Carpometacarpal Arthroplasty and Ulnar Collateral Ligament Reconstruction in a Patient with Suspected Ehlers-Danlos Syndrome: A Case Report and Review of the Literature

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Abstract

Keywords

- carpometacarpal arthritis
- Ehlers-Danlos syndrome
- ulnar collateral ligament

Ehlers-Danlos syndrome (EDS) is a disorder that presents with a heterogeneous constellation of symptoms, ranging from clinically silent to rapidly deteriorating. It is a multisystemic connective tissue disorder that may result in any number of manifestations, with joint hypermobility being a classic manifestation. We present a case of a 58-year-old woman, with suspected EDS, who presented with several years' history of bilateral thumb pain, with imaging and physical examination findings consistent with bilateral thumb carpometacarpal (CMC) arthritis with metacarpophalangeal joint (MPJ) hypermobility. The Beighton hypermobility score was consistent with suspected EDS. Our patient underwent thumb CMC arthroplasty with ulnar collateral ligament (UCL) reconstruction.

Introduction

A 58-year-old woman presented to us with complaints of bilateral thumb pain, left greater than right, present for several years that had progressively worsened over time. She also complained of pain and “clicking” at her right thumb metacarpophalangeal joint (MPJ), with increasing difficulty and pain with grip and writing. Her medical history was significant for hiatal hernia, periodontal disease, chronic bilateral shoulder dislocations since high school, foot arthrodesis, hip arthroplasty, and removal of bilateral cataracts. She also reported several family members requiring hip replacements at young ages, in addition to other orthopedic problems. She had not previously been diagnosed with any type of collagen disorder.

On physical examination, the patient was noted to have bilateral thumb carpometacarpal (CMC) grind with tenderness as well as severe hypermobility and laxity of the left thumb at the MPJ, with 45 degrees of radial deviation (►Fig. 1). She was also noted to have general laxity of her skin and was found to have the Beighton hypermobility

score of 7. These findings along with her medical and family history increased suspicion for undiagnosed Ehlers-Danlos syndrome (EDS). The patient refused formal testing. X-rays were obtained demonstrating significant osteoarthritis of the first CMC joints bilaterally and resting radial deviation of the left metacarpophalangeal joint (►Fig. 2).

At this time, secondary to the instability of the left thumb ulnar collateral ligament (UCL), the decision was made to proceed with left thumb CMC arthroplasty with UCL reconstruction. An incision was made overlying the CMC joint. A trapeziectomy was performed. A 3-mm burr drill was used to create an oblique pathway through the metacarpal base. A volar forearm incision was made, and half of the flexor carpi radialis (FCR) tendon was harvested and passed through the first metacarpal base and secured to the FCR insertion at the second metacarpal base. The anchovy was rolled and placed in the trapeziectomy site with a piece of Gelfoam.

Next, a small incision was made over the MPJ, and drill holes were made in the metacarpal head and P1 base. The palmaris longus tendon was harvested and passed through the drill holes, and the tendon was secured on the ulnar side

Fig. 1 Laxity of the thumb metacarpophalangeal joint ulnar collateral ligament.

Fig. 2 Preoperative radiograph demonstrating thumb carpometacarpal joint arthritis and deviation of the metacarpophalangeal joint.

of the MPJ by tying it to itself using a surgeon's knot. The ends were then passed back through the drill holes and again tied in a surgeon's knot on the radial side of the MPJ (►Fig. 3). Both radial and ulnar knots were secured with braided polyester suture. The incisions were then closed with absorbable suture, and the patient was placed in a thumb spica splint.

The patient returned for her first postoperative visit 2 weeks later, and the splint was removed. She was doing

Fig. 3 Intraoperative reconstruction of the ulnar collateral ligament.

Fig. 4 Postoperative result.

well overall with minimal soreness, and incisions appeared well healed. She was placed in a fiberglass thumb spica cast for an additional 4 weeks. At her 6-week postoperative visit, the cast was removed. Again, the incisions appeared well healed, and she had near full range of motion (ROM) and no pain (►Fig. 4). She was started on a physical therapy regimen and was scheduled to return for her next postoperative visit in an additional 6 weeks. At the 3-month postoperative visit, the patient was very pleased with the

outcome of her surgery, and CMC arthroplasty for the right side was scheduled to take place in 1 month. The patient subsequently underwent CMC arthroplasty of the right thumb in similar fashion to the previously done left side. The patient had no evidence of right thumb MPJ instability, and so only a CMC arthroplasty was performed. She was seen at her 2-week follow-up appointment and was doing well from the right-sided surgery and continued having improved strength and function of the left thumb without any complaints.

Discussion

Our patient exhibited signs and symptoms often associated with EDS. This may manifest itself in the hand as trapeziometacarpal instability with subluxation.¹ Ligamentous instability has been proposed as a source of development of arthritic changes in the trapeziometacarpal joint, often as a result as excessive shearing forces.² Several treatment options have been proposed, including arthroscopic tendon interposition arthroplasty³ and joint arthrodesis⁴ with promising results. EDS patients who undergo surgical procedures for joint stabilization have been found to have increased risk of recurrence of instability; however, this risk is lessened for

the thumb, a non-weight-bearing joint.⁵ EDS presents as a spectrum with varying symptomatology. As this disease process becomes better understood, treatment modalities will continue to evolve and improve.

Conflict of Interest

None declared.

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