Primary synovial osteochondromatosis of the first interphalangeal joint of the foot: A case report

Keywords: Primary synovial chondromatosis; Osteochondromatosis; Synovium

Primary synovial osteochondromatosis (PSO) is characterized by cartilaginous metaplasia of the synovium causing sub-synovial production of multiple hyaline cartilage nodules [1]. It has long been thought of as chondroid metaplasia of the joint synovium, although recent cytological studies have shown that it is in fact a benign neoplastic process involving clonal chromosomal abnormalities [2], secretion of fibroblast growth factors and their receptors [2], dysregulation of progenitor mesenchymal cell differentiation [3], and increased type II collagen synthesis [4].

It is a rare disease (1 per 100,000 people) [5], and is benign but frequently recurrent [6].

Patients usually complain of mechanical pain (85 of 100% cases), joint swelling (42%–58%) and restricted joint mobility (38%–55%) [1].

We describe a novel case of PSO of the interphalangeal joint of the first digit of the right foot in a 46-year-old male patient.

Observation

A 46-year-old man, an army parachutist, described intermittent pain of his right hallux, inflammatory in timing and present for around 10 years, which was hindering him in his job. Clinical examination revealed pain on palpation with slight swelling of the interphalangeal joint. No erythema or skin rash was seen over the remainder of his right foot and he had no active or passive limitation in joint movements. Pulses were present and neurological examination revealed no abnormalities.

Standard right foot radiographies showed multiple oval radiopaque ossified lesions peripheral to the interphalangeal joint with swelling of the adjacent soft tissues. The joint lines were preserved and he had no bony erosions (Fig. 1).

He gave no clinical history of trauma, infection or rheumatological disease.

All of his laboratory investigations were normal and in particular, he had no acute phase reaction.

The diagnosis of PSO was made and further investigations were performed before surgery.

CT showed multiple free millimeter-sized osteochondromas of bone density within and around the joint, with no bony erosions (Fig. 2).
MRI showed osteochondromas, which were hypo-intense on T1- and T2-weighted images and peripheral uptake, delineating the joint cavity on T1-weighted fat saturation gadolinium enhanced images. There were no image abnormalities within the bone (Fig. 3).

Ultrasound confirmed mobile sloping chondromas (Fig. 4).

Surgery involved open synovectomy after excising all of the chondromas (Fig. 5).

Histological analysis confirmed the diagnosis of PSO, showing multiple hyaline cartilage lobules beneath the synovial membrane.

His postoperative course was uncomplicated.

Discussion

PSO is classically mono-articular, affecting the large joints, the knee in 50% of the cases [7].

To our knowledge, only one case of interphalangeal PSO of the hallux has been reported to date in the literature [8]. Four other cases have been reported but all involved the metatarsophalangeal hallux joint [9–12].

Milgram described 3 successive phases of the disease [10]:

- an active phase with sub-synovial cartilage proliferation but without intra-articular release of chondroma;
- a transitional phase with active or inactive sub-synovial cartilaginous nodules with free, occasionally calcified, intra-articular chondromas;
- a late inactive phase (40% of the cases) with quiescent or slightly inflamed synovium and multiple free intra-articular osteochondromas.

With respect to the MRI findings in synovial chondromatosis, the sub-synovial cartilaginous proliferation results in tissue surrounding the joint cavity which is iso- or hypo-intense on T1-weighted images and hyper-intense on T2-weighted images. Small focal hyper-intense areas on T1-weighted images are occasionally seen within the synovium [13]. These represent either proliferation of macrophages with lipid laden cytoplasm (foamy histiocytes) or fatty bone marrow if there is not sufficient surrounding cortical bone to be detected [13].

The osteochondromas are hypo- or hyper-intense on T1- and T2-weighted images depending on their calcium and fatty bone marrow content delineated by a fine hypo-intense line. They do not enhance with contrast. On T1-weighted images after gadolinium enhancement, the inflamed synovium enhances clearly and moulds round the following bodies, which are unchanged in appearance [14,15]. In our case, the contrast enhancement represented reactive synovitis, as the spatial resolution of the investigation cannot differentiate the synovial from sub-synovial tissue [1]. Occasionally, bony edema reactive to the pressure of the foreign bodies can be seen.

Milgram recommends simple excision of the foreign bodies in the late inactive stages of the disease although adding synovectomy and foreign body removal if the disease is active or transitional [8].

The recurrence rate varies depending on the series from 3% to 23% of the cases [5], and is usually due to incomplete resection. Its incidence depends partly on the joint and on the Milgram histological stage.
Malignant transformation into synovial chondrosarcoma is a rare disease [1,5,17–19]. Two studies, however, have shown a malignant transformation rate of around 5% [13–16]. This is classically seen in longstanding PSO after several local recurrences and involves the large joints.

Conclusion

Involvement of the interphalangeal joint of the first digit of the foot is a very rare sight for primary synovial osteochondromatosis. This diagnosis should be considered in refractory mono-articular synovitis even if the site is uncharacteristic.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


N. Brucher b,*, M. Faruch-Bilfeld a, F. Molinier b, A. Brouchet-Gomez b, F. Lapegue a, N. Sans a

a Department of radiology, Toulouse-Purpan University Hospitals, place du Docteur-Baylac, TSA 40031, 31059 Toulouse cedex 9, France

b Department of Orthopedic Surgery, Toulouse-Rangueil University Hospitals, 1, avenue du Pr. Jean-Poulhès, TSA 50032, 31059 Toulouse cedex, France

c Cellular pathology laboratory, Toulouse-Rangueil University Hospitals, 1, avenue du Pr. Jean-Poulhès, TSA 50032, 31059 Toulouse cedex, France

* Corresponding author.

E-mail address: bruchernicolas@gmail.com (N. Brucher)