

## A Rare Case of Concomitant Intra-Articular and Extra-Articular Synovial Chondromatosis of the Knee Joint

### Dear Editor,

Synovial chondromatosis is an uncommon metaplastic disease in which foci of ectopic cartilage develop from the synovium.<sup>1</sup> Despite the known benign nature of this disease, loose bodies can occur when these osteo-cartilaginous lesions grow and detach from the synovium. This may result in disabling mechanical symptoms as well as accelerated degeneration of the affected joint due to irritation and wear. The symptoms can otherwise be non-specific, with patients complaining of pain, swelling or limitation of motion. It is known that synovial chondromatosis occurs more frequently in men, with increasing frequency during the third to fifth decades of life.<sup>2</sup> The disease most commonly affects the knee joint, with the typical location being intra-articular.<sup>3,4</sup> Extra-articular involvement of the bursae and tenosynovium is less common.<sup>5</sup> Combined intra- and extra-articular involvement of the knee joint is very rare with limited reports in the existing literature.<sup>4,6,7</sup> We describe a case of a patient who presented with coexisting intra- and extra-articular synovial chondromatosis of the knee joint and highlight the importance of adequate imaging prior to surgical treatment.

### Case Report

A 63-year-old man presented with right popliteal fossa swelling for 3 years. He noted a recent increase in posterior knee pain accompanied by enlargement of the popliteal swelling over 3 months. He had increasing difficulty in squatting. There was no history of trauma or constitutional symptoms and the patient had an unremarkable medical history. In terms of social history, the patient worked as a taxi driver and did not participate in any sports. On physical examination, the popliteal mass was found to be bony hard, lobulated and tender on palpation. The patient had restricted knee movement with a range of motion of 10 to 100 degrees and mild medial joint line tenderness.

Plain antero-posterior and lateral radiographs of the right knee (Fig. 1) revealed multiple calcified bodies in the popliteal fossa and at the posterior aspect of the right knee on a background of moderate osteoarthritic changes. Magnetic resonance imaging (MRI) (Fig. 2) revealed tri-compartmental osteoarthritis, as well as multiple calcified bodies that were largely extra-articular. Intra-articular loose bodies with stippled calcifications were also noted.



Fig. 1. Antero-posterior and lateral radiographs of the right knee showing calcifications in posterior aspect of the knee on the background of osteoarthritis of the knee.



Fig. 2. A) T1-weighted sagittal image of the right knee showing intra-articular chondromatosis (arrow). B) Proton density fat suppressed sagittal image of the right knee showing more extensive extra-articular chondromatosis (arrow).

A Baker's cyst measuring 7.1 cm x 3.1 cm x 4.4 cm was found communicating with the knee joint posteriorly.

A diagnosis of synovial chondromatosis was made based on the clinical findings and imaging. Surgical excision was advised and the patient underwent a 2-stage operation at the same sitting. In the first stage, open excision of the right knee Baker's cyst was performed in the prone position. A lazy S-shaped incision was made over the popliteal fossa. The cyst was completely excised at its origin from the knee joint. More than 20 chondromatous bodies of various sizes (measuring up to 1.5 cm in diameter) were found in

the popliteal cyst, some of which were still attached to the cyst wall (Fig. 3).

The second stage involved arthroscopic removal of the intra-articular loose bodies (Fig. 4). Standard anteromedial and anterolateral knee portals were created with the patient in supine position. Degenerative changes were noted in the medial and patella-femoral compartment. Loose bodies were identified at the posterior aspect of the knee. A third posterolateral incision was made to facilitate the removal from the posterior compartment and 5 loose bodies were removed: 2 posteromedial, 2 posterolateral and 1 posterior to the posterior cruciate ligament.

Postoperatively, the patient's recovery was uneventful. Posterior knee pain improved and the patient discharged well. At 3 years follow-up, there was no clinical recurrence of the disease. The patient had a range of motion of 5 to 110 degrees, which was equivalent to that of his left knee. There was residual mild medial joint line tenderness from the underlying osteoarthritis.

### Discussion

Synovial chondromatosis is an uncommon benign disease where ectopic foci of cartilage form in synovial tissue. The exact aetiology is unclear, although the theory of synovial metaplasia is favoured.<sup>1</sup> The chondromas enlarge on the synovium and may subsequently break off to form loose bodies. A proportion of these chondromas undergo ossification to form calcified nodules—often termed as osteochondromas. The typical manifestation is usually mono-articular.<sup>3</sup> In addition, it often occurs



Fig. 4. Intra-operative arthroscopic photo showing removal of the loose body.

in an intra-articular location, although extra-articular involvement of the bursae and tenosynovium has been reported.<sup>5</sup> For intra-articular manifestations, the knee is the most common site of disease occurrence, followed by the hip,<sup>1</sup> while extra-articular disease tends to affect the hands and feet.<sup>8</sup> Clinically, disease manifestations are non-specific and include swelling, pain, crepitus and limitation of joint motion. Plain radiographs are a simple first-line investigation that can be used to detect ossified nodules. As demonstrated in our case, it is important to obtain further imaging to better delineate the origin of the disease. X-rays are unable to define the exact location of the disease and furthermore, may not be able to detect uncalcified chondromas. If the physician has any doubt regarding the intra- or extra-articular nature of the lesions, it is prudent to obtain MRI or computed tomography to better visualise the extent prior to surgery. The location of the lesions will affect preoperative planning and counselling to the patient. MRI can also rule out conditions such as tumours that may mimic loose bodies with symptoms of locking.<sup>9</sup>

Primary and secondary forms of the disease have been described. The 2 entities are histologically distinct, although both share the common pathway of synovial metaplasia. Primary synovial chondromatosis (PSC) occurs spontaneously without an inciting agent whereas secondary synovial chondromatosis (SSC) arises against a background of joint disease (e.g. osteoarthritis, inflammatory arthritis, osteochondritis dissecans, trauma or infection) and results from the implantation of chondral debris onto synovial tissue, stimulating metaplastic cartilage proliferation. At the microscopic level, PSC is characterised by discrete nodules of disorganised metaplastic cartilage with marked nuclear atypia, mitotic activity and irregular calcification (in contrast to secondary synovial chondromatosis where



Fig. 3. Clinical image of several chondromas up to 1.5 cm in diameter. Several chondromas are seen attached to the cyst wall.

the chondrocytes are without cytogenic aberrancy and demonstrate an even, uniform arrangement and a concentric pattern of calcification).<sup>10</sup>

This patient's history of osteoarthritis and intra-articular involvement suggests a diagnosis of SSC. Histopathological examination of the nodules showed laminar arrangement of dispersed chondrocytes with variable mineralisation and lack of chondrocyte atypia, pleomorphism or mitotic activity, confirming SSC as the underlying pathology. Chronic inflammatory changes were seen in the synovium from both the Baker's cyst and knee joint. There was no evidence of malignancy.

In our case, the diagnosis of SSC correlates poorly with the macroscopic findings of a large number and small, relatively homogeneous size of chondromas, which are more consistent with PSC. In contrast to the current presentation, chondromas in SSC are typically larger, less numerous and of greater size variation due to their different times of origin.<sup>11</sup> Significantly, the chondral bodies found attached to the popliteal cyst wall intraoperatively likely suggest an extra-articular bursal metaplastic origin. It is the authors' opinion that the chondromatosis originated from the bursal synovium, with subsequent extrusion into the posterior joint space via direct communication with the bursa. Incidental intra-articular involvement is further supported by the comparatively fewer loose bodies found in the joint, sole involvement of the posterior joint compartment and the lack of visible foci of chondral proliferation on the intra-articular synovium. Again, this is atypical of SSC where intra-articular synovial involvement precedes extra-articular dissemination, if any.

In the literature, there are very few reported cases of combined intra- and extra-articular synovial chondromatosis of the knee joint.<sup>4,6,7</sup> Bassir et al<sup>7</sup> reported a case of bilateral intra- and extra-articular involvement in the knee joints, with prominent intra-articular involvement. Two other reports also described extensive disseminated disease, predominantly with intra-articular involvement.<sup>4,6</sup> The surgery offered was similar to our case, being a 2-stage procedure with knee arthroscopy and open exploration of the popliteal fossa. This case differs with its more prominent bursal involvement and active bursal intra-synovial disease. It also presents an atypical macroscopic manifestation of SSC.

Management of symptomatic disease is surgical, with arthroscopic treatment preferred for intra-articular manifestation and open approaches reserved for extra-articular or extensive involvement.<sup>7,12-14</sup> Removal of the chondromatous lesions provides relief from mechanical symptoms and prevents progression of articular damage by irritant loose bodies. The role of synovectomy in preventing disease recurrence is controversial. Milgram described different treatment strategies based on the presence of intra-

synovial disease.<sup>1</sup> For intra-synovial disease without loose bodies, he advocated synovectomy whilst simple removal of loose bodies without synovectomy was reserved for the inactive phase without intra-synovial disease. The literature is divided on the effect of synovectomy on recurrence outcomes. Oglivie-Harris et al<sup>14</sup> showed decreased recurrence rates in patients treated with synovectomy whilst Maurice et al<sup>15</sup> and Shpitzer et al<sup>16</sup> reported no differences in disease recurrence rate regardless of whether synovectomy was performed in tandem with chondroma removal.

In our case, knee joint synovectomy was not performed due to minimal intra-articular involvement. Active synovial disease in the popliteal bursa was managed by complete excision of the cyst containing the osteochondral bodies. The patient remains well 3 years postoperatively with no recurrence of disease. Should there be disease recurrence, a possible consideration in the treatment of SSC on the background of painful disabling osteoarthritis is arthroplasty. This may benefit the patient by removing disease as well as improving pain.

## Conclusion

Concomitant intra- and extra-articular synovial chondromatosis of the knee joint is a rare entity. Adequate imaging should be performed prior to operative treatment in order to accurately delineate the location of the lesions. Such extensive disease can be treated effectively with knee arthroscopy and open resection of the posterior compartment.

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