

Upper ankle ligament rupture and long term problems in a patient with Ehlers Danlos Syndrome – a case report

Case Report

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Abstract: Rupture of the upper ankle joint ligaments is a common injury. Therapy in patients with Ehlers Danlos Syndrome (EDS) is a challenge due to the autosomal-dominant collagenopathy. Therapeutic recommendations and long term results in treatment of patients with EDS are rare. Therefore uncertainty exists. We present a case of clinical and radiologic long term results after ligament rupture of the upper ankle joint in a patient with EDS. A literature review tries to reveal current therapeutic strategies in patients with EDS.

Keywords: Ehlers Danlos Syndrome • Upper ankle joint • Ligament rupture

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1. Introduction

Rupture of the upper ankle joint ligaments is a common injury. Early treatment concentrates on pain relief, short term immobilization and cryotherapy. The golden standard is a functional ankle brace for six weeks and stepwise weight bearing to full weight. Full stress can be put on the injured area about three months afterwards. Patients with Ehlers Danlos Syndrome (EDS) do suffer from autosomal-dominant collagenopathy. Main symptoms are hyper elastic, easy injured skin and hypermobile joints [1]. The prevalence is low with about 0.01-0.02% and literature about treatment at ligamentous injuries of EDS patients is rare. Therefore uncertainty exists, if standard treatment should be advised to patients with EDS and ligament ruptures. We present a case of long term results after ligament rupture of the upper ankle joint in a patient with EDS and try to prepare advice for physicians treating this rare combination.

2. Case presentation

We present a 42 years old male patient that suffered ankle sprain on the right side in 2008. A ligament rupture of the anterior talofibular ligament was suspected initially and confirmed by MRI. The patient was treated with a functional ankle brace for six weeks and received physiotherapy afterwards. However the patient did never fully recover. Due to chronic fulgurant pain sensation, unsteady gait in combination with persistent fear to fall down as well as frequent anterior talus luxation, the patient was sent for MRI in 2011 which revealed a non-union of the anterior talofibular ligament and a rudimentary calcaneofibular ligament. The patient was asking for help and he was offered a ligament reconstruction according to the algorithm introduced by Zwipp et al. in 1994 [2] but he finally refused to be operated.

In 2012 the same patient was reexamined and symptoms did not vanish. On the surface the right upper ankle joint did not show dramatically abnormality to the uninjured side (Figure 1). On reexamination the patient showed normal gait and normal foot roll. One leg standing on the right side was possible while bouncing was denied because of fear of pain. A squatting position was causing pain within the upper ankle area. Compared to

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Figure 1. Inconspicuous superficial view comparing injured (right) with uninjured side (left), maximum inversion



Figure 2. Lateral widening of the upper ankle joint on stress view

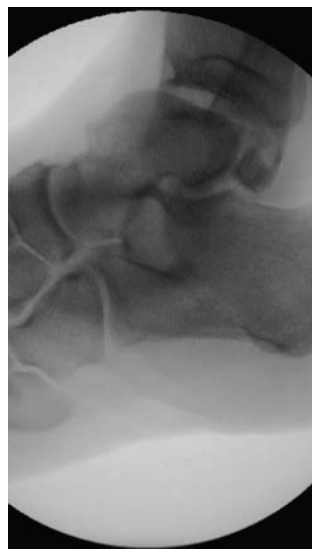


Figure 3. Talar shift on stress view

the uninjured side, the right upper ankle joint unfolded noticeably more (Figure 2). This was verified with fluoroscopy. Also talar shift was seen exceeding physical values (Figure 3). Range of motion was limited compared to the uninjured side.

3. Discussion

Ankle sprain of the upper ankle joint is a common injury and concomitant rupture of the ligaments does occur regularly. Degenerative changes do have an influence on ligament rupture [3]. Conservative treatment shows sufficient restauration of ligamentous stability. Surgery is not superior to conservative treatment [4], but offers a good alternative in persisting instability [5]. However nearly 50% of all patients with chronic ligamentous instability show good results after being consequently immobilized by ankle orthosis including a lateral raised sole section and following physiotherapy [6].

In our case we were confronted by a systemic disease that made treatment more complex. While conservative treatment failed, the patient was offered a tendon reconstruction by Watson-Jones [7] aiming on strengthening the joint and improving the patients' symptoms by reconstruction of the anterior talofibular ligament. Diseases that modify the metabolism and therefore do have an influence on connective tissue such as EDS, Marfan Syndrome or Homocysteinuria are responsible for approximately 1% of chronic tendon disease [8]. Literature on treatment options and long term results in ligament ruptures for patients with EDS is rare. Conservative treatment is performed as usual and indications for operative treatment in acute injury do not differ. Furthermore chronic instability, second stage injuries and re-ruptures are normally indications for surgical treatment [9].

Due to more than 80 surgical techniques that are known, following the algorithm introduced by Zwipp *et al.* 2,5 is recommended. Complication rates in patients not suffering from EDS but undergoing ligament surgery of the upper ankle joint were about 10% in acute injuries and 2% after reconstructive procedures [2]. Infection and reconstructive failure were most commonly seen.

To our knowledge there is no modified algorithms for patients with EDS or similar diseases that affect collagen metabolism. Literature research revealed reports about operative treatment options for glenohumeral stabilization using Achilles tendon [10]. Furthermore patellar tendon rupture in patients with EDS was reported. In one report the authors describe synthetic patellar ligament replacement [11]. They suggested the use of an additional Trevira®-Band augmentation in combination with

an end-to-end suture of the debrided tendon stumps and McLaughlin cerclage. Another case reports exists on usage of suture anchor and temporary protection of the suture with cerclage wiring in case of spontaneous bilateral patellar tendon rupture [12]. Although there are publications describing successful use of autologous ligament replacement in patients with EDS [13,14], it must be taken into account that poor collagen synthesis in EDS patients leads to delayed wound healing and increases the risk for mechanically insufficient scarring which is one of the major causes in transplant failure [15]. Rombaud et al. reported about altered passive properties of the plantar flexors muscle-tendon tissue in patients with EDS. Furthermore he found a significantly lower Achilles tendon stiffness in the affected patient group than in the control group [16].

Due to the fact that long term instability and altered joint kinematics cause preterm joint arthrosis [5,17,18], full restoration of ligamentous structures is necessary. However our patient failed conservative therapy and refused operative treatment. Due to the underlying disease and MRI-results, showing non-union and rudimentary ligamentous structures without a scarring tendency, we conclude that the modified collagen synthesis in EDS worsened the initial prognosis. Treatment of acute and chronic ligamentous injuries in patients with altered collagen metabolism are challenging and timing of conservative and surgical treatment needs to be reflected.

4. Conclusion

To the best of our knowledge, this is the first report of long term consequences of upper ankle ligament rupture in an EDS-patient. This case highlights the need of better understanding of ligament injuries in patient with chronic laxity because of their unpredictable outcome.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Author's contributions

The work presented here was carried out in collaboration between all authors. KH, TD and MP analyzed and interpreted the patient data and were major contributors in writing the manuscript. PK and RMS co-worked on interpretation and discussed analyses, interpretation, and presentation. HCP gave critical and final approval. All authors have contributed to, seen and approved the manuscript.

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