Congenital, Symptomatic Glenoid Hypoplasia: A Case Report and Review of Literature

Sreekanth Reddy Rajoli¹, Vibhu Krishnan Viswanathan², Umesh Meena³, Sudesh Pebam⁴
¹Clinical Fellow, Spine Surgery, Ganga Hospital, Coimbatore, Tamilnadu, India
²Registrar, PGIMER, Chandigarh, India
³Consultant, PGIMER, Chandigarh, India

*Corresponding Author:
Name: Dr. Vibhu Krishnan Viswanathan
Email: drvibhu007@gmail.com

Abstract: Glenoid hypoplasia is a rare congenital anomaly characterised by abnormal development of the inferior portion of the glenoid fossa. It may be seen as a primary isolated lesion, or in association with other anomalies or as a part of syndromes. It is predominantly bilateral, but few unilateral cases are reported in the literature. We present a case of unilateral, symptomatic glenoid hypoplasia in a 6-year-old female child, who had presented to us, due to the restricted shoulder abduction, as well as unsightly appearance of her right shoulder. The uniqueness of the case and the relevant literature has been appropriately highlighted.

Keywords: Glenoid hypoplasia, Restricted shoulder abduction

INTRODUCTION
Glenoid hypoplasia is a relatively rare congenital anomaly characterised by abnormal development of the inferior portion of the glenoid fossa [1]. It may be seen as a primary isolated lesion, in association with other anomalies or as a part of syndromes [1, 2]. Exact prevalence is not known, as the cases may remain asymptomatic in a majority of patients and never manifests clinically in their entire life [3, 4]. It is predominantly bilateral, but a few unilateral cases are reported in the literature [2, 5]. Radiologically, glenoid hypoplasia may show widening of the glenohumeral joint space, labral hypertrophy, hyperplasia of the coracoid process and acromion, hooking of the distal clavicle, and hypoplasia of the humeral head [6]. Early development of osteoarthritis has been described, which may require arthroplasty later in these patients [7, 8]. In this report, we describe a rare, interesting case of unilateral, symptomatic glenoid hypoplasia in a 6-year-old child, with associated humeral head hypoplasia. The uniqueness in the presentation of the condition has been well foregrounded; and the relevant literature, appropriately discussed below.

CASE REPORT
A six-year-old female child, right hand dominant, was seen at our hospital due to the restricted shoulder abduction as well as unsightly appearance of her right shoulder. Patient had no pain. Though the parents of the child were concerned about the appearance of the shoulder and the limitation of the overhead abduction, the child did not have any difficulty performing her daily activities. Past history was unremarkable and her birth was by normal, uncomplicated, vaginal delivery. Developmental milestones were within normal limits. Her parents denied the presence of any similar pathology in any family member.

Physical examination revealed limitation of overhead abduction without any instability of the shoulder. All the other movements at the affected right shoulder joint were the same as that of the left shoulder. Motor power in all the muscle groups seemed to be equal to the opposite arm, but there was deltoid atrophy noticed. There was a loss of normal contour of the involved shoulder with abnormal prominence of the acromion process and the head of the humerus in comparison to the contralateral shoulder. Other examination findings included negative anterior apprehension test, normal load and shift test, absent inferior sulcus sign, and normal posterior load shift. No generalized ligament laxity could be found according to the Brighton’s index. Radiological examination revealed flat, shallow, narrow glenoid fossa, and a small hypoplastic humeral head. Blood and urine investigations were within normal limits.

DISCUSSION AND REVIEW
Giongo [9] and Heupke [10] were the first to describe this entity; and till date as many as 100 patients with bilateral hypoplasia have been described in the literature. Although, multifarious factors including birth...
fractures, growth plate injuries, obstetrical palsy, multiple epiphyseal dysplasia, and neuro motor disorders have been purported to be associated with this condition, the precise etiology and pathogenesis still remains obscure hitherto.

During the normal shoulder development, the scapula develops by intramembranous ossification from about eight centres. At birth, although most of the scapula is ossified, the glenoid along with acromion, corocoid process, vertebral border and inferior angle of scapula remains cartilaginous. Two secondary centres, a superior sub-coracoid centre which develops at age of 10 and fuses by about age 15yrs; and an inferior horse-shoe shaped centre which appears near puberty, ossify to form the bony glenoid. Hypoplasia can result from the aberration in the normal development of any of these processes [1]. Fibrocartilaginous tissue or fat tissue replacing the hypoplastic glenoid and scapular neck have been described in a few reports, signifying that factors other than aberrant ossification might be involved [11]. Failure of the inferior center seems to be one of the most important causes. Although a hereditary background has been described, the exact genetics are not lucid, heretofore [4]. Other clinical differential diagnoses, including avitaminosis C and D, Erb’s palsy, muscular dystrophy, neonatal septic arthritis, however, need to be excluded especially in unilateral cases [4].

Patients with glenoid hypoplasia may be asymptomatic or may develop a range of symptoms like pain (severity of which may vary), stiffness of the shoulder causing restriction in the range of motion or instability [2, 4]. In their largest series of glenoid hypoplasia, Smith et al described four children were asymptomatic throughout, while the rest of the eight developed symptoms later on, in adulthood [12]. While, in childhood, the condition is mostly incidental discovered, adults present with the afore-mentioned symptoms [4, 12]. They may develop early osteoarthritis too. Our patient, a child of 6yrs of age, presented with the symptom of restricted shoulder abduction. This condition is usually bilateral [2, 5]. Less than 10 unilateral cases have been reported, as yet [13].

Humeral head hypoplasia has also been associated with glenoid hypoplasia [12]. Resnick et al. [9], Munshi et al. [11] had documented 2 cases each with humeral head dysplasia, while Smith et al. [12] did not discover any humeral abnormality in their series. They had mentioned variety of associated findings such as hooking or bossing of the distal clavicle, prominent coracoids etc. In conclusion, to our knowledge only few cases of unilateral glenoid hypoplasia, especially with associated humeral head hypoplasia has been reported.

This report details the findings of a symptomatic child with unilateral glenoid hypoplasia with humeral head hypoplasia. In younger patients, physiotherapy only is sufficient where as elderly patients with symptoms may require arthroplasty [2]. Our patient is currently under a supervised neglect regime, on regular physiotherapy protocols.

Fig. 1: Clinical photographs of the patient showing gross wasting of the right shoulder as well as decreased range of motion of the shoulder

Fig. 2: X-ray showing glenoid and humeral head hypoplasia
Fig. 3: Cross sectional T1 and T2 MRI images showing both glenoid and humeral head hypoplasia

Fig. 4: Coronal T1 and T2 MRI images showing both glenoid and humeral head hypoplasia

Fig. 5: Coronal T1 images of the right shoulder
REFERENCES