

Atypical, polyarticular lipoma arborescens in a child

Robert Semnic^{1,2}, Radoje Simić^{3,4}, Slaviša Djuričić^{3,5}, Oto Adjić¹, Filip Vanhoenacker^{6,7,8}

¹Vojvodina Oncology Institute, Sremska Kamenica, Serbia;

²University of Novi Sad, Medical Faculty, Novi Sad, Serbia;

³Mother and Child Health Care Institute of Serbia "Dr Vukan Čupić", Belgrade, Serbia;

⁴University of Belgrade, School of Medicine, Belgrade, Serbia;

⁵University of Banjaluka, Faculty of Medicine, Banja Luka, Republic of Srpska, Bosnia and Herzegovina;

⁶AZ Sint-Maarten Duffel-Mechelen, Department of Radiology, Mechelen, Belgium;

⁷Antwerp University Hospital, Department of Radiology, Edegem, Belgium;

⁸University of Ghent, Faculty of Medicine and Health Sciences, Ghent, Belgium

SUMMARY

Introduction Lipoma arborescens is a rare, tumor-like lesion commonly involving synovial joints and less commonly bursae and synovial tendon sheaths.

Case Outline We report a case of a 12-year-old boy with symmetric involvement of the bicipitoradial bursae, synovial sheaths of extensor compartments of both hands and medial ankles. The diagnosis of polyarticular lipoma arborescens was proposed on magnetic resonance (MR) imaging and this diagnosis was histologically proven after biopsy of the bursae and later by open surgery of the synovial sheath of the right ankle tendons. Literature search was performed and twelve cases with polyarticular involvement were analyzed. Lipoma arborescens commonly involves suprapatellar recess of the knee and very rarely other joints or bursae. Histological analysis revealed an accompanying non-necrotizing granulomatous synovial inflammation.

Conclusion Polyarticular lipoma arborescens is a rare entity and symmetrical involvement of the joints other than the knees is exceedingly rare. MR imaging plays a significant role in the diagnostic protocol, and the characteristic fatty signal on MR imaging is highly suggestive of lipoma arborescens.

Keywords: lipoma arborescens; synovium; tendon; MR imaging

INTRODUCTION

Lipoma arborescens (LA) is a rare, tumor-like lesion involving the synovial joints and less commonly bursae and synovial tendon sheaths. The suprapatellar recess of the knee is the most frequent location followed by the shoulder, elbow, hip, wrist, and hand [1–4]. Only twelve cases of multifocal polyarticular LA (not including bilateral knee involvement) have been reported in the English language literature [3–11]. We present a brief discussion on the polyarticular LA and the role of magnetic resonance (MR) imaging in the diagnostic algorithm of this rather infrequent tumor-like lesion.

CASE REPORT

A 12-year-old boy presented with a four-year history of gradual-onset bilateral swelling of the bicipitoradial bursae, extensor compartments of hands and wrist, and ankles, involving flexor hallucis longus, flexor digitorum longus, and tibialis posterior tendons, bicipitoradial bursae and extensor compartments of the hand (Figure 1).

There was a mild pain in the wrists but no morning stiffness, and no history of trauma. Laboratory investigations were all within the normal ranges. Rheumatological examination

revealed no abnormalities. MR imaging of both arms and ankles was performed (Figure 2, A–D).

A biopsy of the right ankle was performed and the histological changes were consistent with LA associated with a mild chronic synovial inflammation and several non-necrotizing sarcoid granulomas containing rare multinucleated giant cells (Figure 3, A–B). At follow-up, the lesions remained unchanged and there was neither pain nor any functional disability. Six months later, a partial synovectomy of the right ankle was attempted but on the opening of the tibialis posterior tendon sheath, fat tissue unusually firmly adherent to the tendon was found (Figure 4) and the surgeon decided against further radical excision.

A third surgical resection of the wrist LA on the right side was performed one year later, with successful outcome.

DISCUSSION

English language literature search was performed using the subject term "Lipoma Arborescens" in the PubMed database in order to identify articles published from 1950 through July 2015. Among 137 articles yielded by the search criteria, all articles describing monoarticular involvement as well as 16 articles reporting bilateral knee involvement were excluded

Correspondence to:

Robert SEMNIC
Vojvodina Oncology Institute
Put dr Goldmana 4
21204 Sremska Kamenica
Serbia
robert.semnic@mf.uns.ac.rs



Figure 1. Photograph of the elbows, hands, and ankles showing bilateral soft-tissue swelling

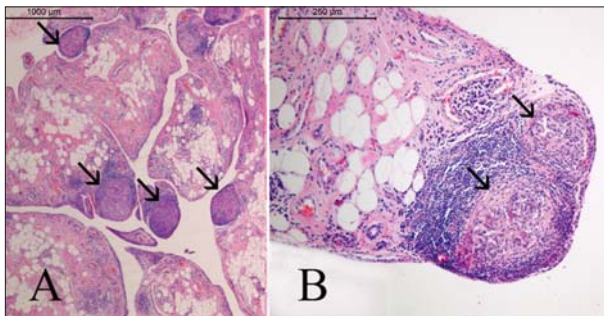


Figure 3. Photomicrographs of the specimen of the excised synovium shows elongated synovial folds containing small lobules of mature fat tissue typical for LA; several sarcoid granulomas and a mild lymphoplasmacytic infiltration are easily visible in the synovial folds (A – hematoxylin-eosin, $\times 25$); the top of the fold contains two non-necrotizing epithelioid cell granulomas, partially surrounded by dense lymphocytic infiltration; there are also several foci of discrete, perivascular lymphoplasmacytic infiltration (B – hematoxylin-eosin, $\times 13.75$)

because they were beyond the scope of this paper. Data of 12 patients reported in nine published articles [3–11] and the present report were analyzed (Table 1).

LA is not a true tumor but rather a villous synovial hyperplastic process that affects mostly men aged 40–60 years, and manifests itself as a slow-growing monoarticular painless swelling, usually in the suprapatellar pouch of the knee. Polyarticular involvement is rare and most frequently affects the knee, whereas bilateral involvement of other joints or a combination of the knee and another joint is far less common (Table 1). Our patient had several atypical features: 1) early age of disease onset; 2) involvement of joints that are otherwise infrequently affected by

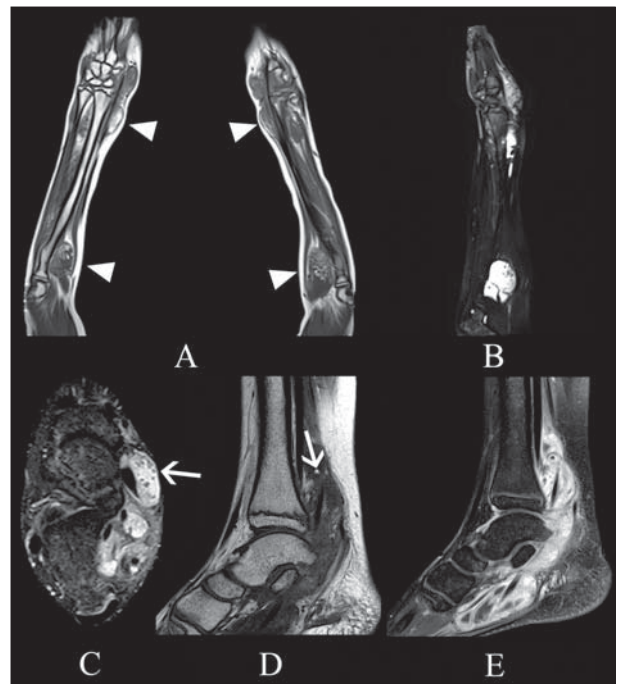


Figure 2. Coronal T1-weighted MR image of forearms shows fatty tumor-like lesions within bicipitoradial bursae and both wrists (A, arrowheads); sagittal fat saturated T2-weighted image – fluid-filled bicipitoradial bursa and frond-like fatty proliferations of the synovium; mild distension of the synovial sheaths of the extensor compartment tendons of the wrist (B); axial STIR image of the right ankle shows fluid within tendon sheaths of the FHL, FDL, and TP, with intralesional foci of fat (C, arrow; D, arrow); fat-saturated contrast enhanced T1-weighted sequence of the right ankle shows thickened synovium with significant contrast enhancement (E), consistent with tenosynovitis

FHL – flexor hallucis longus; FDL – flexor digitorum longus; TP – tibialis posterior



Figure 4. An intraoperative photograph of the right ankle with visible lipomatous mass after opening of the TP tendon sheath

LA, in a combination that has not been reported to date; 3) symmetrical swelling without significant restriction of movement.

Etiology of LA is unknown. However, the majority of cases are primary, although some may represent a secondary reaction to chronic rheumatoid arthritis [12], psoriatic arthritis [13], sarcoidosis [14], joint trauma [15], and the proposed hypothesis is that subsynovial fatty infiltration may reflect a reaction to chronic inflammation [16].

Howe and Wenger [11] proposed a primary form of LA in younger patients without a detectable cause of chronic

Table 1. Multifocal LA, excluding bilateral knee involvement*

Authors	Location	Onset age/sex	Bilateral involvement	Duration of symptoms	Associated diseases	Biopsy (+), synovectomy or excision
Bejia et al. [3]	Knee, hip	24/M	+	6 months	-	+
Siva et al. [4]	Wrist, hand, knee	35/M	+	20 years	Congenital short bowel syndrome	+
Gaede [5]	Knee, ankle	14/M	-	6 months	-	+
Dinauer et al. [6]	Bicipitoradial bursa	37/M	+	1.5 year	-	Synovectomy
Silva et al. [7]	Knee, ankle	45/M	-	9 years	Knee trauma	Excision of the ankle LA
Santiago et al. [8]	Knee, hip	29/F	+	12 years	Rheumatoid arthritis	+
Pandey and Alkhulaifi [9]	Subdeltoid bursa	57/M	+	3 years	Prior shoulder trauma, osteoarthritis	+
Martin et al. [10]**	Hip (iliopsoas bursa)	N/A	+	Not specified	Osteoarthritis, AVN of both femoral heads	Not specified
Howe et al. [11]** (3 patients)	- Knee and ankle - Wrist extensor compartment - Bilateral knee and bicipitoradial bursa	N/A	-	Not specified	Not specified	Not specified
		N/A	+			
		N/A	-			
Current report	- Bicipitoradial bursa, wrist, ankle	10/M	+	5 years		Biopsy, 3 excisions later on

* The table has been used and further modified with permission from Siva et al [4].

** Information about onset age, gender, symptom duration and surgical procedures from articles by Martin et al. [10] and Howe et al. [11] have not been included in the table due to lack of necessary data.

M – male; F – female; N/A – not applicable; AVN – avascular necrosis

joint inflammation [10]. This form can involve more multiple joints [3], tendon sheaths [17], and bursae [6, 9, 10, 11]. Other cases of LA with polyarticular involvement but without an identifiable cause are found in the literature [3].

Two groups of fatty lesions that affect joints, bursae, and tendon sheaths are described as follows: a solid fatty tumor (a synovial lipoma) and a “lipoma-like” lesion in the form of diffuse, hypertrophic synovial villi distended with mature fatty tissue [18]. The second form is present in our case.

Mild lymphoplasmacytic synovial infiltration with non-necrotizing sarcoid granulomas in our patient may suggest a previous chronic synovial inflammation as an expression of arthritis as the underlying joint pathology. A primary inflammatory synovial process was also identified in all five cases reported by Martin et al. [10]. Arthritis and periartthritis as joint manifestations of sarcoidosis may occur in 14–38% of patients with sarcoidosis. They may be a presenting feature or appear later in the course of the disease when multiple large joints are involved more frequently [19]. The histology in our patient was consistent with a chronic synovial granulomatous inflammation seen in sarcoidosis. However, we did not register any clinical, radiological or laboratory features of sarcoidosis. In addition, we found neither anamnestic, clinical and laboratory

diagnostic elements of tuberculosis and fungal infection, nor trauma with potential impaction of foreign bodies in our patient – infections and disorders in which granulomas could appear along with chronic synovial inflammation [20]. Only in one 24-year-old patient with rheumatoid arthritis LA-associated scattered multinucleated giant cells were found in the accompanying lymphoplasmacytic synovial infiltration [12]. In our patient, we found for the first time entirely formed sarcoid granulomas in the synovial folds. Pathological entities with granulomas are classified into infections, vasculitis, immunological aberrations, leukocyte oxidase deficiency, hypersensitivity, chemicals, and neoplasia. However, histopathologists can detect granulomas in many different pathological disorders that are outside of this classification and that probably indicate a good defense and a satisfactory outcome against an unknown antigenic aggression [20]. We have no plausible explanation for sarcoid like granulomas occurring in the setting of LA.

In general, open synovectomy is suggested as curative treatment [10].

In conclusion, polyarticular LA is a rare condition and MR imaging has a pivotal role in the correct diagnosis, and more widespread use of MR imaging in the examination of joints will probably reveal more cases of polyarticular LA in the future.

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Атипични, полиартикуларни *lipoma arborescens* код детета

Роберт Семниц^{1,2}, Радоје Симић^{3,4}, Славиша Ђуричић^{3,5}, Ото Ађић¹, Филип Ванхунакер^{6,7,8}

¹Институт за онкологију Војводине, Сремска Каменица, Србија;

²Универзитет у Новом Саду, Медицински факултет, Нови Сад, Србија;

³Институт за здравствену заштиту мајке и детета Србије „Др Вукан Чупић“, Београд, Србија;

⁴Универзитет у Београду, Медицински факултет, Београд, Србија;

⁵Универзитет у Бањој Луци, Медицински факултет, Бања Лука, Босна и Херцеговина;

⁶AZ Sint-Maarten Duffel-Mechelen, Одељење за радиологију, Мехелен, Белгија;

⁷Универзитетска болница у Антверпену, Одељење за радиологију, Едегем, Белгија;

⁸Универзитет у Генту, Факултет медицинских и здравствених наука, Гент, Белгија

КРАТАК САДРЖАЈ

Увод *Lipoma arborescens* представља ретку, тумору сличну лезију која захвата синовијалне зглобове и ређе бурзе и овојнице тетива.

Приказ болесника У раду се приказује случај 12-годишњег дечака са симетричним захватањем биципиторадијалних бурзи, синовијалних омотача екстензора обе шаке и медијалних аспеката оба скочна зглоба. Након начињеног прегледа магнетном резонанцом (МР) постављена је сумња на полиартикуларни *lipoma arborescens*, а дијагноза је доказана хистолошки након биопсије бурзе и касније након отворене хирургије синовијалних овојница десног скочног зглоба. Хистолошка анализа је додатно утврдила постојање

не-некротизирајуће грануломатозне синовијалне упале. Извршена је анализа дванаест случајева из литературе са полиартикуларним *lipoma arborescens*.

Закључак *Lipoma arborescens* је ретка, тумору слична промена која захвата супрапателарни рецесус колена и врло ретко друге зглобове и бурзе. Полиартикуларни *lipoma arborescens* је редак ентитет, а симетрично захватање зглобова је изузетно ретко. МР игра врло важну улогу у дијагностичком протоколу код оваквих пацијената, а карактеристичан сигнал масне компоненте тумора сугерише постојање *lipoma arborescens* са високом сигурношћу.

Кључне речи: *lipoma arborescens*; синовијум; тетиве; магнетна резонанца