Surgical Management of a Spontaneous Black Tendon Rupture

Spontan Siyah Tendon Rüptürünün Cerrahi Tedavisi

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ABSTRACT

Spontaneous non-traumatic rupture of Tendo-Achilles (TA) as a presenting feature of alkaptonuria is unusual. In this disorder, there is deposition of homogentisic acid in the cartilage of major joints and intervertebral disc is responsible for the degenerative changes. Infrequently, it also gets deposited in major tendons that renders them structurally weak ensuing in pathological ruptures. A case is presented with non-traumatic TA rupture and its surgical management in alkaptonuria.

Key Words: Tendoachilles, alkaptonuria, spontaneous, rupture, repair

INTRODUCTION

Non-traumatic spontaneous rupture of a tendon should lead to suspicion of a pathological tear and it should be thoroughly evaluated. Alkaptonuria is a rare form of in born error of metabolism of amino acids (phenylalanine and tyrosine), in which urine turns black due to presence of excess of Homogentisic Acid (HA) (1). This breakdown product also gets deposited in various soft tissues like cartilage, ligaments, intervertebral discs, bones and heart valves (Ochronosis). The deposition of HA in the Tendo-Achilles (TA) and subsequent rupture is an unusual incident. There is paucity of cases in the literature describing such an event and its outcome after surgical management.

CASE REPORT

An apparently healthy 35 years old male presented to our Foot and Ankle clinic with pain in the left ankle and difficulty in walking since twenty days. Twenty days back, while getting down a bus he felt a sharp pain in his left ankle. After some time, he continued to walk with pain and experienced some difficulty in movements at the left ankle. There was no similar complaints in the past. There was no significant medical and family history. General examination was unremarkable. There was decreased power of plantar flexion of the left ankle as compared to the opposite side. However, Thompson’s test for a ruptured TA was negative.

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An MRI study of the left ankle revealed a partial thickness intra-substance tear of TA in the distal fibres with retraction of fibres proximally from insertion site, suggestive of a Type I TA tear (Figure 1). Surgical management was considered in view of clinical suspicion of a pathological rupture.

Under spinal anaesthesia and tourniquet control, patient was placed in prone position. A 10cms midline linear incision was made over the posterior aspect of ankle. On exposure, the distal end of proximal torn part (for about 2cms) of the tendon appeared blackened (Figure 2).

Rest of the tendon on either sides appeared normal in appearance and texture. On palpation, a grating sensation of this segment was appreciated. This pathological segment was excised and sent for histopathological examination. The two ends were repaired together by Krackow suture technique using high strength Orthocord sutures. Paratenon was repaired with 3-0 vicryl and patient was shifted out in an above knee slab with knee in 20° flexion and ankle in 20° plantar flexion. This position was maintained for a period of three weeks in an above knee cast and later converted to below knee cast for another three weeks period.

Patient was mobilised non-weight bearing with support for six weeks followed by gradual partial weight bearing as tolerated. Follow-up at sixteen months his American Foot and Ankle score is 97.

Post-operatively, patient gave history of passing dark urine (Figure 3) and in childhood he and his brother had history of staining the diapers with dark urine, however, no further investigations or treatment were pursued. His urine examination for HA was strongly positive. Histopathology suggested features of calcific tendinitis. His liver function tests, renal function tests, ECG, Echocardiography and skeletal survey were all essentially normal.
Alkaptonuria is a rare autosomal recessive disorder that occurs due to deficiency of an oxidase enzyme necessary for the metabolism of HA. The incidence of this condition is reported to be 1 in 250000 to 1 in 1000000 live births (2), however there is no Indian subcontinent data available.

In childhood, urine turning black from exposure is one of the earliest symptoms to be presented. Patient and his brother reported to have a childhood history of dark staining of the diapers. Urinary and plasma HA levels can be measured using spectrophotometric and chromatographic method.

Joint pains usually begin in the adulthood and weight bearing joints are frequently affected. Radiographic screening of the spine and pelvis is useful to determine the onset of ochronotic arthropathy. In our patient, the disc spaces were well maintained and there were no abnormal calcification, osteopenia or degenerative changes observed. The facet joints and the sacro-iliac joints were also unaffected. Similarly, other peripheral joints were essentially normal. By the fourth or fifth decade the disease usually progresses from simple alkaptonuria to alkaptonuric arthropathy in approximately 30% of subjects (3).

Scleral pigmentation (Osler sign) and pigmentation of the ear cartilages are also frequently seen sometime around 3rd decade. Furthermore, discoloration of the forehead, axilla, groin, teeth, palms, and soles and dark staining of the nails can also be seen. However, none of these signs were present in the index case. Cardiac involvement is usually seen in the form of mitral or aortic valve calcification or regurgitation which needs to be screened by echocardiography.

Pathological or spontaneous rupture of tendons due to the deposition of HA in the substance of tendons are extremely rare presentation. Large tendons such as quadriceps tendon and TA are the ones that are frequently involved. The accumulation of HA inhibits collagen cross-linking, leading to a reduction in the structural integrity of collagen, and thus increasing the likelihood of spontaneous rupture (4).

The diagnosis of TA rupture can be comfortably made based upon clinical examination. This can be further confirmed by either Ultrasonography or Magnetic Resonance Imaging studies. There are four types of TA injuries (5). Accordingly, Type I tears (as seen on MRI in the index case) can be managed by non-operative methods. Nevertheless, based on patient’s symptoms and suspicion of pathological rupture, surgical repair was contemplated. Aggressive mobilisation is generally avoided in order to prevent the risk of re-rupture. A midterm follow-up in the index case showed favourable outcomes. Alkaptonuria patients should avoid strenuous exercises and foot straining especially in patients developing early orthopaedic manifestations (6).

There is no effective therapy available as a cure for this condition, however, early detection of the disease is essential to treat involvement of other systems (cardiovascular and renal system). There is a limited role for dietary modification (avoiding food such as meat, milk and nuts, which are rich in phenylalanine and tyrosine). Role of vitamin C has not been clearly defined in the management. Regardless, our patient was advised dietary modifications and vitamin C (1gm OD for two months), as an antioxidant.

A high index of suspicion and a prompt evaluation is warranted for a pathological rupture of TA. Reasonable outcome can be expected with surgical management of a TA rupture in patients with Alkaptonuria.

Conflict of interest
No conflict of interest was declared by the authors.

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