JSES Reviews, Reports, and Techniques 1 (2021) 127–129

Contents lists available at ScienceDirect

JSES Reviews, Reports, and Techniques

journal homepage: www.jsesreviewsreportstech.org

Unilateral congenital absence of the acromion: a case report and literature review

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ARTICLE INFO

Keywords:
Acromion
Congenital absence
Shoulder instability
Musculoskeletal disorders
Acromioclavicular dysplasia

The acromion is an important anatomic landmark of the shoulder, serving as the muscular attachment site of the deltoid and trapezius, and ligamentous attachment site of the coracoacromial ligament and acromioclavicular joint capsule.[17] The acromioclavicular joint also facilitates both normal shoulder range of motion and stability by allowing for superior and inferior gliding motion of the clavicle, while preventing superior translation of the humeral head.[12] The acromion is formed by three ossification centers, the meta-acromion, meso-acromion, and preacromion, which typically appear on radiographs at 10 to 15 years of age, with full fusion occurring at 19 to 25 years of age.[7] In 1.3-15% of the population, fusion of the acromion fails and children develop os acromiale, more commonly seen in blacks.[2,18] Os acromiale is a well-documented condition that has known associations with subacromial impingement and rotator cuff tears.[2] However, complete congenital absence of the acromion is rare, with only two reported cases of congenital bilateral acromial absence[9,12] and 1 reported case of bilateral hypoplastic acromia.[1] Here, we report the first case of unilateral congenital absence of the acromion, in an adolescent boy with concurrent ipsilateral musculoskeletal anomalies and truncus arteriosus.

Case report

History/presentation

The patient is an 11-year-old boy, born preterm, who grew up in the Democratic Republic of the Congo. As per discussions with him and his father, he had no known cognitive abnormalities, though he had limited medical care during his childhood. When he was initially evaluated by orthopedic surgery, he was nearing the conclusion of a hospitalization for surgical management of truncus arteriosus. His father requested the evaluation owing to concern about many years of atraumatic limp and limited right shoulder range of motion. He had not been evaluated by an orthopedic surgeon before this point.

Physical examination

On examination, he was a happy, engaged child who responded to directions. He did not exhibit dysmorphic facial features. He was slim and tall.

Upper extremities

The right shoulder sat higher than the left, without bony or soft-tissue tenderness to palpation. Right shoulder passive abduction was limited to 100 degrees, active abduction was limited to 90 degrees secondary to pain, and he was able to internally rotate to mid-thoracic region. Compared with the left side, there was increased, 1+ anterior/posterior and inferior/superior translation of the proximal humerus relative to the glenoid, with a firm endpoint. Right deltoid strength was 4/5 compared with 5/5 on the left side. No pain was elicited with rotator cuff testing or the Hawkins impingement test. Neer test could not be performed owing to decreased range of motion. The remainder of the bilateral upper extremity strength, range of motion, and neurovascular examinations were within normal limits.

Lower extremities

No gross abnormalities were present on inspection. No tender areas were identified to palpation. Hip flexion and extension were full bilaterally. Hip abduction was symmetric bilaterally. Hip

Institutional review board approval was not required. Consent was obtained from both patient and father for publication of case report and de-identified images.

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https://doi.org/10.1016/j.xrrt.2020.12.004

internal and external rotation were measured supine and revealed a bias to internal rotation. A right-sided Galeazzi test was positive, with right side shorter than left side by approximately 2 cm. Right hamstrings strength was 4/5, but normal muscle strength was verified in the remaining musculature of the lower limbs. The right knee had a 10-degree flexion contracture and hyperactive reflexes L4 and S1 were present on the right side. Spasticity and ankle clonus were absent, and he had a normal neurovascular examination on the lower limbs. On standing, he exhibited pelvic obliquity with the right iliac crest approximately 2 cm lower than the left iliac crest. Gait assessment by observation revealed an antalgic pattern with a limp on the right side.

Radiographic findings

Radiographs demonstrated right unilateral absence of the acromion, without fracture or dislocation, and maintenance of the glenohumeral joint (Fig. 1). Left shoulder radiographs demonstrated no fractures or abnormalities. An anterior-posterior standing spine series radiograph demonstrated a dysplastic right acetabulum with incomplete coverage of right femoral head consistent with developmental hip dysplasia, gracile bone, and possible dextroscoliosis (Fig. 2).

Literature review

To our knowledge, only two cases of bilateral congenital absence of the acromia and 1 case of bilateral hypoplastic acromia have been described previously.[9,12] Unilateral congenital absence of the acromion has not been previously described in the extant literature (Table I). All described cases have been managed nonoperatively.

Discussion

Herein, we present the case of a 11-year-old boy with unilateral congenital absence of the acromion, with concurrent ipsilateral abnormalities of the lower extremity. This case merits presentation as it is (1) the first documented case of unilateral congenital absence of the acromion, (2) only the third case report of any type of congenital acromial absence or hypoplasia, and (3) associated with concurrent ipsilateral developmental hip dysplasia.

In the patient younger than age 25 years, some discussion may be put toward the differentiation of developmental delay in acromial ossification vs. complete congenital absence or os acromiale. Previously, this distinction is described by Sammarco[18] on the basis of morphologic characteristics, defining os acromiale as a clean, linear joint horizontal to the acromial axis, whereas delayed development demonstrates a rough, crescent-shaped border with osseous spicules interdigitating with the proximal aspect of the acromion. This distinction had previously been described in 1893 and 1993 by Macalister[15] and Edelson et al,[4] respectively, although all in cadaveric specimens. Similar to the findings noted in the 11-year-old case described by Bichler, the young age of the patient makes it possible that this hypoplasia is some type of developmental variant.[1] However, this is unlikely owing to (1) the extent of the hypoplasia, (2) the morphology, and (3) the unilateral nature of the hypoplasia in this patient, presenting a direct comparison to the opposite shoulder which demonstrates age-appropriate acromial development.

While the presentation of the patient in question is not consistent with established presentations of musculoskeletal syndromes, perhaps the most closely related syndrome is the Sprengel deformity, first described in 1891.[8,13] Sprengel deformity is characterized by congenital elevation of the scapula limiting shoulder abduction, typically to less than 90 degrees, with several associated

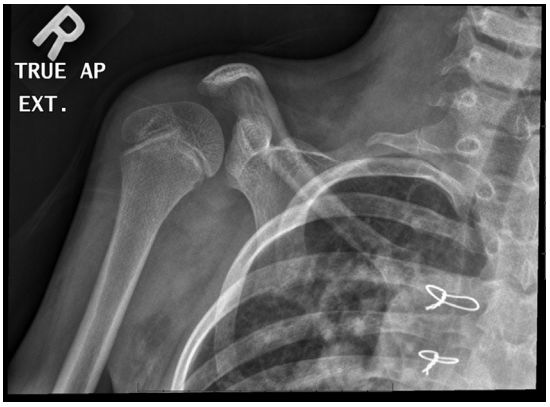

Figure 1 Anterior-posterior radiograph of the right shoulder showing complete absence of the acromion.

anomalies such as scoliosis (35-55%), omovertebral bone (20-50%), and in rare occasions, congenital dislocation of the hip (1-4%) and femoral shortening (1%).[8] At least 1 case report of Sprengel deformity in a 16-year-old boy describes shoulder anomalies similar to this case, including limited abduction; however, the acromion was elongated, rather than absent.[3] Other previously described scapulailiac dysostoses such as Nail-Patella syndrome and Kosenow syndrome were considered.[5,14] However, given that these are associated with characteristics not present in our case, such as absent fingernails, bony dysplasia of elbow and knee, significantly more dysmorphic features, or features such as Turner syndrome, they did not adequately fit with the patient's presentation.[5,14] It is of interest to note that all the patient's abnormalities, acromial absence, hip dysplasia, and knee contracture were ipsilateral.

As so few cases of acromial absence have been described, we can only posit the biomechanical issues that such patients may incur by reviewing clinical anatomy. For example, the patient exhibited weakness in shoulder abduction. As the intermediate fibers of the deltoid typically insert on the superior aspect of acromion and are

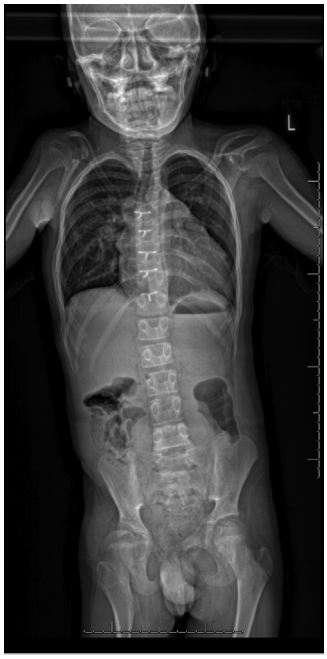

Figure 2 Anterior-posterior spine series radiograph shows right dysplastic acetabulum with incomplete coverage of the femoral head.

Table I
Reports of congenital absence of the acromion or hypoplastic acromion.

Author	Year	Sex	Age	History/ Presentation	Diagnosis	Management
Kim[12]	1994	F	31	Unilateral shoulder pain without inciting trauma	Bilateral congenital absence of the acromia with anterior translation of humeral heads	Conservative
Hermans[9]	1999	M	57	Unilateral shoulder pain without inciting trauma	Bilateral familial congenital absence of acromia, also diagnosed in brother (65) and sister (60)	Conservative
Bichler[1]	2014	M	11	Unilateral shoulder pain following jumping on trampoline	Bilateral hypoplasia of clavicle and acromia	Conservative

responsible for abduction throughout the shoulder range of motion[11], this weakness is likely because of acromial absence. The acromion also acts as a shoulder stabilizer by limiting vertical displacement of the humerus and clavicle.[6] This expected disruption to functional anatomy was observed in the patient's radiographs. Finally, the patient exhibited firm endpoints with anterior/posterior and inferior/superior translation of the glenohumeral joint on examination suggesting no gross ligamentous deficiency. This may be why the patient's shoulder function is largely preserved, and should a magnetic resonance imaging be clinically indicated in the future, it would be of interest to note the modified attachment sites of the coracoacromial ligament and deltoid.[6]

While evident that his absent acromion contributed to his abnormalities during physical examination, he was not significantly distressed by his limited shoulder abduction and denied significant functional limitations of his right shoulder. This is consistent with other reports of congenital acromial absence in which the anomaly was only discovered later in life, in patients aged 31 and 57 years, when they presented with atraumatic pain and no evidence of concurrent osteoarthritis, respectively.[9,12] All described cases were managed nonsurgically.[1,9,12] Relatively minor functional impairment secondary to acromial absence is consistent with acromial resections[10,19] in which the acromion may be surgically excised in cases of subacromial impingement and rotator cuff arthropathy with improvement in pain and only minor functional limitations.[10,16] As he is significantly younger at age 11 years than the other described patients and is already presenting with pain in shoulder abduction, it is likely that his long-term management may differ. Notably, he was more concerned with his limp secondary to his hip dysplasia and ongoing management of his truncus arteriosus. Regarding his right dysplasia, surgical management will likely be indicated.

Conclusion

The patient reported in this article has unilateral congenital absence of the acromion with ipsilateral hip dysplasia. Congenital unilateral absence of the acromion has not been previously documented in the literature. Based on prior case reports of bilateral congenital absence and hypoplasia of the acromia, as well as our experience with our patient thus far, it appears these anomalies are largely asymptomatic and can be managed conservatively.

Funding

No funding was disclosed by the author(s).

Conflicts of interest

The authors, their immediate families, and any research foundations with which they are affiliated have not received any financial payments or other benefits from any commercial entity related to the subject of this article.

Patient consent

Obtained.

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