

Bilateral digital xanthomatosis: a case report

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Abstract: Triki R: Trikiramy0816@gmail.com

Xanthomas are physical manifestation of hyperlipidemia. These lesions can occur in multiple regions of the body. Their location in the hand is rare and usually well tolerated. Their excision can be necessary either for functional or cosmetic reasons. Their presence can be a sign for future coronary diseases. Our case is a man whose both hands were affected.

Keywords

Xanthomatosis ; Hand ; hyperlipidemia

Introduction:

Xanthomas are non-neoplastic tumors that represent a physical manifestation of hyperlipidemia. Particularly, tendon xanthomas often appear as a clinical manifestation of familial hypercholesterolemia [1]. It can precede other signs or complications of hyperlipidemia, such as coronary atherosclerosis. Therefore, these lesions shouldn't be neglected. Xanthomas often affect the Achille tendon and rarely the hands. It is known that they can affect flexor or extensor tendons. Around 4 articles were reported in the English literature focusing on xanthomas affecting the extensor tendons. We report here a case of a patient whose hands were both affected with a review of the literature.

Case report

Our case is a 42-year-old man, right handed, manual worker, who has hypertension, familial hypercholesterolemia type 2 treated with

plasmaphereses since the age of 12 years-old. He had a triple coronary bypass at the age of 27. His parents are not related.

Since 3 years ago, he had multiple and bilateral small tumors on the dorsum of his both hands. These tumors were painless and soft. In his right hand, these lesions were located regarding the 3rd and 4th metacarpophalangeal joint and the 2nd phalange of the middle finger. In his left hand, these lesions were located regarding the 3rd and 4th metacarpophalangeal joints (**Figure 1**). His range of motion were slightly affected. These tumors were growing slowly during these 3 years. There was no arcus cornea. His X rays were normal.

The surgical biopsy of these tumors was performed under local anesthesia (**Figure 2**). The tumors had lipidic aspect. Their size was around 3 x 3 cm. These lesions were adherent to the extensor tendons.

At the last follow up of 6 months, there was no recurrence of these lesions. There was a good cosmetic and functional result, with a complete range of motion (**Figure 3**). The anatomopathology confirmed the diagnosis.



Figure 1: Clinical aspect before the surgery

Figure 2: Clinical aspect during the surgery



Discussion:

Cases in the literature about hand xanthomatosis are rare. To our knowledge, 4 articles were focusing on xanthomas in the extensor tendons in the English literature [1,2,3,4] (**Figure 4**). One case is reported to affect the range of motion [4].

Xanthomas are considered as reactive lesions, associated with familial hypercholesterolemia, which is an autosomal dominant, highly penetrant (90%), inherited condition caused primarily by a mutation in an LDL cholesterol receptor gene. During familial hypercholesterolemia, the heterozygous form is the most common [5].

Xanthomas are defined by the deposition of yellowish cholesterol rich material in tendons or other body parts [4]. They are subcutaneous and tendinous deposits of cholesterol within macrophages (foam cells). Histologically, they consist of clusters of foam cells containing cholesteryl ester, triglycerides and phospholipid [6]. The detection of xanthomas is critical to an early diagnosis of the disease, which is extremely important to be able to alter the course of the disease before the onset of coronary artery disease [4]. In our patient, he has already had coronary complications at an early age.

The prevalence of xanthomas rises after the age of 30 in people with familial hypercholesterolemia [5]. They are generally seen in the elbow, knee, hip, ankle and other surfaces of the extensor large joints, including the Achilles Tendon [6,7,8]. The tendons which are affected are usually weakened and tend to be more susceptible to injury [9,10]. Once they infiltrate the tendons, they are more likely to cause pain and stiffness of the joints.

These lesions arise from the tendon. Therefore, their removal can cause its disruption. Most of the time, the patient motive is cosmetic. In our patient, lesions became larger by the time, and thus less cosmetic. They also altered his range of motions.



Figure 3: Clinical aspect at the last follow-up

Xanthoma can mimic rheumatoid arthritis, like the case report of Dan Li [11]. More importantly, xanthomas are a visible sign of familial hypercholesterolemia. Patient with xanthomas are more likely to have early coronary complications. the presence of tendon xanthomatosis was correlated with an over-risk of 3.2 early cardiovascular complications: 2.3 and 4.5, respectively, in men and women [5]. Aside from excising the lesions, we have to keep in mind the importance of prevention of these complications.

Conclusion:

Xanthomas in the hand are exceptional and well tolerated most of the time. The surgical excision of these lesions is indicated when the extensor tendons are affected and interfering with the function of the hand. Cosmetic discomfort is evaluated according to the size of the lesions. Coronary and general complications should be kept in mind.

Article	Number of cases	Sex	Age	Side affected
J.R Doyle [1] (1988)	3	F M M	55 50 66	- Right -
G.T. Lin [2] (1999)	1	M	42	Right
D.J. Bozentka [3] (2001)	2	-	-	-
Huri G [4] (2013)	1	M	42	Bilateral
Our case	1	M	42	Bilateral

Figure 4: Comparison of the cases in the English literature

References:

- [1] J. R. Doyle, "Tendon xanthoma: a physical manifestation of hyperlipidemia," *Journal of Hand Surgery*, vol. 13, no. 2, pp. 238–241, 1988.
- [2] G. T. Lin, "Surgical excision of the tendon xanthoma in familial hypercholesterolemia—a case report," *The Kaohsiung Journal of Medical Sciences*, vol. 15, no. 7, pp. 441–446, 1999.
- [3] D. J. Bozentka and B. M. Katzman, "Two cases of surgically treated hand tendon xanthomas," *American Journal of Orthopedics*, vol. 30, no. 4, pp. 337–339, 2001.
- [4] Huri G, Joachim N. An unusual case of hand xanthomatosis. *Case Rep . Orthop* 2013; 2013: 183018.
- [5] Bento Da Costa A, Le Besnerais M, Cailleux-Talbot N, Armengol G, Levesque H, Marie I. Nodules involving hands and feet. *Rev Internal Medicine*. July 2015; 36(7):498-500.
- [6] Aljenedil S, Ruel I, Watters K, Genest J. Severe xanthomatosis in heterozygous familial hypercholesterolemia. *J Clin Lipidol*. July 2018; 12(4):872-7.
- [7] C. Mirzanli, C. Z. Esenyel, K. Ozturk, A. Baris, and Y. Imren, "Cerebrotendinous xanthomatosis presenting with bilateral achilles tendon xanthomata: a case report," *Journal of the American Podiatric Medical Association*, vol. 103, no. 2, pp. 152–155, Mar-Apr 2013.
- [8] E. K. Hassett, K. Kulig, and P. M. Colletti, "Xanthomatous tendinosis," *Journal of Orthopaedic and Sports Physical Therapy*, vol. 42, no. 4, article 379, 2012.
- [9] H. S. Kruth, "Lipid deposition in human tendon xanthoma," *American Journal of Pathology*, vol. 121, no. 2, pp. 311–315, 1985.
- [10] J. D. Lenn, "High cholesterol and tendon injury," *American Association of Orthopaedic Surgeons*, vol. 4, no. 7, 2010.
- [11] Li D, You L, Fan S, Tan L. Xanthomatosis in bilateral hands mimicking rheumatoid arthritis: A case report. *Medicine (Baltimore)*. Dec 2017; 96(51):e9399.