Case Report

A Proximal Humerus Fracture Dislocation in a Patient with Stiff Person Syndrome

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Keywords

Stiff person syndrome · Shoulder dislocation · Orthopedics · Fracture dislocation · Shoulder arthroplasty

Abstract

We present a case of a 47-year-old female who presented with an atraumatic posterior proximal humerus fracture dislocation secondary to episodic spasms, later diagnosed to be caused by stiff person syndrome (SPS). She underwent a shoulder hemiarthroplasty as well as a subsequent revision for recurrent dislocation and instability. SPS is a challenging diagnosis and we recommend optimization of medical management prior to surgical intervention.

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Introduction

Posterior shoulder dislocations are relatively rare and are usually a result of high-energy trauma, electrocution, or seizure. Dislocations caused by seizures are thought to be related to a muscular imbalance where the pectoralis major and latissimus dorsi overpower the external rotators. The humeral head displaces superiorly and posteriorly against the acromion and medially towards the glenoid, ultimately resulting in a posterior dislocation [1, 2]. We present a

case of a 47-year-old female who sustained a posterior shoulder dislocation associated with a comminuted fracture of the humeral articular surface. Her only presenting symptoms were pain and spasms of her right arm. She was later diagnosed with stiff person syndrome (SPS).

SPS is an autoimmune neuromuscular disorder characterized by insidious onset of muscular rigidity and uncontrolled spasms, which predominately affect the proximal and axial musculature. It is a rare disorder which typically presents in the fifth decade of life and affects less than 1 person per million, predominately females. Antibodies to glutamic acid decarboxylase and amphiphysin are characteristic of the disease and may aid in diagnosis [3–5]. SPS may present as a paraneoplastic syndrome [6, 7]. Medical treatment varies, but outcomes are generally poor.

Orthopedic manifestations of SPS are rare. A review of the literature resulted on only one brief case report of a woman who sustained bilateral hip fractures from spasms secondary to SPS [8]. The present case was particularly challenging as a delay in diagnosis and lack of effective medical therapies led to recurrent instability.

Case

A 47-year-old female who was admitted to the hospital complained of new-onset arm pain secondary to spasms. Imaging revealed a posterior shoulder dislocation with significant impaction and comminution of the humeral head (Fig. 1).

Prior to this injury, the patient had been admitted to the hospital for lower extremity weakness, which was initially treated as Guillain-Barré syndrome. Plasmapheresis and IVIG resulted in minimal improvement and she began to have upper extremity spasms. The patient underwent a neurological workup including an EEG to rule out seizure as the cause of her dislocation. Given the age of the patient and the degree of comminution of the articular surface, the patient was initially treated with an uncemented hemiarthroplasty. The rotator cuff appeared to be intact but glenohumeral ligaments were found to be disrupted and primarily repaired. Unfortunately, 4 weeks postoperatively, the patient reported continued spasms which led to an anterior dislocation of her prosthesis (Fig. 2). The patient was treated with a revision cemented hemiarthroplasty and repair of the greater and lesser tuberosity. Recurrent instability occurred due to continued spasms resulting in dislocation of the prosthesis (Fig. 3).

A further workup revealed positive anti-amphiphysin antibodies consistent with SPS. As the anti-amphiphysin antibody is related to paraneoplastic syndromes, she had an oncological workup which has been negative. Since her diagnosis, she has been treated with various medications and therapies including steroids, plasmapheresis, IVIG, and rituximab. She has had an improvement in her upper extremity spasms with continued lower extremity weakness. Presently, the patient's symptoms are manageable, and a reverse total shoulder arthroplasty (RTSA) has been considered dependent upon improvement of her lower extremity weakness.

Discussion

SPS is an extremely rare neurological disorder characterized by axial and proximal limb muscle spasms and rigidity. Though the exact pathophysiology is unknown, it is thought to be an autoimmune disorder. Approximately 60% of patients with SPS have antibodies to glutamic acid decarboxylase [3]. A subset of individuals have antibodies to amphiphysin. SPS

related to anti-amphiphysin antibodies has an association with paraneoplastic syndromes and breast cancer [3, 4, 9].

SPS is rarely seen in the orthopedic literature. It presents a challenge to the surgeon because the underlying problem persists after treatment of the orthopedic manifestation. Implants are likely to come under increased stress due to persistent spasms, which theoretically places them at a high risk of repeated fracture or instability. Medical treatments for SPS are poor and include IVIG, plasmapheresis, corticosteroids, baclofen, benzodiazepines, antiepileptics, dantrolene, and rituximab [3, 4].

Our patient not only had a spastic disorder but was also very young, 47 at the time of presentation. Therefore, the risk of dislocation and need for revision surgery was particularly worrisome. We considered the possibility of revising to an RTSA, which has been shown to be an option in patients at a higher risk of dislocating such as spastic paraplegics and patients with Parkinson's [10, 11]. While RTSA can relieve pain and provide glenohumeral stability, these patients generally have high complication rates and less predictable improvements in range of motion [11, 12]. Salvage procedures such as a fusion of the glenohumeral joint have shown good outcomes in patients with spasticity and recurrent dislocations [13, 14]. We recommend clinicians to engage patients in a thorough discussion detailing the complexity of their condition, increased risk for infection, and possibility of multiple revision surgeries including a glenohumeral fusion.

This case illustrates the difficulty of managing orthopedic manifestations of SPS. If medical treatment fails, then the orthopedic implant will continue to experience the repetitive spasms which led to the initial injury. Though rare, SPS should be considered in the differential diagnosis of patients with symptoms such as uncontrollable muscle spasms. Based on our experience, we would recommend an attempt at optimal medical management prior to any surgical intervention. During the surgical procedure, implants should be selected for maximal stability or the use of robust implants in the case of fracture. Clinicians and patients should be prepared for the possibility of revision surgeries including a glenohumeral fusion.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and associated images.

Disclosure Statement

The authors have no conflicts of interest to disclose.

References

- 1 Goudie EB, Murray IR, Robinson CM. Instability of the shoulder following seizures. *J Bone Joint Surg Br.* 2012 Jun;94(6):721–8.
- 2 Rouleau DM, Hebert-Davies J, Robinson CM. Acute traumatic posterior shoulder dislocation. *J Am Acad Orthop Surg.* 2014 Mar;22(3):145–52.
- 3 Ciccoto G, Blaya M, Kelley RE. Stiff person syndrome. *Neurol Clin.* 2013 Feb;31(1):319–28.
- 4 Baizabal-Carvallo JF, Jankovic J. Stiff-person syndrome: insights into a complex autoimmune disorder. *J Neurol Neurosurg Psychiatry.* 2015 Aug;86(8):840–8.

- 5 Gallien P, Durufle A, Petrilli S, Verin M, Brissot R, Robineau S. Atypical low back pain: stiff-person syndrome. *Joint Bone Spine*. 2002 Mar;69(2):218–21.
- 6 Sarva H, Deik A, Ullah A, Severt WL. Clinical Spectrum of Stiff Person Syndrome: A Review of Recent Reports. *Tremor Other Hyperkinet Mov (N Y)*. 2016 Mar;6:340.
- 7 Esplin NE, Stelzer JW, Legare TB, Ali SK. Difficult to Treat Focal, Stiff Person Syndrome of the Left Upper Extremity. *Case Rep Neurol Med*. 2017;2017:2580620.
- 8 Podobinski TK, Mimbella PC, Irvine MJ, Verduzco-Gutierrez M. Poster 237 Bilateral Hip Fracture During Hospitalization for Spasm Exacerbation in an Adult with Stiff Person Syndrome: A Case Report. *PM R*. 2016 Sep;8(9 9S):S237–8.
- 9 Murinson BB, Guarnaccia JB. Stiff-person syndrome with amphiphysin antibodies: distinctive features of a rare disease. *Neurology*. 2008 Dec;71(24):1955–8.
- 10 Skedros JG, Smith JS, Langston TD, Adondakis MG. Reverse Total Shoulder Arthroplasty as Treatment for Rotator Cuff-Tear Arthropathy and Shoulder Dislocations in an Elderly Male with Parkinson's Disease. *Case Rep Orthop*. 2017;2017:5051987.
- 11 Hattrup SJ, Cofield RH. Shoulder arthroplasty in the paraplegic patient. *J Shoulder Elbow Surg*. 2010 Apr;19(3):434–8.
- 12 Cusick MC, Otto RJ, Clark RE, Frankle MA. Outcome of reverse shoulder arthroplasty for patients with Parkinson's disease: a matched cohort study. *Orthopedics*. 2017 Jul;40(4):e675–80.
- 13 Thangarajah T, Alexander S, Bayley I, Lambert SM. Glenohumeral arthrodesis for the treatment of recurrent shoulder instability in epileptic patients. *Bone Joint J*. 2014 Nov;96-B(11):1525–9.
- 14 Diaz JA, Cohen SB, Warren RF, Craig EV, Allen AA. Arthrodesis as a salvage procedure for recurrent instability of the shoulder. *J Shoulder Elbow Surg*. 2003 May-Jun;12(3):237–41.

Fig. 1. **a** AP radiograph of shoulder. **b** Axillary radiograph of shoulder demonstrating a comminuted posterior shoulder fracture dislocation. **c** Axial CT scan demonstrating significant comminution of the humeral head extending to the articular surface. **d** Coronal CT scan further characterizing the articular comminution.

Fig. 2. Radiographs taken approximately 2 weeks (a) and 1 month (b) postoperatively. One-month follow-up radiographs demonstrate anterior prosthetic dislocation with associated displaced greater and lesser tuberosity fractures as well as an associated coracoid fracture.

Fig. 3. AP radiographs of shoulder 1 day postoperatively (a) and at follow-up (b), the latter demonstrating a dislocated prosthesis.