Case Report

Xanthoma of Tendoachilles: Management and Follow-Up

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Abstract
Xanthomas are rare pseudotumors of connective tissue, consisting of histiocytes with lipid deposits. Usually, it manifests as encapsulated lobular yellowish mass, often associated with familial hypercholesterolemia (FH). We present a case of xanthoma of tendoachilles just proximal to its insertion site, secondary to FH. The patient was a 28-year-old female presenting with a 2-year history of difficulty ambulating and complaints of swelling over the posterior aspect of her left lower leg just proximal to her heel. She had a deranged lipid profile level and was started on a lipid lowering agent for 3 weeks followed by total excision of the lesion with flexor hallucis longus tendon graft augmentation. The patient was able to bear full weight at 6 weeks and could stand on her toes by 4 months. No recurrences were noted at 1 year follow-up.

Introduction
Xanthomas are rare non-neoplastic lesions that are frequently found in the tendons and synovium [1]. They appear as hard and nodular structure which is covered by skin. The common sites are the tendoachilles (TA) and the patella tendon [2]. These are most commonly seen in the 3rd decade of life with female predominance (4:3) as compared with males [3]. Usually, tendinous xanthomatosis is found to be associated with hyperlipidemia. It occurs due to a defect of the LDL receptor and is associated with familial hypercholesterolemia (FH) [4]. Histologically tendinous xanthoma contains multinucleated giant cells, histiocytes, and hemosiderin deposits. Xanthoma of TA may lead to pain, loss of function, and shoe-related problems [5].
This lesion makes the tendon weak resulting in weakness of plantar flexion. There are no clear-cut guidelines about the management of such cases and most of the cases are managed conservatively in the literature. The graft used for reconstruction purposes was also different in the different study and there is no definite consensus. We present a case of xanthoma of left side TA just proximal to its insertion site, secondary to FH. It was managed with total excision of the diseased segment and reconstruction with autologous flexor hallucis longus (FHL) transfer.

**Case**

A 28-year-old female presented to the outpatient department with complaints of swelling over posterior and distal aspect of the left leg just above the heel for 2 years and pain for 2 months with difficulty on walking. It was insidious in onset without any history of trauma, gradually progressive and aggravated on walking and relieved on rest.

On examination, a firm swelling of size 6 × 3 × 2 cm on the TA tendon was present 2 cm proximal to the insertion site (Fig. 1a). It was globular in shape, the skin over the swelling was smooth with the normal surrounding skin. There was no local rise of temperature, the swelling was non-tender, non-reducible and non-translucent, and non-adherent to skin. The lesion was in the tendon and was moving along the movement of the tendon. In the left ankle, terminal dorsiflexion was painful and restricted by 10° in comparison to the opposite side (30°). The patient had an antalgic gait. There was no motor weakness or sensory loss of the bilateral leg.

X-ray of heel showed soft tissue shadow without the involvement of bone (Fig. 1b). Ultrasonography of the swelling showed a homogenous hypoechoic lesion inside the TA. The magnetic resonance imaging of the left ankle showed swelling (hyperintensity structure) of 6 × 3 × 2 cm, arising from TA (Fig. 1c) suggestive of tendon xanthoma (TX). After clinical and radiological examination, the provisional diagnosis was made as TX with differential diagnosis as cholesterol accumulation, tendonitis, peritendinitis or bursitis, trauma, nodules from rheumatic arthritis, or gout tophi. The patient was advised for general medicine consultation for further investigation and medical management because of TX. Her Total cholesterol level was 361 mg/dL, and the total LDL-C level was 277 mg/dL as per the laboratory report. Her mother was having hyperlipidemia and under treatment for the same. She was diagnosed as...
heterozygous FH and she was put on atorvastatin 20 mg once daily as suggested by the concern physician. Her LDL-C level was 198 mg/dL after 3 weeks of atorvastatin therapy. There was no reduction in the size of the swelling after 3 weeks. There was a pain due to the mechanical block on the dorsiflexion of the foot and the patient was uncomfortable during walking. The swelling was intervened surgically due to persistent pain and gait disturbance. Intraoperatively, full-thickness involvement was found and the total excision of the affected part was done through the posterior approach and sent for histopathological examination. In the neutral position of the ankle, a gap of about 6 cm was created due to excision of the affected part and that was reconstructed by harvesting short FHL graft and transfixing with bioabsorbable interference screw through the calcaneum (Fig. 2b). Histopathological examination confirms our diagnosis of TX (Fig. 2a). Macroscopically, it showed cut section which was firm, solid, and greyish-yellow in color. On the microscopic section, fibrocartilaginous tissue infiltrated by foamy histiocytes was seen. There were large areas of cholesterol cleft surrounded by foreign body giant cells.

A below-knee splint was applied dorsally in plantar flexion position for 2 weeks. The dressing was done on the 2nd and 5th day, which was clean and healthy, and the suture was removed on day 14th. A boot cast in plantar flexion was applied for the next 2 weeks followed by in neutral position for 2 weeks with partial weight mobilization with a walker. The patient was allowed for full weight-bearing mobilization after 6 weeks of surgery. The patient was able to stand on her toe at 4 months. The patient was followed up every month and at the end of 1 year, there was no recurrence.

Discussion

Xanthoma is defined as pseudotumors of connective tissue, consisting of histiocytes with lipid deposits, out of which lipid constitutes around 33% of dry weight and collagen constitutes 24% of dry weight [6]. These tumors manifest as encapsulated lobular masses which is yellow in appearance. Most often, TX associated with FH [7]. The size of the lesion increases with age [8]. As per existing literature, among whites, 1/500 are heterozygous for FH and
1/1,000,000 are homozygous [9, 10]; however, in most countries, a larger population is not even diagnosed [11]. The extent of underdiagnosis and undertreatment of individuals in the general population with FH is largely unknown, and there was no uniform registry maintained in each region [12].

Plain X-ray is usually the first imaging modality showing the soft tissue component (thickened TA with or without associated calcification) [13, 14]. CT scan can be useful to measure the thickness of the TA [15–17]. Ultrasonography is the investigation of choice, and it can able to detect focal hypoechoic areas of lipid deposit inside the normal hyper-echogenic tendon along with the measurement of tendon diameter [18–20]. Magnetic resonance imaging scans can be useful in delineating collagen fiber (low signal intensity structure) from infiltrating foam cells and inflammatory cells (high signal intensity structure) [21].

The initial modality of treatment for TX is usually with hypolipidemic medications (statins) which may help in the regression of the disease [22–30]. A newer drug-like Ezetimibe with a diet low in sterol content is also showing the promising result. Estrogen therapy has also shown to have some impact on the decrease of the tendon size [31]. In advanced cases, surgical resection is required when the patient presented with mass with pain and gait disturbance [32]. The rate of recurrence is 12–15% if partial resection is done [32]. Total resection of the tendon from musculotendinous junction and calcaneum is recommended to decrease the recurrence [33]. Use of various tendons to supplement the gap after excision has been advised by various authors. The tensor fascia lata, FHL, gracilis, and semitendinosus are the usual options for reconstruction [34]. Dinçel et al. [35] recommend the use of quadriceps tendon graft with bony fragments when the defect is large. FHL transfer was described by Pearce et al. [36]. Reconstruction with peroneus brevis was proposed by Gallant et al. [37]. Scagenelli et al. [38] have found the reconstruction by using allograft satisfactory in their study. Dinçel et al. [35] have shown good results in bilateral xanthoma excision and reconstruction done by quadriceps tendon with bone fragment in 1 side and other side reconstruction with FHL transfer in different stages. FHL tendon has definite advantages in the Achilles tendon reconstruction than other existing options as (1) it is close to the Achilles tendon; easy to harvest, (2) the FHL muscle has the same function as the triceps surae; same excursion, (3) it has adequate strength; hypertrophy of FHL occurs to bear the strength of TA, and (4) it has the same axis of moving with the Achilles tendon [39]. There is a 2–8% chance of re-rupture of Achilles tendon treated surgically as a complication [40–43].

In this case, we have done total excision of xanthoma, and reconstruction was done with FHL transfer at 1 stage. There was no recurrence till 1 year of follow-up period with very good functional outcomes. She was able to stand, walk independently at a 6-week postoperative period. Power was adequate, and she was able to stand on her toes at 4 months after the surgery. We believe the total excision of the tumor mass with reconstruction with FHL autograft is a valid option with a low recurrence rate and good outcome.

**Conclusion**

Xanthoma is a rare pseudotumors of connective tissue, consisting of histiocytes with lipid deposits. Most often, TX associated with FH, which can be established with clinical examination, biochemical tests, and radiological imaging. Although statins are the drug of choice in such conditions, in the advanced stage, when it is painful and associated with gait disturbance, surgical intervention is needed. The total excision of the lesion followed by augmentation with FHL tendon graft is a valid option with good functional outcome.
Statement of Ethics

Consent was taken from the patient at the time of surgery. The patient gave her written informed consent to publish their case (including publication of images). The study protocols have been approved by the committee on human research of Kalinga Institute of Medical Sciences, Bhubaneswar, Odisha, India.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

S.N.N., S.S., and S.K.T. were the operating surgeons. Data were collected by S.S. and A.K.G. N.K.S. and S.N.N. wrote up the paper. M.T. did the histopathological study for diagnosis. All authors read the manuscript and agreed on the content.

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