

Ochronosis of Achillis Tendon Enthesopathy- Rare Case

Dr Asha Lata Jr, Dr Azeez Q and Dr S N Patil*

Anil Neerukonda Hospital, NRI Institute of Medical Sciences, Sangivalasa. Visakapatnam, India

***Corresponding Author:** Siddaram Patil, Professor & HOD, Anil Neerukonda Hospital, NRI Institute of Medical Sciences, Sangivalasa. Visakapatnam, India.

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Case History

A 43 year old male patient presented with pain on left tendoachillis region, with swelling and difficulty in walking .unable to planter flex ion of 15 days duration .History of sudden jerk when trying to get up from chair and heard a snap, followed by pain which lasted for 10 days

On clinical examination: weakness in limb, difficulty in walking and driving car. Recent past he had similar complain on other leg, but improved by conservative treatment.

Swelling, step sign, positive Thompsons test ,unable to actively dor-siflex the ankle joint , pain sub sided by medication, Routine inves-tigation where normal , ultra sound showed tear in Tendo-Achillis tendon , Planned for Tendon repair surgically.

On questioning the patient admitted that his urine was dark on standing and his mother having brown pigmented patch on both

cheeks and blackening of urine on standing. Urinary chromatogra-phy showed elevated levels of homogenitistic acid.there is history of consanguinous marriage of their parents.

Intra-operative findings – Black colour crystals were found in and around the tendo-achillis

Histopathology

Gross: Multiple grey brown to grey tan ,gritty bits ,all together mea-suring 3 cms by 2 cms.

Microscopy: cartilage and dense fibrous tissue fragments with lo-cally scatered deposits (Figures 1,2) of pale amber –coloured pig-ment intermingled with in collagen fibers.At foci the pigment is bro-ken into crystalline fragments with pointed ends.(Figures 4,5).

H&Estaining: Focal ares of dystrophic calcification and congested blood vessels with scanty inflammatory cell collections are also noted. Figures 5.6)



Figure 1:

Figure 1:



Figure 2:

Figure 2:

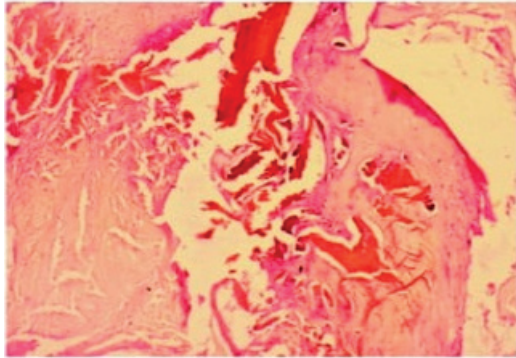


Figure 3: 10X, H&E staining

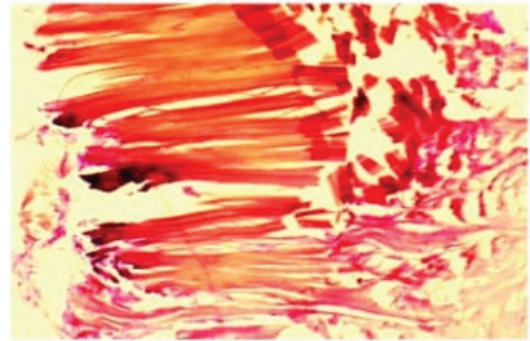


Figure 6: 10X, H&E staining

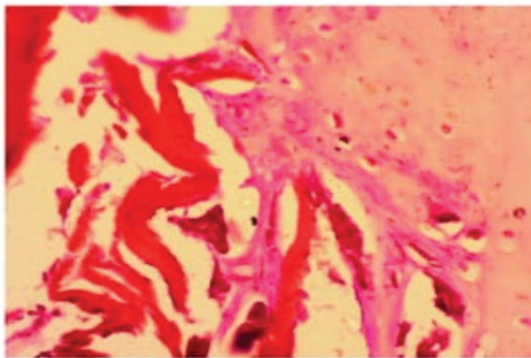


Figure 4: 10X, H&E staining

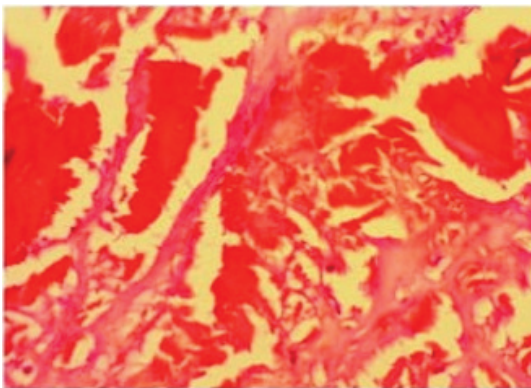


Figure 5: 10X, H&E staining

Dignosis

Featur are consistant with OCHRONOSIS OF TENDO- ACHILLIS ENTHESOPATHY

Discussion

Ochronosis is a rare metabolic disease, autosomal disorder caused by defect in tyrosine metabolism due to congenital absenc of homogentisic acid oxidase present on chromosome 4q23 with a prevalnce of about 1:1,000,000. Spontaneous tendon ruture are common in these patients with tendoachillis rupture as a clinical menifestation. When middle aged patients present with chrinic bach ache and enthesopathy, one should keep in mind the possibility of ochronosis, which can be confirmed by urine chromatography for homogentisic acid, active survilance for cardiac, renal and prostate complications should be done after the 4th decade of life.

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