

# Synovial Lipomatosis (Lipoma Arborescens) Affecting Multiple Joints in a Patient with Congenital Short Bowel Syndrome

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**ABSTRACT.** Synovial lipomatosis is a rare, synovial based disorder that typically affects a single knee. Magnetic resonance imaging and synovial biopsy established this diagnosis in a patient who presented with a 20 year history of painless swelling of the knees, wrists, and hands. Such extensive involvement, particularly of hand joints and tendon sheaths, has not been described previously. Superimposed on this chronic soft tissue joint swelling were acute knee effusions, which responded to intraarticular steroid injections. The co-occurrence with congenital short bowel syndrome may have etiological implications for this disorder of inappropriate fat deposition. (J Rheumatol 2002;29:1088–92)

*Key Indexing Terms:*

SYNOVIUM

LIPOMATOSIS

LIPOMA ARBORESCENS

VILLOUS PROLIFERATION

CONGENITAL SHORT BOWEL

Synovial lipomatosis (SL) is a rare, articular condition of unknown etiology, characterized by hypertrophic fat infiltration beneath the synovium. The proliferative synovium is filled with mature adipose cells, often strewn with enlarged or congested hyperemic capillaries<sup>1,2</sup>. The condition is also commonly known as lipoma arborescens, referring to the frond-like appearance of the synovial proliferation, although “lipoma” has a misleading connotation. Hallel, *et al* proposed the term “villous lipomatous proliferation of the synovial membrane” to reflect the characteristic gross, radiographic, and histologic appearance of the process<sup>1</sup>. However, it may not be villous in all cases, so recent authors have suggested naming the disorder SL<sup>3</sup>. Typically, SL affects a single knee joint. We describe an individual with several unusual features including extensive disease of the joints and tendon sheaths of the wrists and hands.

## CASE REPORT

A 35-year-old Caucasian male presented with worsening bilateral knee swelling. Painless, intermittent swelling of knees, wrists, and hands,

considered by the patient to be of only a minor inconvenience, had been present for more than 20 years. In the past, he had ignored transient acute increases in the size of the knee(s), but the recent massive enlargement led to decreased range of motion, prompting him to seek medical attention. No rheumatologic investigation had previously been undertaken, although soft tissue enlargement and effusions had been noted on knee radiographs 10 years earlier. His history was significant for frequent gastrointestinal complaints during childhood. An exploratory laparotomy at St. Louis Children's Hospital at age 15 revealed the length of his small intestine to be 90 inches, one-third of normal. Since then he has received supplemental parenteral nutrition. He denied joint trauma, morning stiffness, constitutional symptoms, or family members with swollen joints.

Examination showed boggy, noninflammatory appearing soft tissue swelling of both wrists and hands (metacarpophalangeal and interphalangeal joints). There was thickening of the extensor tendon sheaths over the wrist and dorsum of the hands. Symmetric tense knee effusions were not warm, erythematous, or tender but did restrict range of motion. The remainder of the musculoskeletal evaluation and the general physical examination were normal. About 1000 ml of yellow tinged fluid was aspirated from each knee. The fluid was not milky or chylous in appearance. The total cell count was 7840 with 1900 white blood cells (WBC) (85% segmented neutrophils, 11% lymphocytes, 4% macrophages). Similar fluid (300 to 500 ml each time) was aspirated from each knee weekly for the next 21 days. WBC counts ranged from 20,000 to 80,000 with roughly 90% segmented neutrophils. Crystal analyses, gram stains, and cultures were negative. Complete blood counts, electrolytes, uric acid, serum protein electrophoresis, cryoglobulins, serum lipid profile, and urinalysis were normal or negative. Antinuclear antibodies and rheumatoid factor were negative on 2 occasions 8 months apart. Erythrocyte sedimentation rate was slightly elevated at 29 mm/h. Radiographs of hands, wrists, and knees were remarkable for the soft tissue swelling but there were no erosions or other abnormalities.

Because of the inflammatory features of the effusions, triamcinolone acetonide (80 mg) was injected into each knee at the 4th week. This prevented reaccumulation of the effusions; however, soft tissue swelling of the knees persisted. Over the next 6 months, because of the inflammatory characteristics of the effusions and the marked soft tissue swelling, a trial of methotrexate (up to 20 mg) was administered, but this had no effect on the soft tissue swelling of any joint.

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On magnetic resonance imaging (MRI), synovial masses with frond-like lobules protruded into the knee joints, most prominently in the suprapatellar bursae. The signal intensity of these lobules was high on T1 images (Figure 1A) and very low on T2 images (Figure 1B), but was consistently isointense with subcutaneous fat. Examination of the wrists showed similar fat lobules within the synovial sheaths of both dorsal and volar tendons (Figure 2). There was also thickening of the quadriceps tendons. Fluid surrounded the fat lobules in the knee joints and within all synovial sheaths of the wrists. Overall, the MRI appearance was specific for SL and the diagnosis was confirmed by a synovial biopsy of the wrist (Figure 3). Methotrexate was discontinued and the patient declined synovectomies.

## DISCUSSION

Most cases of synovial lipomatosis have been reported in the last decade. In our literature review, we found 63 cases including those reported in several recent radiologic series. Age at the time of presentation ranged from 9 to 90 years, without a predilection for an age group. Males represented 70% of the patients. The suprapatellar pouch was the most common site, with 43 patients having single knee involvement and 12 additional patients with lesions in both knees. Only 8 cases with involvement of other locations have been described (Table 1). To our knowledge, SL affecting hand joints and tendons has not been reported before and there has been only one report of wrist involvement. In 1957,

Napolitano described a 47-year-old women with wrist lesions that were attributed to tuberculosis, which may have been SL<sup>4</sup>. In addition to bilateral wrist and knee involvement, our patient had prominent synovial swelling affecting multiple small joints of the hands. Moreover, he had extensive extensor and flexor tendon sheath involvement in the hands. There was also profound thickening of quadriceps tendons, which has been described once before<sup>5</sup>.

The usual clinical presentation of SL is one of a slowly progressive, painless knee swelling that may last for decades. In a few patients, like ours, there have been intermittent exacerbations secondary to large effusions, which can cause pain and a decrease in range of motion<sup>2,5,6</sup>. Intermittent trapping of the fatty masses between moving joint surfaces likely produces these effusions. Rapid reaccumulation of the knee effusions in our patient was controlled with intraarticular steroids. Synovectomy — open<sup>3,7</sup> or arthroscopic<sup>8</sup> — is the suggested curative treatment for SL.

Most patients do not have an associated musculoskeletal disorder, although osteoarthritis (OA)<sup>1,6,9,10</sup>, rheumatoid arthritis (RA)<sup>11</sup>, gout<sup>12</sup>, psoriatic arthritis<sup>2,13</sup>, sarcoidosis<sup>14</sup>, and joint trauma<sup>12</sup> have been described. Bone erosions have occasionally been noted in knee MRI of patients with SL,

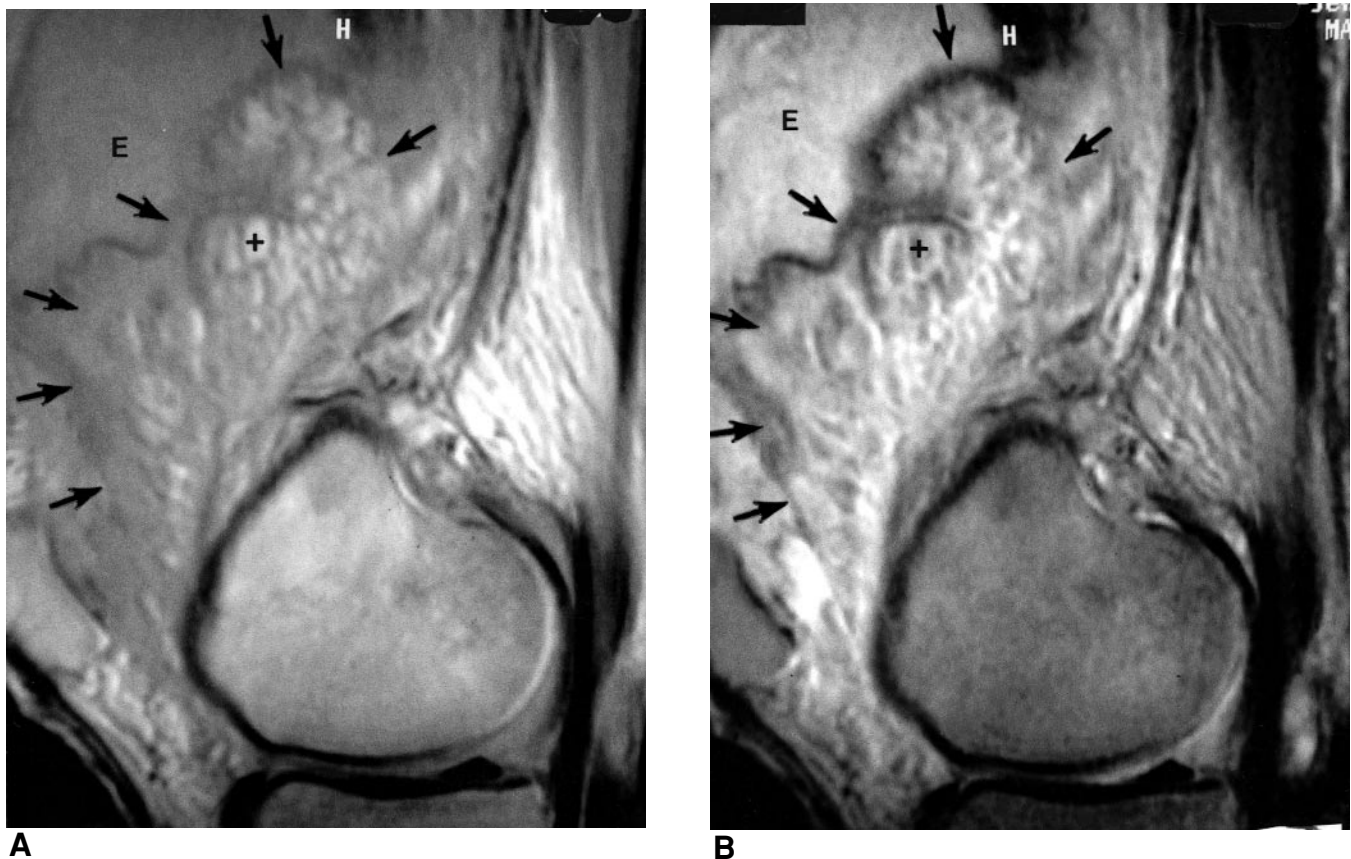


Figure 1. Sagittal images through the medial femoral condyle. A. T1 weighted image shows a large joint effusion (E) and lobulated (single lobule marked “+”) fat containing mass within the inferior portion of the suprapatellar bursa (outlined by arrows). B. Corresponding fat saturated T2 weighted image again shows the joint effusion (E), now higher in signal intensity. The high signal components observed on the T1 weighted image are now seen as low signal lobulations (marked “+”) on the fat saturated image, indicating that they are fat.

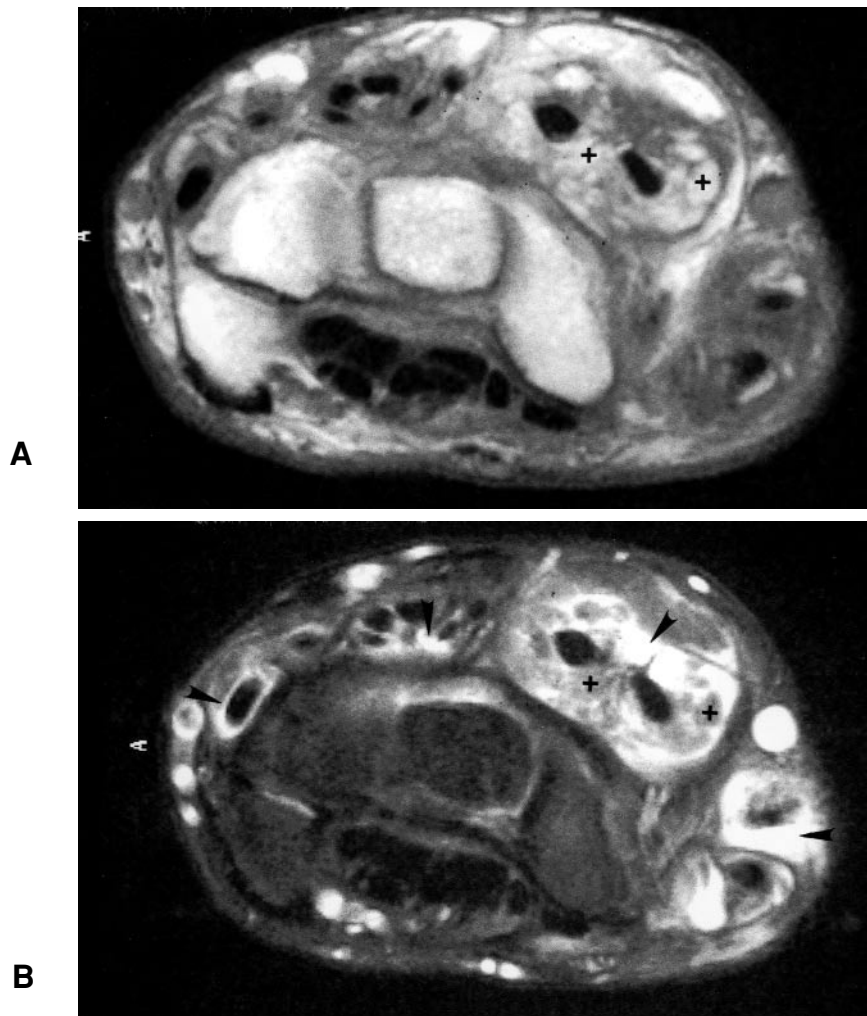


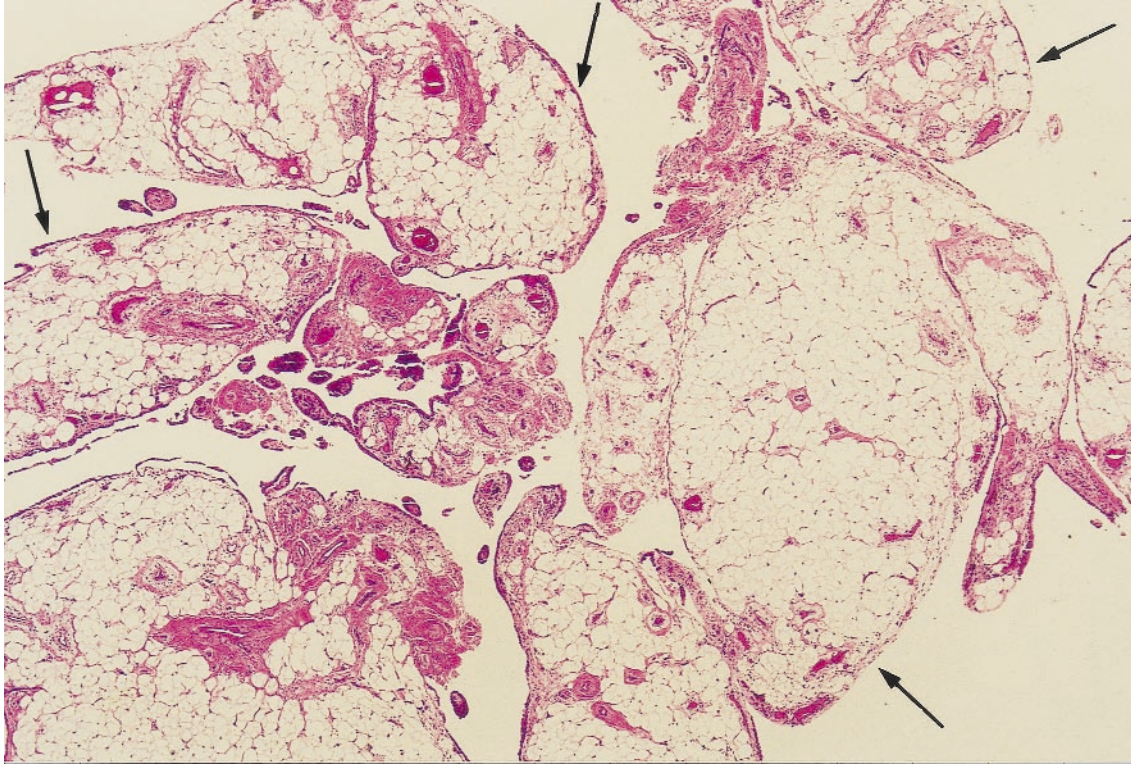
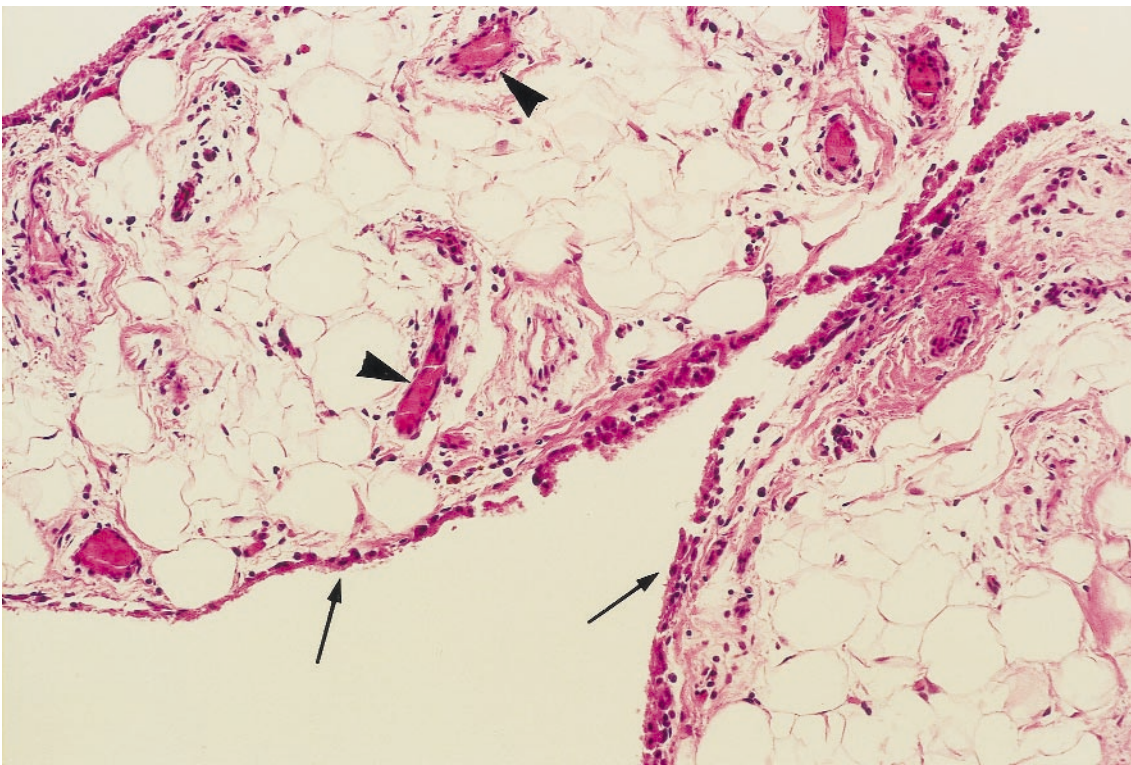
Figure 2. Axial images through the proximal carpal row. A. T1 weighted image shows fatty lobulations (marked “+”) within the synovial sheath of the 2nd and 3rd dorsal carpal compartments. B. Corresponding fat saturated T2 weighted image shows that the high signal components seen on the T1 weighted image (marked “+”) are now low in signal, again indicating their fatty content. Fluid is also visible within the tendon sheaths of all dorsal compartments (arrowheads).

even in the absence of a history of an inflammatory arthritis<sup>9</sup>. Some authors have speculated that SL might also contribute to the development of OA<sup>1,9</sup>. Although the severity of OA seems to correlate with the duration of the disease<sup>1,9</sup>, it is unclear whether the co-occurrence of OA and other arthritic conditions is more than coincidental.

The characteristic MRI features of SL include (1) an intraarticular mass containing fatty lobules surrounded by fluid, (2) a large joint effusion, (3) absence of nonfat soft tissue component and hemosiderin, and (4) possible chemical shift artifacts at the fat-fluid interface (dark-light signal lines at the edge of the fat adjacent to joint fluid). Pigmented villonodular synovitis (PVNS), atypical synovitis of RA, and synovial chondromatosis may resemble SL, but can usually be differentiated by MRI<sup>5,9</sup>, since none show fat lobules. The RA and PVNS masses are of soft tissue signal intensity, low on T1 images and intermediate to high on T2

images. The signal intensity of PVNS (contains hemosiderin) is very low on both T1 and T2 images, and that of synovial chondromatosis (appearing like cartilage) low on T1 and high on T2 images. The chondromatous bodies may contain fat within the center as they enlarge and may have rim-like calcifications. These calcifications have low signal intensity on all sequences, and are visible on plain radiographs (osteochondromatosis). The prefemoral fat pad seen in normal knees may be lobulated occasionally, but does not form a frond-like mass protruding into the knee, and is smooth and uniform with no fluid tracking into it.

Our patient has a second rare condition, congenital short bowel syndrome, of which only 28 cases have been described (reviewed by Schalamon, *et al* in 1998<sup>15</sup>). It usually results in death in early childhood and the longest survivor was reported to have lived for more than 7 years. Our patient is doing well at age 35 with a reasonable quality

**A****B**

*Figure 3.* Synovial biopsy of the wrist. A. Low magnification ( $\times 10$ ): Villi or nodules (arrows) of unremarkable mature adipose tissue. B. High magnification ( $\times 40$ ): The synovial lining consists mostly of a single layer of cells (arrows), but in some areas there is mild hyperplasia in association with a chronic inflammatory infiltrate. The engorged blood vessels are prominent (arrowheads).

Table 1. Synovial lipomatosis affecting locations other than the knee.

Author	Age/Sex	Location	Duration of Symptoms, yrs	Knee Involvement	Associated Diseases	Biopsy
Napolitano <sup>4</sup>	47 F	Wrist	0.5	–	–	+
Gaede <sup>16</sup>	14 M	Knee, ankle	0.5	+	–	+
Weston <sup>11*</sup>	33 F	Subdeltoid bursa	NA	–	RA	+
Noel <sup>18**</sup>	29 M	Hip	3	–	–	+
Hubscher <sup>7</sup>	26 M	Hip	1	–	Mild OA	+
Dawson <sup>17</sup>	65 M	Subdeltoid bursa	1.5	–	–	+
Laorr <sup>10</sup>	90 F	Subdeltoid bursa	NA	–	OA	–
Martin <sup>5</sup>	NA	Hips	NA	–	–	NA
Current report	35 M	Wrists, hands, knees	> 20	+	Mild OA	+

\* Although the author did not use the term “lipoma arborescens,” the lesions described in subdeltoid bursa were felt to be SL by others<sup>17</sup>, and the elbow lesions described by the same author are synovial lipomas. \*\* The hip lesion described in this report seems to be a solitary synovial lipoma rather than SL, as pointed out by Hubscher *et al*. NA: Not noted in the report.

of life. We speculate that aberrant handling of fat substrates by his short bowel may be etiologically related to synovial lipomatosis. However, we found no reports of association between short bowel (congenital or acquired) syndrome and SL.

The patient described here had several unusual features of synovial lipomatosis including the most extensive joint involvement ever reported, superimposed acute knee effusions responsive to steroid injections, and an association with congenital short bowel syndrome.

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