Bilateral Acromioclavicular Joint Subluxation in an Adolescent with Ehlers-Danlos Syndrome: Case Report and Literature Review

Luis F. Colón, Anthony M. Padgett, Charles W. Powell, Dillon L. Morrow, Jeremy R. Bruce

Department of Orthopaedic Surgery, University of Tennessee College of Medicine, Chattanooga, Chattanooga, TN, USA; University of Tennessee Health and Science Center, College of Medicine, Memphis, TN, USA

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Abstract
Bilateral acromioclavicular joint (ACJ) dislocation is a rare occurrence, with only one reported case in the literature published in 1984. We present the case of a 15-year-old male with Ehlers-Danlos syndrome (EDS) who presented with complaints of subsequent nontraumatic bilateral ACJ subluxations and pain. ACJ reconstruction via an open mini-Mumford procedure was performed on this patient on two separate occasions with successful outcomes. The patient demonstrated decreased pain and increased stability during the postoperative period. EDS is a rare collagen disorder that is usually characterized by abnormal skin elasticity, bleeding tendencies, and, most importantly for this case, joint hypermobility. It is important to have an elevated index of clinical suspicion for potential joint subluxations and injuries in patients with a known history of collagen disease.

Correspondence to:
Anthony M. Padgett, apadget7@uthsc.edu

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Introduction

Acromioclavicular joint (ACJ) dislocation is a shoulder injury that has a higher predilection in men than in women [1]. Usually, this type of injury occurs secondary to trauma. However, nontraumatic or voluntary ACJ dislocations can rarely occur [2]. Furthermore, there has only been one published case study pertaining to this rare event occurring in both shoulders. In 1984, Richards et al. [3] published a case report about a 14-year-old male patient who presented with bilateral AC joint dislocations. His patient had a history of seizures but no history of Ehlers-Danlos syndrome (EDS) or collagen disease. The patient presented in this case report sustained nontraumatic bilateral ACJ subluxations with a pertinent history of EDS.

EDS is a rare multisystemic connective tissue disease with at least 13 subtypes [1]. The most common subtype of EDS is the hypermobile subtype, which is also known as EDS type III, and it is inherited in an autosomal dominant fashion [4]. Other subtypes of EDS include types I and IV, which are known as classical EDS and vascular EDS, respectively. Both of these subtypes are also inherited in an autosomal dominant fashion. However, less common subtypes may be inherited in an autosomal recessive manner. The underlying pathogenesis of EDS is most commonly due to mutations in the genes encoding for collagen, which leads to structurally deficient collagen [4]. The main complaints observed in patients with EDS are musculoskeletal pains, joint instability, bursitis, and tendonitis [5, 6]. While orthopedic ailments might be the chief complaint in a patient with EDS, it is important to recognize that many of these patients also suffer from neurological, gastrointestinal, and cardiovascular issues, and a multidisciplinary approach should be adopted for clinical management [7]. Nonoperative management remains the mainstay of treatment for EDS patients who present with joint pain and should be exhausted prior to recommending surgical interventions [8]. To date, literature regarding the management of ACJ dislocations and subluxations in patients with EDS is missing.

A comprehensive literature search was performed to identify all reported cases of bilateral and atraumatic ACJ dislocations. The propensity for ACJ dislocations or subluxations and hypermobility in patients with EDS was also thoroughly investigated through a literature search. All relevant studies in the Cochrane Library, Medline/PubMed, and EMBASE (Elsevier) were searched from inception to January 18, 2022, using medical subject headings and text words without limitations on language or study type.

Case Presentation

The patient is a 15-year-old male with a history of EDS who presented with complaints of right-sided shoulder pain, popping, and tenderness that had been worsening for months. The patient was active in high school and club soccer. Radiographs revealed normal joint spaces without any fractures or dislocations but did reveal subluxation of the right ACJ; dynamic views with the arm at different degrees of abduction followed during this office visit (shown in Fig. 1a, b).

On physical exam, the patient was able to dislocate and reduce his shoulder with active rotation. The patient also had winged scapula, pectus excavatum, and hypermobility in multiple joints. The patient displayed no neurological abnormalities.

The patient had undergone a 3-month trial of conservative management including physical therapy without improvement. A surgical discussion was had with the patient and the family and we decided to proceed with an open mini-Mumford procedure with right ACJ reconstruction because of the debilitating pain in a young active patient. The patient...
Fig. 1. a Grashey view of the right shoulder at the first office visit. X-ray shows subluxation of the AC joint with the distal clavicle migrated superiorly. No other acute fractures or dislocations were noted. b AP X-ray of right shoulder with arm abducted at 90° on the first office visit. X-ray shows superior subluxation of the acromion at the AC joint with abduction of the shoulder. c Grashey view of the left shoulder showing subluxation of AC joint with superior migration of the distal clavicle.
underwent the surgery successfully without any complications. At a 6-week postoperative visit, the patient complained of mild right pericapsular shoulder pain that started after playing soccer again. The ACJ felt stable with no subluxation. Radiographs showed adequate anatomic alignment and expected postoperative changes at the distal clavicle (shown in Fig. 2a, b). At about 3 months postoperatively, he returned to the office and stated that he no longer had any painful popping or subluxation of this shoulder.

Nine months later, the same patient (who is now 16 years of age) presented to the clinic with complaints of pain and instability in his left ACJ. He reported constant pain lasting all day and endorsed the same painful popping sensation that was present on the right side. Radiographs of the left shoulder revealed similar findings of subluxation in the ACJ (shown in Fig. 1c). Following 3 months of failed nonoperative management, we proceeded with an open mini-Mumford and left-sided ACJ reconstruction. The surgery was successful without any intraoperative complications. The patient returned to the clinic 3 weeks later and displayed

Fig. 2. a Postoperative Grashey view of right shoulder showing expected changes of the distal clavicle after Mumford procedure with good alignment of AC joint. b Postoperative Scapular-Y view of right shoulder showing appropriate alignment of AC joint with no other noted dislocations or subluxations.
full range of motion and normal motor strength in the left upper extremity without any painful popping or subluxation of the left ACJ. At 1 and 2 years from his left and right Mumford procedures, respectively, the patient was seen in office and had no complaints of shoulder pain or restrictions to his activities.

**Discussion**

The incidence of bilateral AC joint dislocation is rare, with only one report published in the literature. Our patient’s atraumatic subluxations were likely secondary to his inherited collagen-deficient disease. Patients with EDS are more susceptible to axial skeletal pathologies such as cervical instability and lumbar spondylosis, in addition to dislocation and subluxation of joints such as the hip and patella. As a result, they are also susceptible to peripheral nerve injury and chronic pain [7]. More research needs to be conducted to weigh the risks and benefits of prophylactically performing stabilizing procedures on patients with EDS.

The Mumford procedure is a distal clavicular resection that can be performed either openly, as in this patient, or arthroscopically. Indications for the Mumford procedure include Rockwood grade I or II unreduced ACJ dislocations in which the coracoclavicular ligaments are intact [9]. The technique involves the resection of approximately 2.5 cm of the lateral end of the clavicle. The superior margin of the remaining lateral clavicle is smoothed with a file to reduce any sharp bony ridge beneath the epidermis. Postoperative complications can include ACJ stiffness, heterotopic ossification, and infection [10]. Overresection of the distal clavicle can lead to ACJ instability [9, 10].

Joint dislocations and joint hyperlaxity are often the initial presenting symptoms of patients affected by EDS. Nourissat et al. [11] demonstrated that painless atraumatic glenohumeral dislocation with pain following reduction in a young patient can suggest EDS. Similarly, the patient in this case report presented with a Grade I ACJ injury (according to the Rockwood classification) with subluxation and associated progressively increasing painful range of motion [12]. The patient did not have arthritis in the AC joint on initial radiographs but had difficulty maintaining his ACJ reduced when he initially presented to the office. For this reason, we decided to proceed with the distal clavicle excision in addition to ACJ reconstruction as we believed it would alleviate some of the painful clicking symptoms the patient was experiencing. The authors acknowledge that there may be other surgical options for this type of pathology, such as an ACJ reconstruction without bone resection, although none have been described specifically for EDS.

Because musculoskeletal symptoms are often the first manifestations of EDS, orthopedic surgeons are frequently the first contact for such patients. Providers should maintain a high index of suspicion for collagen disease in patients with atraumatic joint dislocations and subluxations, especially when they are recurrent or affect multiple joints. Because it is a multisystemic disorder, orthopedic surgeons can help coordinate the appropriate multidisciplinary care for patients with suspected EDS. Gastrointestinal symptoms include both mild dysfunction of the gastrointestinal tract as well as more severe complications such as hernias and perforation [13]. While cardiovascular complications are primarily associated with the vascular subtype of EDS, they can include life-threatening phenomena such as arterial dissections and aneurysmal ruptures [14]. Throughout the intervention of our patient’s AC joint abnormalities, we consulted the expertise of a geneticist, a cardiologist, and a pediatric surgeon to evaluate the patient’s nonorthopedic ailments. This multisystemic approach will hopefully increase his long-term quality of life.
Statement of Ethics

This retrospective review of patient data did not require ethical approval in accordance with national guidelines. Written informed consent was obtained from the parent/legal guardian of the patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors declare that we have no conflicts of interest.

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Author Contributions

Luis F. Colón was an associate provider in the patient’s clinical course, performed literature review, and critically revised the manuscript. Anthony M. Padgett performed literature review and critically revised the manuscript. Charles Powell critically reviewed the manuscript for important intellectual content and revised the manuscript. Dillon Morrow critically reviewed the manuscript for important intellectual content and revised the manuscript. Jeremy R. Bruce was the primary provider throughout the patient’s clinical course, coordinated and supervised literature review, and critically reviewed the manuscript for important intellectual content.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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