Recurrent Anterior Shoulder Dislocation Before Epiphyseal Closure in a Patient with Shprintzen-Goldberg Syndrome

Naoya Taki[1], Hideyuki Sasanuma[1], Yuki Iijima[1], Yuji Kanaya[1], Hideaki Watanabe[2], Katsushi Takeshita[1]

[1]Department of Orthopedic Surgery, Jichi Medical University

[2]Department of Pediatric Orthopedic Surgery, Jichi Children's Medical Center

Abstract：This is a case of recurrent anterior shoulder dislocation before epiphyseal closure in a patient with Shprintzen-Goldberg syndrome (SGS), a subtype of Marfan syndrome. The patient was a 13-year-old boy. He was diagnosed with SGS at the age of 4 years, and had experienced more than 10 shoulder dislocations. Radiographs showed an open proximal humeral growth plate, without a clear Hill-Sachs lesion or fracture of the glenoid rim. Preoperative shoulder magnetic resonance arthrography revealed signs characteristic of capsular rupture. Arthroscopy showed complete absence of the middle glenohumeral ligament, which left the subscapularis uncovered. We performed arthroscopic capsular reconstruction by elevating the anterior inferior glenohumeral ligament and suturing it to the superior glenohumeral ligament, thereby reducing capsule volume. At the 24-month follow-up, the patient was satisfied with the results; he maintained his regular schedule of leisure sports and football without pain, limitations, or new episodes of instability. This report shows that arthroscopic stabilization of recurrent shoulder dislocation in a patient with open physes and a Marfan-related disorder provided good functional outcomes without recurrence of instability. As the patient had risk factors including age (under 20 years) at the time of surgery and shoulder hyperlaxity, there is a need for long-term follow-up.

Introduction

Shprintzen-Goldberg syndrome (SGS), which is characterized by craniosynostosis and Marfanoid habitus, was first reported by Shprintzen and Goldberg[9] in 1982. Mutations in exon 1 of SKI have recently been identified as being responsible for approximately 90% of reported individuals diagnosed clinically with SGS[1]. To date, approximately 60 cases of SGS have been reported[7]. The clinical features of SGS are neurological, cardiovascular, connective tissue, and skeletal abnormalities, including some traits also found in Marfan syndrome[8]. The skeletal features of SGS include such as tall stature, arachnodactyly, foot deformity, developmental scoliosis, and joint hypermobility[7]. With joint hyperlaxity and hypermobility characterize several inherited disorders or occur because of local or generalized causes, some patients suffer actu-

Key words：arthroscopy, growth plate, recurrent dislocation, glenohumeral ligament, hyper laxity
Corresponding author：Naoya Taki
Department of Orthopaedic Surgery, Jichi Medical University, 3311-1 Yakushiji, Shimotuke-shi, Tochigi 329-0498, Japan
TEL: +81-285-44-2111
e-mail：naoya.taki.11.2@gmail.com

al clinical dislocation episodes either from injury or excessive normal use[6]. There have been no reports regarding macroscopic alterations due to recurrent dislocations in young patients with SGS. We present the case of an adolescent patient with SGS who underwent arthroscopic surgery for chronic recurrent shoulder dislocations.

Case Presentation

A 13-year-old right-side dominant boy with SGS was referred because of recurrent left shoulder dislocations. He was diagnosed with SGS at the age of 4 years, and the first episode of shoulder dislocation was at the age of 10 when diving into a swimming pool. After that, he had experienced more than 10 anteroinferior dislocations following minor trauma, in abduction and external rotation of the left glenohumeral joint (Fig.1). At each episode, sling immobilization in abduction and external rotation position (ABER position) was performed for 3 weeks after closed reduction, followed by rehabilitation, which yielded unsatisfactory results. He also had bilateral genu recurvatum and recurrent ankle sprains. However, the Beighton score for generalized joint laxity was only 2 of 9[6].

Initially, both shoulders showed similar range of motion : 170° flexion, 165° abduction, T1 vertebral level internal rotation, and 65° external rotation at the side. The left shoulder showed a positive sulcus sign and positive anterior apprehension test at 0–150° abduction; however, posterior or inferior apprehension was absent, no signs of multidirectional instability.

Radiographs (Fig.1) showed an open proximal humeral growth plate without a clear Hill–Sachs lesion or fracture of the glenoid rim. Preoperative magnetic resonance arthrography showed leakage of contrast medium outside of the axillary pouch, suggesting capsular rupture[3] (Fig.2).

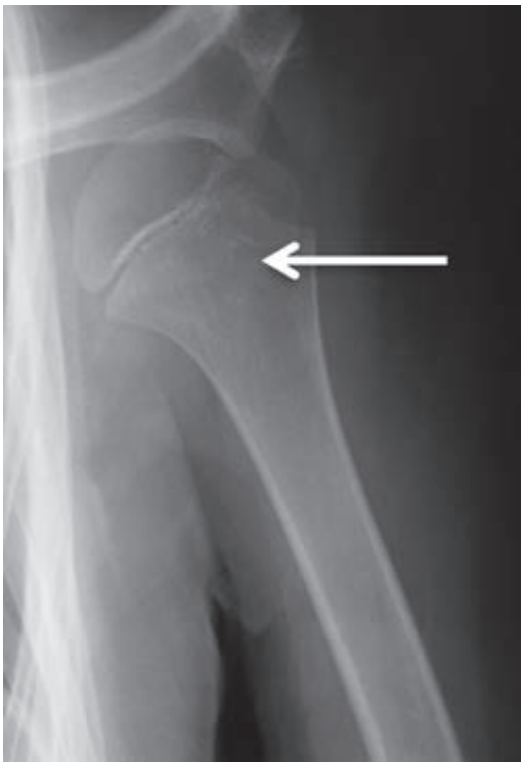

Fig. 1. Radiograph of anteroinferior dislocation before epiphyseal closure Proximal humeral physis (white arrow)

Considering the frequency of dislocation and the patient's desire, arthroscopic stabilization of chronic anterior shoulder dislocation was scheduled. An interscalene nerve block was followed by general anesthesia and placement in a beach chair position. Initial intra-articular exploration through a posterior portal showed complete absence of the mid-anterior capsule, leaving the subscapularis muscle uncovered (Fig.3) ; the inferior insertion of the labrum and anterior inferior glenohumeral ligament (AIGHL) appeared normal. No abnormal lesions were found on the superior labrum, rotator cuff, or chondral surfaces; the superior glenohumeral ligament (SGHL) and biceps tendon were in place. A sec-

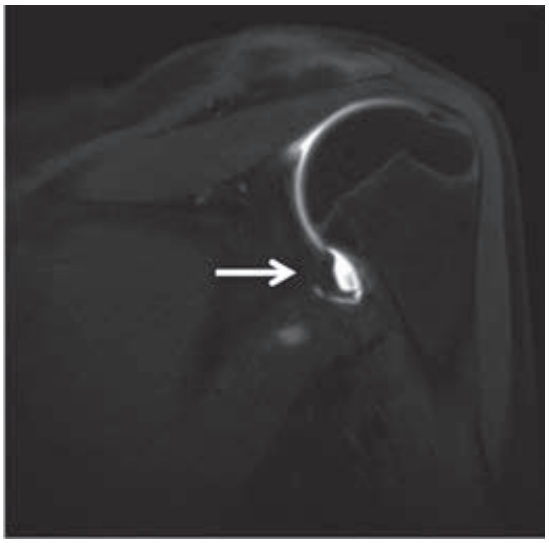

Fig. 2. Leakage of contrast medium outside of the axillary pouch, indicating capsular rupture (white arrow)

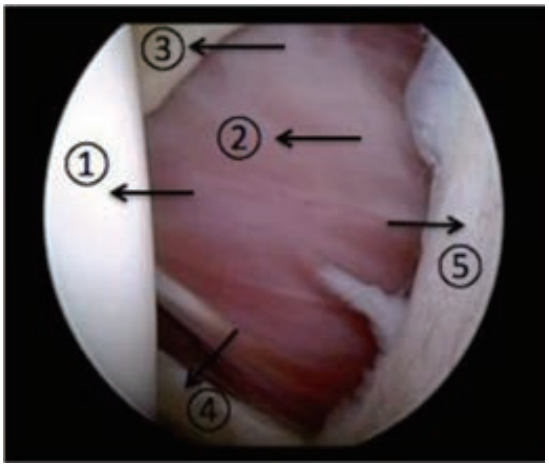

Fig. 3. Arthroscopic posterior portal view : Subscapularis muscle was exposed because of complete absence of the mid-anterior capsule
① Humeral head ② Subscapularis muscle ③ SGHL ④ AIGHL ⑤ Glenoid

ond anterior approach involved an inside-out technique. The operation was performed according to a procedure reported by Gomes et al[5]. Dissection of the AIGHL complex from the glenoid rim and subscapularis muscle was performed with a radiofrequency wand to prepare the capsule. First, the AIGHL complex was pulled up to the anterosuperior part of the glenoid and resutured with four bioabsorbable anchors. Second, the mid-anterior capsule was reconstructed by suturing the SGHL and AIGHL together (2 sutures, No. 2 strong suture), and closure of the large defect (Fig.4). Postoperative rest in a sling was prescribed for 1 month, followed by physiotherapy. Six months later, recreational sports were resumed. At the 24-month follow-up, the mobility of both shoulders was as follows : 180°/170° flexion, 180°/160° abduction, T1/T8 vertebral level internal rotation, and 65°/60° external rotation. Although slight difference of range of motion between both shoulders existed, he continued leisure sport activities and football without pain, limitations, or new episodes of instability.

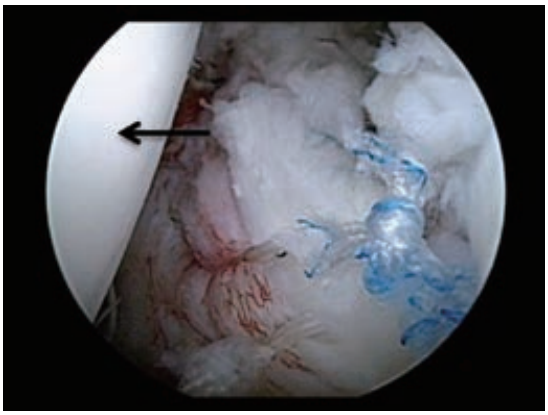

Fig. 4. Capsular reconstruction by suturing SGHL and AIGHL together Final aspect of reconstruction Humeral head (black arrow)

Discussion

This report showed that arthroscopic stabilization of recurrent anterior shoulder dislocation in a patient with open physes and a Marfan-related disorder provided good functional outcomes without recurrence of instability.

Gomes et al. reported arthroscopic treatment of an adult with Marfan syndrome and recurrent anterior shoulder dislocation with complete absence of the mid-anterior capsule with the

subscapularis uncovered[5]. Capsular reconstruction through anteroinferior dissection from the glenoid rim, followed by superior drift of the capsule was performed. As the intraarticular findings were similar to ours, we selected their procedure for treatment. While both of them are just as good postoperative course without pain, limitations, or new episodes of instability, our patient is much younger and carefully follow-up is essential.

Gigante et al. reported remarkably fewer elastic fibers in the knee capsules of three Marfan syndrome patients compared with control tissues obtained from three adolescents who had no stigmata of Marfan syndrome at the time of death[4]. Ultrastructurally, they appeared fragmented and indented, indicating discontinuous elastin aggregates among randomly dispersed filaments. Therefore, we hypothesized that absence of the mid-anterior capsule after repetitive trauma may be attributed to failure of normal remodeling because of lack of competent elastic fibers.

The following risk factors of recurrent instability have been identified: age under 20 years at the time of surgery and shoulder hyperlaxity[2]. Arthroscopic Bankart repair has a high risk of failure in athletic adolescents compared with the results in adults[10]. Long-term follow-up is needed because our patient has risk factors for recurrent shoulder instability.

REFERENCES

1) Au PY, Racher HE, Graham JM Jr et al : De novo exon 1 missense mutations of SKI and Shprintzen-Goldberg syndrome : two new cases and a clinical review. Am J Med Genet A **164A** : 676-684, 2014.
2) Balg F, Boileau P : The instability severity index score. A simple pre-operative score to select patients for arthroscopic or open shoulder stabilisation. J Bone Joint Surg Br **89** : 1470-1477, 2007.
3) Bui-Mansfield LT, Banks KP, Taylor DC : Humeral avulsion of the glenohumeral ligaments: the HAGL lesion. Am J Sports Med **35** : 1960-1966, 2007.
4) Gigante A, Chillemi C, Greco F : Changes of elastic fibers in musculoskeletal tissues of Marfan syndrome : A possible mechanism of joint laxity and skeletal overgrowth. J Pediatr Orthop **19** : 283-288, 1999.
5) Gomes N, Hardy P, Bauer T : Arthroscopic treatment of chronic anterior instability of the shoulder in Marfan's syndrome. Arthroscopy **23** : 110.e1-5, 2007.
6) Murray KJ : Hypermobility disorders in children and adolescents. Best Pract Res Clin Rheumatol **20** : 329-351, 2006.
7) Pagon RA, Adam MP, Ardinger HH et al (editors) : Gene Reviews [Internet], University of Washington, Seattle, 1993-2016.
8) Robinson PN, Neumann LM, Demuth S et al : Shprintzen-Goldberg Syndrome : Fourteen New Patients and a Clinical Analysis. Am J Med Genet A **135A** : 251-262, 2005.
9) Shprintzen RJ, Goldberg RB : A recurrent pattern syndrome of craniosynostosis associated with arachnodactyly and abdominal hernias. J Craniofac Genet Dev Biol **2** : 65-74, 1982.
10) Shymon SJ, Roocroft J, Edmonds EW : Traumatic anterior instability of the pediatric shoulder : A comparison of arthroscopic and open Bankart repairs. J Pediatr Orthop **35** : 1-6, 2015.