

Reconstruction of bilateral tendoachilles with fascia lata graft

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ABSTRACT

A 19 year old male presented with progressive enlargement of both tendoachilles for 2 years and difficulty in walking for 3 months. The neurological history and examination revealed progressive mental deterioration and ataxia. The blood investigation revealed hypercholesterolemia. We report this rare case of cerebrotendinous xanthomatosis with bilateral tendoachilles enlargement, which was treated by excision of bilateral tendoachilles and reconstruction with fascia lata. The American Orthopedic Foot and Ankle Society hindfoot score was 93/100 bilaterally and the subjective evaluation of the patient showed very good results.

Key words: Fascia lata graft, tendoachilles, xanthomatosis

INTRODUCTION

anthomas are benign lesions characterized by the local collection of lipid laden macrophages, giant cells and other inflammatory cells in response to the deposition of cholesterol in tissue.¹ Xanthomas usually occur in the second or third decade as yellow patches, papules or nodules in the skin, tendons and soft tissues. Bilateral enlargement of tendoachillis has been reported in xanthomatosis and associated with high levels of plasma lipids.² This has also been reported in cerebrotendinous xanthomatosis (CTX).³⁻⁵ CTX is a rare, autosomal recessive, lipid storage disorder characterized by accumulation of cholestanol and cholesterol, predominantly in the brain, spinal cord, peripheral nerves, tendons, lungs, liver and kidneys.⁶ Infantile onset diarrhea, childhood onset cataract, adolescent to young adult onset tendon xanthoma and adult onset progressive neurologic dysfunction (dementia, psychiatric disturbances, pyramidal and cerebellar signs, dystonia, peripheral neuropathy and seizures) are the

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most prominent features of this disease.⁷ Tendoachillis xanthomas can cause pain, swelling, loss of function and problems with footwear.⁸ Several procedures have been described for the reconstruction of chronic degeneration of Achillis tendon. The majority of reconstructive procedure involves autogeneous tissue transfers or heterogeneous homologous grafts.⁸⁻¹⁰ We report a rare case of bilateral xanthomatosis of tendoachilles tendon sheath which was managed by wide surgical excision and reconstruction with tensor fascia lata.

CASE REPORT

A 19 year old male presented to outpatient department with slow and progressive enlargement of tendinous nodule on the posterior aspect of ankle for last 2 years. The disease had progressively worsened in last 3 months with difficulty in walking. The patient was mentally retarded (intelligence quotient 55). Clinical examination did not reveal nodule or mass anywhere else in the body. The patient's blood lipid profile showed hypercholesterolemia (serum cholesterol level 331.7 mg/dl) while serum uric acid level was normal. There was no history of prolonged medication or antecedent trauma. There was no similar history in any of his siblings. Karyotyping of the patient and his parents was normal.

The radiographs of the lower legs and ankles demonstrated bilateral soft tissue shadows on the posterior ankle region [Figure 1]. The masses on both sides extended from the ankle to the lower calf and were located along the expected course of the tendoachilles [Figure 2]. No associated calcification or osseous abnormalities were seen. Magnetic resonance imaging of the brain showed diffuse cerebral and cerebellar atrophy.

Fine needle aspiration cytology showed giant cell rich lesion both side. The patient was taken for bilateral wide local excision of tendoachilles and reconstruction of the tendon with fascia lata graft.

Operative procedure

The curvilinear lazy S incisions were made on the posterior aspect of the lower calf till tendoachilles insertion in prone position. Tendoachilles exposed which revealed large grayish white rounded mass involving $3/4^{th}$ of length on the right side and full length of tendoachilles on the left side [Figure 3]. Abnormal tendoachilles tendons were excised almost completely creating a large gap which was not possible to reconstruct by primary repair. The defect in each limb was almost from musculotendinous





Figure 1: Lateral radiograph of left ankle with leg showing increased soft tissue shadow in tendoachilles region



Figure 2: Clinical photograph of patient showing bilateral tendoachillis enlargement



Figure 3: Peroperative photograph showing appearance of right and left tendoachilles xanthomata



Figure 4: Peroperative photograph showing tendoachilles gap is reconstructed with fascia lata graft tunneled through a drill hole in calcaneum. Note during the repair foot is kept in equinus to reduce the gap

the calcaneum. During the repair, the foot was kept in equinus to reduce the gap and produce repair under adequate tension [Figure 4]. Other end of fascia graft was sutured back to the muscle end of the gastrocnemius and soleus with proline 3-0. The closure was done in layers. Thompson's test was done prior to dressing to check the integrity of reconstruction. The procedure was repeated in other limb in the same sitting. The excised specimen was sent for histopathological examination.

Pathological evaluation

Gross specimen showed large cylindrical grayish white soft tissue mass that involved the tendoachilles completely on the left side and almost 3/4th of tendon on the right side. Specimen was measuring 12 cm \times 3 cm \times 3 cm on the right and 13 cm \times 4 cm \times 4 cm on the left side in craniocaudal, transverse and anteroposterior dimension [Figure 5].

Microscopic examination showed a large number of granuloma composed of giant cells with foamy cytoplasm and lymphocytes in subepithelial tissue. Giant cell were mostly langhans type [Figure 6]. Adjacent tissue was with a smaller number of granuloma.

Postoperatively, the ankle was immobilized in nonweight bearing plaster at 15° of plantar flexion for first 6 weeks and then in plantigrade cast for another 4 weeks. Stitch removal was done at 3 weeks postop and showed no evidence of wound dehiscence, infection or discharging sinus.

After removal of plaster cast the patient was allowed to begin active physiotherapy. Weight bearing was delayed as this was a bilateral case. After 3 months, patient was allowed to increase weight bearing progressively within the limits of comfort. At 5 months followup the patient was able to walk without crutches. At 1 year followup, range of motion in both ankle joints was 15° of plantar flexion and 10° of dorsiflexion. The patient was able to stand and walk on toes, which he was able to since 6 months postsurgery [Figure 7]. Despite low intelligence quotient, at 1 year followup, the patient was independently mobile without pain with good cosmetic outcome. The functional outcome was satisfactory in all aspect except moderate limitation of sagittal and hindfoot motion with an American Orthopedic Foot and Ankle Society ankle hind foot score of 93/100.

DISCUSSION

Giant cell tumor of tendon sheath is a benign soft tissue tumor which arises from the complex of the tendon sheath and periarticular soft tissues of small joints and often associated with history of trauma.¹¹ Fibrous histiocytoma of synovium, pigmented nodular synovitis, localized nodular tenosynovitis, benign synovioma, fibrous xanthoma of the synovium and pigmented villonodular



Figure 5: Gross specimen showing large cylindrical grayish white soft tissue mass involving tendoachilles



Figure 6: Microscopic examination showing a large number of granulomas composed of giant cells with foamy cytoplasm and lymphocytes



Figure 7: Clinical photographs showing (a) the patient is able to squat (b) the patient is able to stand on the tip of toes

bursitis are all synonymous names of the giant cell tumor of the tendon sheath.^{12,13} Other differential diagnosis to be considered are ganglion, epidermal inclusion cyst, lipomas, rheumatoid nodule, xanthoma, hemangioma and neurofibromatosis.^{5,14-16}

Bilateral tendoachilles enlargement has been reported in xanthomatosis. Xanthomas usually appear as yellow patches, papules or nodules in skin, tendon and soft tissues and associated with high plasma lipid levels. There is a well-established association between tendinous xanthomas and hyperlipidemia. In particular, type IIa hyperlipidemia and type III hyperlipidemia have shown a strong association with tendinous xanthomas. Bilateral tendoachilles involvement has also been reported in CTX, an autosomal recessive, rare lipid storage disease described by Van Bogaert in 1937. CTX is a rare disease and its symptoms vary, some cases may have atypical manifestations, which may lead to their misdiagnosis as foot and ankle tumors. In patients with CTX, bile acid synthesis is abnormal because of a defect in the activity of the hepatic mitochondrial enzyme sterol 27 hydroxylase (CYP 27). This condition usually occurs in the first 30 years of life and the clinical presentation can be very variable with neurological symptoms, cataract, atherosclerosis and tendinous xanthomas. The firm masses of Achilles tendons may be the first symptom the patient recognizes because it can jeopardize his or her ability to walk.

Magnetic resonance imaging of the foot and ankle is the modality of choice as it allows soft tissue tumors to be assessed anatomically and is helpful in establishing accurate preoperative diagnosis.^{17,18}

Several procedures have been described for the reconstruction of chronic degeneration of Achilles tendons. The majority of reconstructive procedure involve autogeneous tissue transfers or heterogeneous homologous grafts.⁸⁻¹⁰ However, the latter was not commonly used because of limited suitable donor material availability and higher rates of immunological rejection and infection.⁷ Among autogeneous tendon transfer, peroneus brevis and flexor hallucis longus transfer are commonly used, although tibialis posterior is also been used for reconstruction. Other available therapy option for reconstruction are V-Y tendon alignment with plantaris weaving, gastrocnemius free turndown flaps, fascia lata, flexor digitorum longus transfer and bone tendon bone autograft harvested from the knee extensor mechanism. The use of cadaveric tendoachillis allograft has also been advocated,^{8,10} but it was not possible in our case as tissue banking facilities were not available at our center. In our case, reconstruction following resection of both tendoachilles was difficult as following removal there was large defect in the Achilles tendon, which was too extensive to be reconstructed satisfactorily by any of the available methods. We used fascia lata strip to bridge the defect because of limited donor site morbidity and sufficient length of graft, further we were not sure whether other local tendons were free from the disease. However, we could achieve satisfactory results bilaterally with this procedure.

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