

A case of recurrent hyperostotic macrodactyly

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Macrodactyly and its variation hyperostotic macrodactyly are some of the rarest deformities encountered by foot and ankle specialists. Changing the natural aesthetic shape of the foot, limiting the functionality of the lower extremity, and causing pain are some ways that this condition can affect patients' everyday life and mental health. This study presents a case of recurrent hyperostotic macrodactyly that was managed with surgical intervention in order to debulk the soft tissue and excise excess osseous elements with successful results.

Keywords: congenital deformity, forefoot, lower extremity, foot and ankle surgery

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Macrodactyly is a non-hereditary and congenital deformation pathology of the upper and lower extremities which can be bilateral or unilateral [1-5]. In this deformity both osseous and soft tissue components of the digit can be enlarged in size which can cause functional and esthetic problems for the patient. Two types of this condition that were described by Barsky are static, in which deformity is present at birth and increases in size proportionally to other digits, and progressive, in which a digit grows disproportionately to other digits and is most commonly encountered in the lower extremity [1].

Macrodactyly is believed to be originally described by Von Klein in 1824 in the upper extremity and later in 1925 by Feriz in the lower extremity [6-8]. This disorder is thought to be a rare condition and because of that there is no accurate estimate of the prevalence of this disorder. According to some estimates, macrodactyly accounts for 0.9% of all congenital deformities and it is more prevalent in males [9, 10]. The etiology of macrodactyly has been debated over the years, but no clear conclusion has emerged yet. Some cases of

macrodactyly present in patients with other disorders such as Proteus syndrome, Neurofibromatosis type 1, Klippel–Trenaunay syndrome, lymphangioma and fibrous dysplasia [8,11,12]. However, in many of the case reports published regarding macrodactyly, this condition is observed as an isolated condition with no other associated disorders [2,3,9,13,14].

Throughout the years, different terms such as macrodystrophia lipomatosa progresia, macrodystrophia lipomatosa, megalodactyly, and localized gigantism have been used in the literature to describe macrodactyly or other variations of this deformity [7]. A unique and less discussed type of progressive macrodactyly is hyperostotic macrodactyly which usually has a later onset than typical macrodactyly and is associated with osteo-cartilaginous mass formation in peri-articular areas of the upper and lower extremity [15].

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Figure 1 Preoperative medial oblique X-ray image (on the left) and clinical photo (on the right), prior to second surgery.



Figure 2 Postoperative medial oblique X-ray image, following the second surgery.

The rare presentation of hyperostotic macrodactyly and a lack of literature regarding this topic have provided practitioners with no clear guidelines regarding the management of this disorder. This case report presents a case of a mild recurrent

hyperostotic macrodactyly in a patient with previous surgical interventions to address this deformity.

Case Report

A 53-year-old female with no past medical history other than asthma presented to the clinic with a painful recurrent mass on her left hallux. She related that she has had two prior surgeries. The patient reported that she was originally seen regarding this problem when she was 14 years old and was diagnosed with localized gigantism. She had a surgery at that time to fix her deformity which had satisfying results and resolved her problem for about 35 years.

The second surgery was performed four years ago when the patient was seen by another podiatrist regarding this problem. She had noticed an increase in size of her left hallux and denied any trauma to the area. She had pain both on the plantar and medial side of her left hallux and first metatarsal head with noticeable bony prominences. She changed her shoe gear, using slippers or open sandals to accommodate the prominences (Figure 1). In the second surgery, osteophytes from the left first metatarsophalangeal joint and hallux were excised and a soft tissue mass from the plantar aspect of the left hallux was removed as well (Figure 2). During the postoperative period, the patient stubbed her hallux on a heavy plastic bin which was very painful for her, however no fractures were noted, and the rest of her postoperative course was uneventful.

The patient presented to our clinic two years ago due to noticing the recurrence of her deformity after the second surgery. Physical exam revealed approximately a 4 cm x 3 cm firm soft tissue mass overlying the left first tarsometatarsal joint. There was also tenderness with palpation of a prominent exostosis along the medial aspect of the left hallux Interphalangeal joint. Joint motion at first metatarsophalangeal and tarsometatarsal joints were severely limited as well, but not painful (Figure 3). Magnetic resonance imaging report indicated an ovoid, subcutaneous lipoma measuring 5.2 x 2.8 x 1.1 cm. Mild to severe arthritis of the first tarsometatarsal and metatarsophalangeal joints with ossified bodies in addition to fatty infiltration of abductor and flexor hallucis muscles was also noted.



Figure 3 Preoperative clinical photos (in the left and the middle) and dorsoplantar X-ray image (on the right), prior to the third surgery.

After discussing the possible adverse effects, benefits and alternative therapies to the surgery with the patient, the patient wished to proceed with exostectomy and removal of the soft tissue mass. During the third surgery, osseous masses from the left hallux were removed and the soft tissue mass from the dorsal and medial aspect of first metatarsal of the left foot were excised and both specimens were submitted for pathology evaluation (Figure 4). The osseous masses were clinically equivalent with osteophytes measuring 0.8x0.6x0.5cm and 1.2x0.8.0.5cm.



Figure 4 Intraoperative image of the excised fibro-fatty mass, during the third surgery.



Figure 5 Postoperative clinical photo, following the third surgery.

The soft tissue mass revealed mature adipose tissue with features of a lipoma measuring 4.5x2.5x1.3cm. The patient was kept non-weight bearing in a splint for 2 weeks and then she was allowed to weight-bear as tolerated for 2 weeks in a surgical boot. Postoperative course was uneventful, and the patient made good progress, had no complaints, and was satisfied with the results more than 18 months after the third surgery (Figure 5).

Discussion

Hyperostotic macrodactyly is a distinctive type of macrodactyly in which massive osteo cartilaginous deposits are observed around the joints [2,15]. Early in the formation process of these osteo-cartilaginous bodies around the joints, they are mostly cartilaginous and later they are substituted with osseous elements which leads to motion restriction across the affected joint [2]. As a progressive macrodactyly, it is not uncommon to observe fatty growths or lipomas and fatty infiltrations in this condition. In many reported cases this fatty hypertrophy and infiltration can be observed in both plantar and dorsal aspect of the foot [3,14]. The case presented in this article demonstrated both osseous and fatty enlargement across the first ray of the patient's left foot.

The main goals of treatment for macrodactyly should be providing the patients with a pain free, functional foot that can fit into their shoes [4]. Unfortunately, there are very few reported cases of hyperostotic macrodactyly in the foot, however it appears that the removal and debulking of the excess fibro-fatty tissue and osseous bodies is the best method of management of this disorder which was the way the patient in this case was treated [2,9,13]. In a case report by Katz, the patient presented with lipomas across his right foot and ankle and osteo-cartilaginous growths across his fourth and fifth toes and metatarsals [9]. An surgery was performed on the patient in which the lipomas and osteo-cartilaginous bodies were excised and the fifth toe and half of the fifth metatarsal were resected. The patient had excellent results after the surgery. In another case by Matsuzaki, et al., the patient was suffering from painful and limiting osteo-cartilaginous masses around his left first metatarsal head and ankle [2]. Patient had a previous left second toe and hallux amputation surgery to address his macrodactyly. After removal of these osseous bodies from the patient's left foot, his pain was relieved and the motion across his ankle joint increased and no recurrence was reported.

One curious aspect of the case presented in this article that requires attention is the recurrence of osteo-cartilaginous bodies almost two years after their resection in the second operation. This seems to be too early for a recurrence to happen considering that

it took almost 35 years after the first surgery for her to have problems with her left foot again. A possible explanation for this can be the traumatic event to the area in the postoperative course in which the patient stubbed her big toe straight into a heavy plastic bin. Some studies have suggested that trauma can be the trigger for the osteo-cartilaginous hypertrophy observed in hyperostotic macrodactyly deformity which would explain the recurrence in this case [2,15].

In conclusion, hyperostotic macrodactyly is a rare progressive form of macrodactyly in which massive and limiting periarticular osteo-cartilaginous bodies in addition to fatty tissue hypertrophy can form. Surgical intervention to remove these osseous and fatty masses in cases that they cause pain and functional disability due to blocking joints appears to be the best method to treat this condition. It is also very important to educate patients to avoid trauma to the areas affected by hyperostotic macrodactyly since trauma appears to be one of the causes of this disorder or its recurrence.

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