

XANTHOMATOSIS INVOLVING TENDONS AND SACRAL REGION- A RARE COMBINATION

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PRESENTATION OF CASE

A 33-year-old male patient presented with multiple lobulated, nodular, firm swellings over both Achilles tendons, patellar tendons and sacral region. The lesions started developing around 25 years ago and have gradually progressed to their present size. The patellar and tendoachilles swellings were excised and histopathology proved the swellings to be xanthomas. This 33 years old non-diabetic, non-hypertensive male patient presented with gradually progressive multiple lobulated, nodular, firm swellings over both Achilles tendons, patellar tendons (Figure 1, 2) and sacral region for last 25 years (Figure 3), few of which demonstrated surface scarring due to friction. The patient had social problems with regards to the cosmetic deformity the lesions caused and hence attended our outpatient department. On clinical examination, the swellings were free from the underlying bone at all places. They were not associated with pain, fever, weight loss or any other conditions. The patient also provided with history of similar swellings over both triceps tendons, which were operated two years back outside our hospital. The patient has denied any recurrence of the operated lesions. The patient did not have past history of any other illness. He was non-smoker, non-alcoholic and bowel-bladder habit was regular. General and systemic examinations were within normal limit. There were no other swellings or lesions present in any other part of his body other than those already described.



Figure 2. Swelling over Achilles Tendons



Figure 3. Swelling over Sacral Region



Figure 1. Swelling over Patellar Tendons



Figure 4. Yellowish Lobulated Tendon Surface Peroperatively

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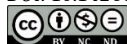




Figure 5. Swollen Tendon Peroperatively



Figure 6. Tendon Infiltrated with Xanthomatous Material



Figure 7. Superficial Wound Infection Post-Operatively

DIFFERENTIAL DIAGNOSIS

The distribution of the xanthomatous lesions may pose difficulty in clinical diagnosis. Important differentials to be considered are: Giant cell tumour of tendon sheath can be a differential diagnosis in case of solitary lesions, especially those involving the hand.^[1] These can be best differentiated by histology, wherein xanthomas are found to have fewer giant cells than giant cell tumours.

Ganglion cysts usually present as visible painless lumps in relation to joints or uncommonly within soft tissues.^[2,3] Most ganglion cysts have a typical location around the hands. Other notable sites being knee, spinoeloid notch and ankle. Ganglion cysts show the typical characteristics of cystic lesions on imaging studies. Aspiration usually shows clear fluid.

Soft tissue haemangiomas are often found within extremities representing up to 7% of all benign soft tissue tumours in the general population.^[4] Diagnosis is aided by radiographs and CT scan, which show soft tissue mass and phleboliths. MRI is the standard for imaging evaluation^[5] showing heterogeneous high signal intensity on T1 weighted images due to adipose tissue and heterogeneous high signal in T2 weighted images due to angiomatous components. Histopathology is confirmatory.

Pigmented villonodular synovitis can be considered if the swellings are adjoining large joints.

Chronic tophaceous gout and rheumatoid nodules can also be considered as differentials.

PATHOLOGICAL DISCUSSION

Laboratory Parameters

- Hb%- 12g%.
- TLC- 10,200/cumm.
- ESR- 18 mm/1st hr.
- Blood Sugar: Fasting- 80 mg/dL, Post-prandial- 110 mg/dL.
- Urea- 18 mg/dL, Creatinine- 0.75 mg/dL.
- Uric acid- 4.5 mg/dL.
- Bilirubin- 0.6 mg/dL.

Lipid Profile

- LDL- 166.23 mg/dL.
- Triglycerides- 177.17 mg/dL.
- HDL- 21.87 mg/dL.

Radiography

Radiographs of ankle, knee and lumbosacral spine revealed no obvious underlying bony abnormality.

MRI Examination of Lumbosacral Spine

MRI was done to exclude any communication of the swelling with the underlying spinal canal. No communication was found.

Surgery

A decision of excision was taken. Each of the lesions (both tendoachilles, both knee and sacral region) were operated on separate sittings. Peroperative Findings:

- Yellowish appearing lobulated surface (Fig. 4).
- Affected tendons appeared swollen (Fig. 5).
- The tendon fibres could not be separately identified- they appeared to have been infiltrated by the xanthomatous material (Fig. 6).

- The sacral xanthoma did not appear to arise from any enthesophytic attachment site. It was confined to the subcutaneous plane.

It was not possible to remove all the xanthomatous tissue, because the tendon fibres were infiltrated with the xanthomatous tissue.

Post-Operative

Post-operatively, there was superficial wound infection in one site (Fig. 7).

Histopathological Examination

Biopsy revealed lipid laden spindle-shaped cells along with multinucleated xanthomatous giant cells.

DISCUSSION OF MANAGEMENT

Xanthoma is derived from the Greek word- "xanthos" meaning "yellow." It is defined as deposition of cholesterol rich material in tendons^[6] and other body parts in various disease states.

To the knowledge of the authors, the combination of tendinous xanthoma in conjunction with sacral region xanthoma has not been reported before, thus providing valuable addition to the current literature. Conventionally, tendinous xanthoma involves metacarpophalangeal joints of fingers, also Achilles, patellar and triceps tendons. Rarely, involve toe extensor tendons and plantar fascia.^[7] Xanthomatous material infiltrates the tendons, which become structurally weak and are more prone to injuries.^[8] Xanthoma is also found commonly in association with hyperlipidaemic states like familial hypercholesterolaemia. The patient did not have any gait ataxia, mental retardation or abnormalities on neurological^[9] examination, thus ruling out cerebrotendinous xanthomatosis.

Surgery was undertaken mainly for cosmetic purpose. However, since these are infiltrative lesions, complete surgical removal is unlikely. The same issue was encountered in our case as well.

Post-surgical recovery of the patient was uneventful except for a superficial wound infection at a single site. On further followup, no obvious recurrence has been noted at the surgical sites yet.

Conclusion

Tendinous xanthomatosis with involvement of the sacral region is a rare entity, which makes this case report a first of its kind.

FINAL DIAGNOSIS

Xanthomatosis involving multiple tendons (bilateral patellar and Achilles tendons) and sacral region is our final diagnosis.

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