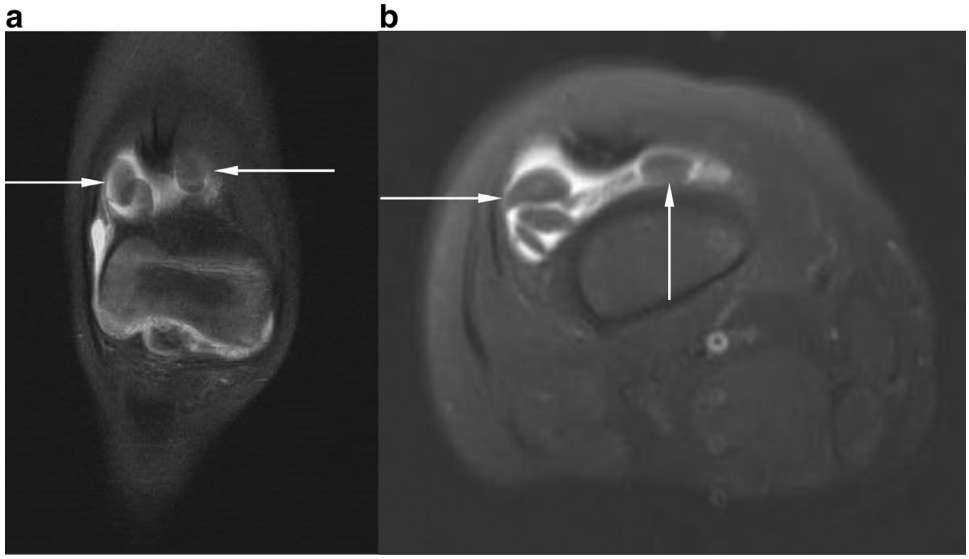

Fig. 1 – (A) Coronal, and Fig 1B, Axial T2, demonstrate the separate ovoid structures with surrounding synovial fluid (arrows).

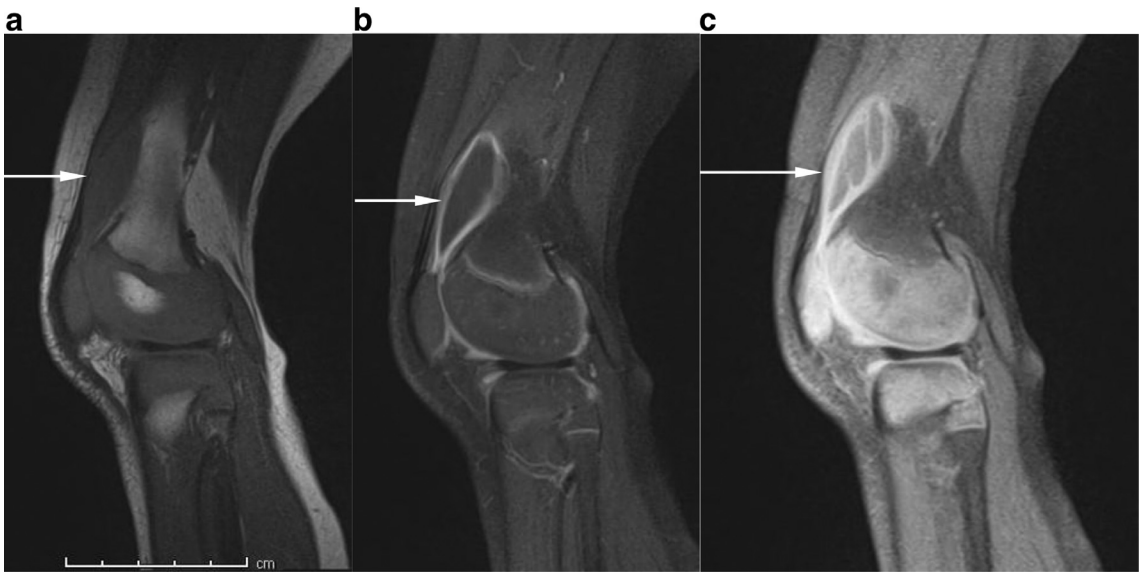

Fig. 2 – (A), Sagittal T1 demonstrates bowing of the prefemoral joint space (arrowhead). (B) Sagittal T1 post Gadolinium shows enhancement of the synovial lining without delineation of the central elements (arrow). (C) Sagittal Proton Density demonstrates separate ovoid structures (arrow) within the suprapatellar recess with similar signal intensity to the adjacent cartilage.

the ovoid structures confirmed that they were "hyperplastic and hypertrophic synovium," with "increased synovial fibroblasts and compact fibrous tissue" and "histiocytes and lymphocytes," consistent with our assertion of a fibroinflammatory state. (Fig. 4) No chondroid elements were noted on the pathologic specimens, and there was no evidence of synovial chondromatosis.

Further pathologic, ophthalmologic, and laboratory workup of yielded several markers of JIA, including synovial hypertrophy and hyperplasia, elevated ESR, ANA, and bilateral cataracts, all consistent with JIA.

In this case, the diagnosis was complicated because the child had imaging findings consistent with SC, and operative findings visually suggestive of chondral bodies. However, the bodies were soft upon forceps grasp, and no chondroid elements were detected on pathologic analysis. Given the imaging features on MRI, and the absence of discernable chondral bodies on arthroscopy or pathology, this patient was determined to have tumefactive synovial hypertrophy and hyperplasia or the *forme fruste* of synovial chondromatosis in the setting of oligoarticular JIA. A comprehensive research query yielded no cases similar to this in the published literature. This case report stresses the importance of the differential diag-

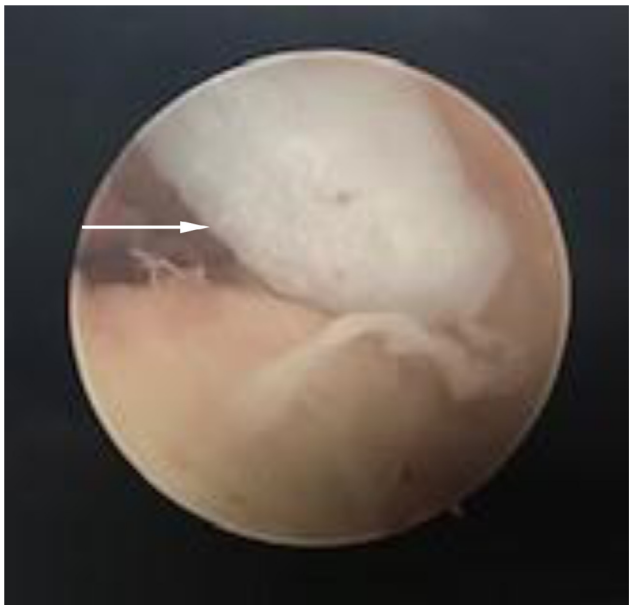

Fig. 3 – Arthroscopic image in the coronal plane demonstrates the pearly white rounded ovoid tissue which was soft and pliable with forceps grasp and easily removable.

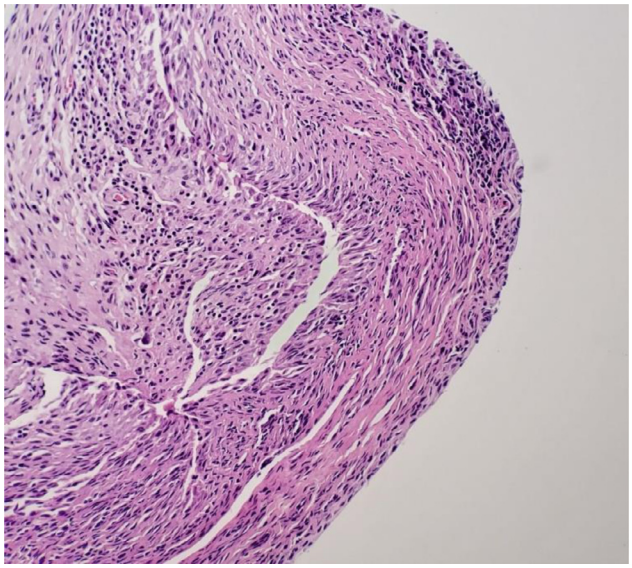

Fig. 4 – Microscopically, the resected synovial membrane demonstrates synoviocyte hypertrophy and disorganized cellular hyperplasia, most apparent at the surface intima and subintima tissue level (pictured). Some histiocytes and scattered lymphocytes are seen, highlighted by immunohistochemical stains CD163 and CD45, respectively. There are also increased numbers of synovial fibroblasts, increased intervening compact fibrous tissue, and a loss of extracellular matrix.

nosis be communicated to the pediatric surgeon for planning purposes.

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