


A new form of shoulder dysplasia in an 11-year-old boy

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ABSTRACT

Pain and impaired function in the shoulder can result from a variety of conditions. Dysplasias of the shoulder girdle are usually asymptomatic; previous literature reports on shoulder malformation have focused on glenoid dysplasia or complete agenesis of the clavicle. We report the case of an 11 year old patient with severe shoulder pain after strenuous physical exercise. Physical inspection showed a prominent clavicle and anteriorly displaced humerus. X-ray demonstrated an abnormally shortened clavicle and a hypoplastic acromion; MRI imaging revealed an elongated deltoid muscle leading over the humeral head. The patient recovered with analgesics and careful mobilization. This is the first reported case of a dysplasia of both the acromion and the clavicle. This developmental abnormality can lead to significant pain, but apparently responds well to conservative treatment. It is possible that this condition is underdiagnosed or represents significantly delayed development.

CASE REPORT

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An 11-year old, right-hand-dominant boy presented to us with a 5-week history of right shoulder pain. He did not report any preceding trauma, but had previously participated in a vigorous trampoline session. Shortly thereafter, he developed increasing pain in his right shoulder over the period of a week, to the point where he was no longer able to use his arm. He had been treated with cold packs, a Gilchrist bandage and ibuprofen as needed. On physical examination, the patient was afebrile with stable medical signs. The contour of both shoulders appeared unusual, with a shortened and prominent clavicle and an anteriorly displaced humerus (Figure 1). The right shoulder was elevated with accompanying pain-induced scoliosis. However, the shoulder was not warm to the touch, with no erythema, but tender on palpation over the deltoid muscle. ROM was limited to 30° abduction with significant pain. External and internal rotation was limited to 70°. The neurovascular examination of the involved extremity was normal.

Imaging Findings

Radiographs of the shoulder were obtained and showed a bilateral, symmetrical hypoplasia of the upper part of the shoulder joint. The roof of the shoulder was formed by an abnormally shortened clavicle and a hypoplastic acromion (Figure 2 and 3). MRI imaging revealed an elongated deltoid muscle originating from the hypoplastic acromion and clavicle and leading over the humeral head (Figure 4). The glenohumeral joint was shown to be intact by MRI imaging (Figure 5). Visual inspection of the parents showed no signs of a similar abnormality, and there were no reports of other family members with similar problems.

Management & Follow-up

The patient was treated with naproxen (15 mg/kg/day p.o.) and physiotherapy with initially passive mobilization. With this treatment, he recovered quickly and after two weeks was able to initiate a program of active exercise aimed at increasing muscle mass at the shoulders.

DISCUSSION

Etiology & Demographics

The shoulder is one of the largest and most complex joints in the body, with the humerus fitting relatively loosely into the shoulder joint. This gives the shoulder a wide range of motion, but also makes it vulnerable to injury. Shoulder problems including pain are one of the most frequent reasons for musculoskeletal symptoms and consultations with primary care providers [1].

Except for Sprengel deformity, scapular dysplasias are rare developmental abnormalities, frequently overlooked and therefore commonly found incidentally on chest radiographs [2]. The exception are dysplasias in the area of the glenohumeral joints, which have been described frequently in the literature and are found in up to 35% of bone specimens in various populations [3-5]. In contrast, dysplasias of the acromion are rare [6].

Clinical & Imaging Findings

There are no previously described cases of acromioclavicular dysplasia. Comparison of the X-ray images of the patient with an age-matched control show a significantly altered anatomy of the shoulder (Figure 2). The MRI imaging demonstrates a deltoid muscle with no apparent defects traveling over the humeral head without a bony roof (Figure 4). As this was the only abnormality in this patient, we speculate that the severe pain was caused by this anomaly, either by unusual exertion of a stretched muscle beyond the physiological limit, or by friction from the relatively unprotected humeral head.

Treatment & Prognosis

Overuse of the deltoid muscle is the probable cause of the pain symptoms of the patient. Conservative treatment with non-steroidal antirheumatic drugs and careful mobilisation led to rapid improvement, supporting this theory. The patient was cautioned to prevent overuse and engage in a program of muscle strengthening of the shoulder to stabilize and protect the humeral head.

Differential Diagnoses

To our knowledge, acromioclavicular dysplasia has not been described previously in the literature. There are several reports of congenital dysplasias in the scapula, usually in the area of the glenoid [3, 4, 7, 8]. Complete, bilateral agenesis of the acromion has been reported only in two symptomatic patients, with additional, asymptomatic family members in one case [6, 9]. Both affected patients exhibited similar symptoms, with moderate shoulder pain slowly developing over time, in one case exacerbated by a traffic accident.

The scapula arises from two or more distinct embryologic origins, controlled by different genes than the upper limb [2]. The adult scapula is formed by at least eight different ossification centers [6, 7]. At birth most of the scapula is ossified, with the exception of the acromion. The acromion itself is created from several ossification centers with fusion usually in the early third decade of life [6, 9]. These three centers coalesce into a triangular epiphyseal bone, connecting

via the acromial base to the scapula [10]. Dysplasias of the acromion result from aberrant fusion or abnormal growth, such as the frequently described os acromiale [6]. It is likely that local aberrant growth of one or more ossification centers have caused the acromial dysplasia in this patient. This is supported by the bilateral symmetrical findings, suggesting an underlying genetic cause. Interestingly, in contrast to complete clavicular dysplasia, the clavicles in this case are hypoplastic rather than hypertrophic [9]. Given the young age of the patient it is possibly that this case represents a developmental variant with extremely delayed growth of the acromioclavicular structures. While development of this area can be delayed until the third decade, the extent of hypoplasia exhibited by the patient makes this unlikely.

In summary, we describe the first case of acromioclavicular dysplasia in an eleven year old male patient. This developmental abnormality does not significantly affect the shoulder function, but can apparently lead to significant pain starting in adolescence; due to the moderate symptoms, reacting well to conservative treatment at least in this case, it is possible that this condition is underdiagnosed.

TEACHING POINT

Acromioclavicular dysplasia appears to be a developmental abnormality resulting in a hypoplastic shoulder roof. This can be a cause of significant pain, but reacts well to conservative treatment.

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FIGURES



Figure 1: 11 year old boy with acromioclavicular dysplasia. A frontal photograph of the patient shows the change in the contour of both shoulders with shortened clavicae and anteriorly displaced humeral heads.

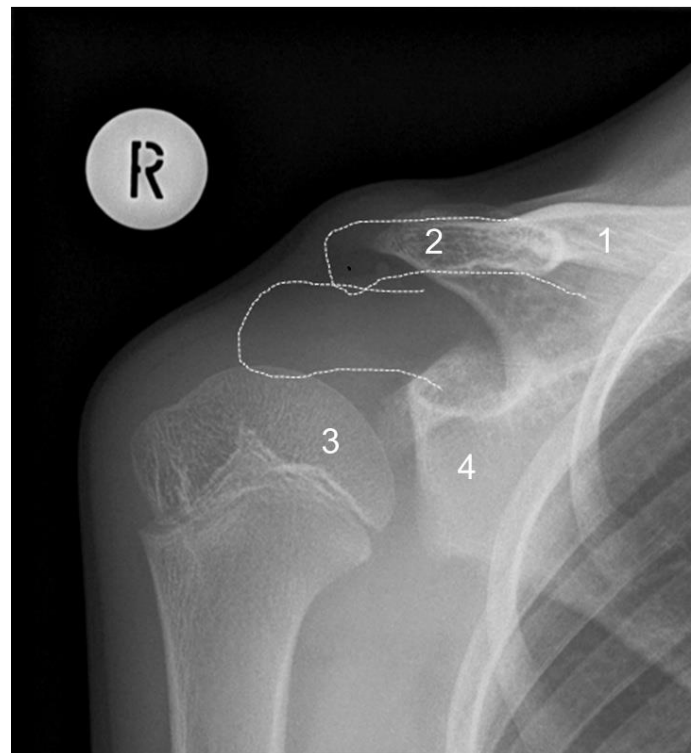


Figure 2: 11 year old boy with acromioclavicular dysplasia. Radiographs of the shoulder show hypoplasia of the clavicle (1) and acromion (2) with normal sized humeral head (3) in contact with the glenoid (4). The white line shows the outline of the lateral contours of clavicle and acromion from an age- and sex-matched healthy control.

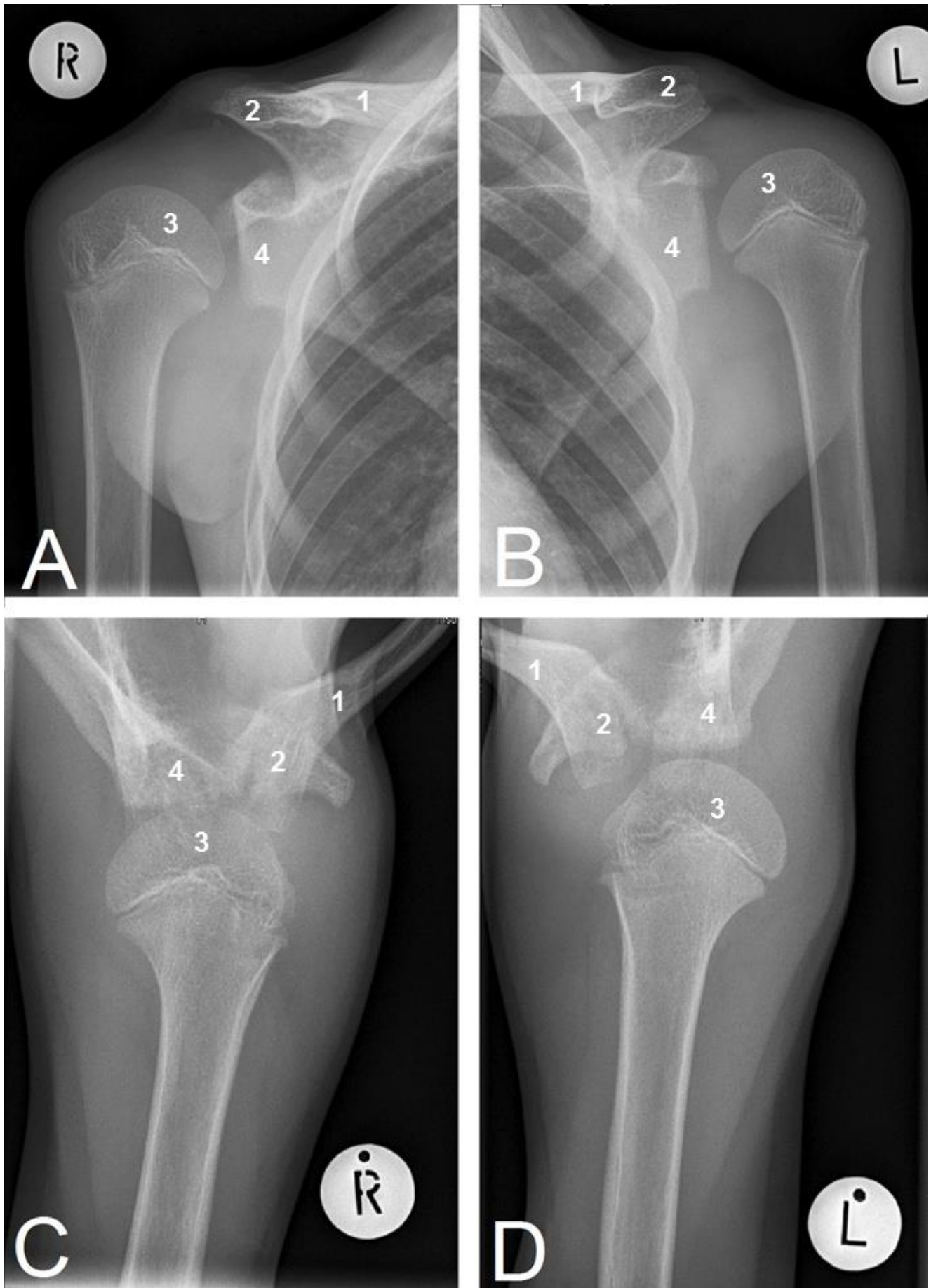


Figure 3: 11 year old boy with acromioclavicular dysplasia. Radiographs of the shoulder, anterior-posterior view, right (A) and left (B), axillary view, right (C) and left (D), show bilateral hypoplasia of the clavicle (1) and acromion (2) with normal sized humeral head (3) in contact with the glenoid (4).

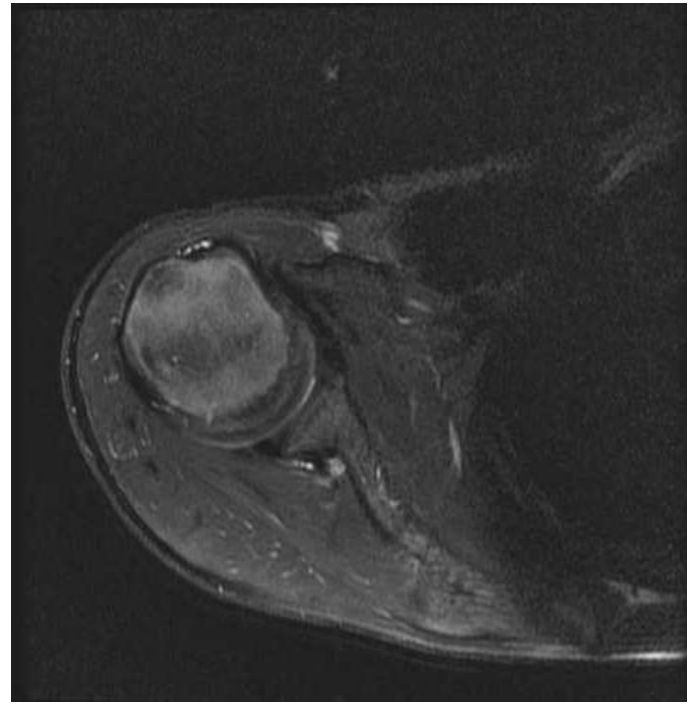


Figure 4: 11 year old boy with acromioclavicular dysplasia. MRI imaging (GE Genesis Signa 1.5T, T1 weighted, TR 500 ms, TE 13 ms, spin-echo, fat suppression, with contrast) of the right shoulder of the patient (frontal view). The white arrows point toward the deltoid muscle completely covering the humeral head and inserting at the hypoplastic acromion (*).

Figure 5: 11 year old boy with acromioclavicular dysplasia. MRI imaging (GE Genesis Signa 1.5T, T2 weighted, TR 4500 ms, TE 52.42 ms, fast spin echo, fat suppression, no contrast) of the right shoulder of the patient (axial view). This shows the glenohumeral joint with an intact glenoid and labrum.

Etiology	Unknown
Incidence	Single case, but possibly underreported
Gender Ratio	Unknown
Age Predilection	Single case of 11 year old boy
Risk Factors	Unknown
Treatment	Conservative treatment
Prognosis	In a single subject, good short-term outcome on conservative treatment
Findings on Imaging	X-ray shows hypoplasia of the clavicle and acromion, MRI shows elongated deltoid head

Table 1: Summary table of acromioclavicular dysplasia

Diagnosis	Imaging Findings X-ray	Imaging Findings MRI
Acromioclavicular Dysplasia	Abnormally shortened clavicle and hypoplastic acromion.	Elongated deltoid muscle originating from the hypoplastic acromion and clavicle and leading over the humeral head.
Agnesis of the Acromion	Absence of the acromion with a hypertrophied distal end of the clavicle and coracoid process [6].	
Glenoid Dysplasia	Decreased ossification of the lower two-thirds of the osseous glenoid and adjacent area of the scapular neck. Associated ill-defined anomalies are possible, including shortening of the limbs, brachymesophalangy and clinodactyly [9].	Posterior sloping of glenoid articular surface and truncation of posteroinferior glenoid rim. Thickening of abnormal low-signal posterior soft tissue. This can be associated with posterior labral tear [5].

Table 2: Differential table of acromioclavicular dysplasia

ABBREVIATIONS

MRI = magnetic resonance imaging
 ROM = range of motion

KEYWORDS

Acromioclavicular dysplasia; shoulder dysplasia; developmental abnormality; shoulder; X-ray; magnetic resonance imaging

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