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Case Report

Arthroscopic Glenoid Labrum Repair of Left Shoulder in a Male With Ehlers-Danlos Syndrome

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Ehlers-Danlos syndrome (EDS) is an inherited connective tissue disorder that has been classified into several primary types. The clinical characteristic of EDS is hypermobility of the joints, hyperextensibility and fragility of the skin, and hemorrhage tendency. For many patients, the hypermobile joints become a serious problem. We present the case of a 19-year-old male diagnosed with EDS, with recurrent dislocations of his left shoulder, hyperextensibility and fragility of the skin, and a carp-mouth–shaped scar of the forearm. After 4 years of nonoperative treatment, we performed an arthroscopic glenoid labrum repair of left shoulder. At a 6-year follow-up, the patient has no instability in the left shoulder. We believe that glenoid labrum repair is a viable method for treating recurrent dislocations for patients with EDS. It is strongly suggested to check coagulation function of patients to avoid substantial bleeding when decorticating the glenoid rim, to ensure a conservative postoperative rehabilitation.

Key words: Ehlers-Danlos syndrome – Arthroscopy – Glenohumeral joint – Glenoid Labrum repair

A 19-year-old male was admitted to our department of sports medicine on May 9, 2006, with the chief complaint of "recurrent dislocations of his left shoulder for 4 years." The patient was diagnosed with Ehlers-Danlos syndrome (EDS), characterized by recurrent dislocations of his left shoulder, hyperextensibility and fragility of the skin, and a carp-mouth–shaped scar of the forearm. He report-

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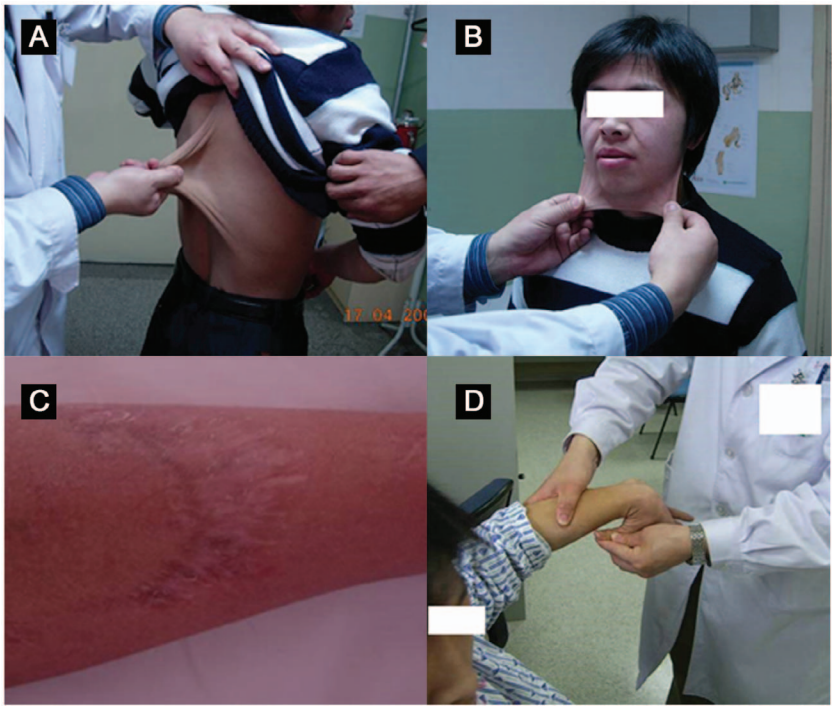

Fig. 1 Body examination shows: (A) hyperextensibility of the dorsum skin, (B) hyperextensibility of the facial skin, (C) a wide carp-mouth–shaped scar on the left forearm and (D) hypermobility of the joints. The patient had demonstrable laxity of his MCP joints and could oppose his thumbs to his forearms.

ed history of shoulder dislocations starting at the age of 15 years. Initially, the left shoulder dislocation was provoked during a badminton smash, which was reset by one of his friends. The patient resumed normal activity after 3 days. However, within the following 4 years, the dislocations reoccurred when pursuing low-energy overhead activities, such as taking off his clothes and arranging his hair. The patient also reported that any slight injuries were often accompanied by subcutaneous hematoma, which would disappear if he bandaged the wound with pressure. The patient's parents were not consanguineous mates, and there was no similar symptom in his family.

Examination showed a healthy-appearing male with normal stature and facies. Body examination showed hyperextensibility of the dorsum and facial skin, presenting a velvet-like appearance (Figs. 1A and 1B). A carp-mouth–shaped scar was noted in the left forearm (Fig. 1C). There was slight muscle atrophy in both shoulders. Focal tenderness was noted in the posterior area of both shoulders and the greater tuberosity area of the left shoulder. An instability examination was significant for a 2+ anterior and posterior drawer test, 2+ sulcus sign, 3+ sulcus in external rotation, and apprehension with anterior and posterior stressing; both elbows and knees of the patient had recurvatum of 5° to 10°. He could also oppose his thumbs to his forearms (Fig. 1D).

Laboratory examination showed that his coagulation function was normal. Plain radiographs of his shoulders revealed normal bony anatomy without dislocations or bony lesions. Magnetic resonance imaging (MRI) contrast showed anterior-inferior glenoid labrum injury and an inferior capacious capsular (Fig. 2A). On May 12, 2006, the patient underwent a left shoulder arthroscopy with anterior glenoid labrum repair. The examination under anesthesia was consistent with preoperative findings. Visualization under arthroscopy showed a grossly normal biceps tendon; rotator cuff; superior, middle, inferior glenohumeral ligament; and anterior capsular—and a ruptured labrum and an inferior capacious capsule. The labrum was separated with the glenoid from 11 to 5 o'clock, but the periosteum of the region was intact. We implanted 4 anchors (G2) in the 12, 2, 3, and 5 o'clock positions of the glenoid, to fix the glenoid labrum (Fig. 2B). The postoperative instability examination was negative for an anterior and posterior drawer test, sulcus sign, and sulcus in external rotation. The patient was then placed in a left shoulder joint orthosis for 2 months. When the patient got out of the orthosis within the 2 months, the patient began physical therapy on his left

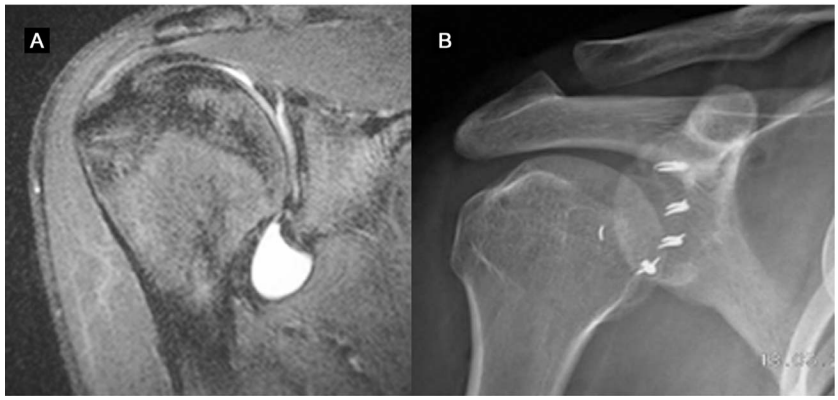

Fig. 2 Pre- and postoperative images of the patient. (A) MRI contrast shows anterior-inferior glenoid labrum injury and an inferior capacious and patulous capsular. (B) Postoperative plain film of the left shoulder.

shoulder. The patient did range of motion and strengthening exercises for the rotator cuff, deltoid muscles, except for excess abduction and external rotation movement.

Until the final follow-up in 2012, 6 years after the surgery, active and passive ranges of motion for his shoulders were normal. There was no apprehension with anterior and posterior stressing. The patient was actively involved in badminton, swimming, and basketball. He has had no dislocations in his left shoulder, and he is pleased with his current level of functioning.

Discussion

EDS is a group of variable clinical manifestations including hypermobility of the joints, hyperextensibility, fragility of the skin, poor healing, and bleeding tendency.[1] The description of EDS was independently reported by Tschernogobow,[2] Ehlers,[3] and Danlos[4] in medical literature. The eponym of Ehlers-Danlos syndrome was first suggested by Poumeau-Delille and Soulie in 1934.[5] EDS is an inherited disorder estimated to occur in about 1 in 5000 births worldwide.[6]

EDS typically affects the joints, skin, and blood vessels. EDS may have the following symptoms: hyperflexible joints[7]; unstable joints that are prone to sprain, dislocation, subluxation, and hyperextension[8]; early onset of advanced osteoarthritis[9]; chronic degenerative joint disease[9]; tearing of tendons or muscles[10]; deformities of the spine such as scoliosis, kyphosis, tethered spinal cord syndrome, occipitoatlantoaxial hypermobility[11]; muscle pain and joint pain[12]; congenital deformity or dislocation[13]; fragile skin that tears easily[9]; easy bruising[7]; and redundant skin folds.[9] In the cardiovascular system, EDS may cause the following symptoms: easy arterial rupture,[7] vascular heart disease,[14] congenital heart disease,[14] and dilation and/or rupture of ascending aorta.[15] EDS may also have other manifestations or complications like hiatal hernia and anal prolapse.[16]

The case we reported had the following signs and symptoms that supported an EDS diagnosis: (1) recurrent joint dislocation, because of the hyperextensibility of the skin and hypermobility of the joints; (2) skin that tears easily caused a wide carp-mouth–shaped scar on the left forearm; (3) frequently occurring subcutaneous hematomas with any slight injury in each joint, because the arterioles can be ruptured easily.

Unlike patients with traumatic shoulder instability, patients with hyperlaxity like EDS are more likely to experience episodes of recurrent subluxation than they are to have recurrent dislocation.[17] In the case reported here, the patient didn't try to get clinical intervention before the unexpected injury caused by the badminton smash at the age of 15 years. The patient is more likely to have instability because of the soft-tissue and osseous lesions associated with the trauma. Therefore, we should first deal with these soft-tissue lesions to treat the dislocation.

It is noteworthy that the bleeding tendency in EDS that caused the subcutaneous hematoma is not the same as coagulation disorders. The coagulation function of patients with coagulation disorders is abnormal. Moreover, patients with coagulation disorders always have family members with the same disorder. Despite having similar clinical manifestations to coagulation disorders, the coagulation function in EDS patients is normal; also, patients with EDS don't have members with the same bleeding symptoms in their families.

The chief complaint of the patient in this case was "recurrent dislocations of his left shoulder for 4 years." Visualization under arthroscopy showed a grossly normal biceps tendon, rotator cuff; superior, middle, and inferior glenohumeral ligament; and anterior capsular—and a ruptured labrum and an inferior capacious capsular. The glenoid labrum injury may be caused by recurrent dislocations of the humeral head. After a comprehensive assessment, we decided only to perform the glenoid labrum repair for the patient, because of the normal anterior capsular and glenohumeral ligaments. We made the following observations:

1. This case was a traumatic injury caused by a badminton smash at the age of 15 years. Before this unexpected injury, the patient never complained of any glenoid-labrum joint dislocation, even when the patient was conducting movements or postures that now make the patient feel apprehension. Moreover, the patient did not have a long history of atraumatic shoulder instability before this unexpected injury at the age of 15 years.
2. When we performed the labrum repair, we observed that the labrum was separated with the glenoid from 11 to 5 o'clock. Although there was some soft-tissue retraction, labrum tension was good enough to pull back to its original location. We didn't see any bone loss in the glenoid, so we assumed that the prospective outcome would be satisfactory if we conducted the labral repair procedure only. Currently, the patient has more than 20% bone loss to the glenoid. We performed the Latarjet procedure without any capsular reconstruction.

We suggested glenoid labrum repair as a viable method for treating recurrent dislocations in patients with EDS, when capsular ligaments were also very stretched out. First, we should check the coagulation function of the patient to rule out coagulation disorders and avoid substantial hemorrhage when decorticating the bone surface. If the anterior capsular are all abnormal, thermal capsulorrhaphy or reefing of joint capsule procedures should be performed. The arthroscopic procedure may need to be changed from traditional (open) surgery if hemorrhage cannot be controlled. Second, given that the collagen fibers of EDS patients are easily ruptured and hard to repair, the humeral head is highly prone to redislocation during the rehabilitation period. Therefore, postoperative rehabilitation should be conservative; rehabilitation of the fixed glenoid labrum will take longer in EDS patients than in patients without EDS. Furthermore, because patients with EDS have a serious risk of hemorrhage, we should ask patients to avoid trauma in the future.

Acknowledgments

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