A vestige of an extra upper-limb skeleton in the left shoulder

Dear Sir,

A 31-year-old woman was unable to flex or abduct the left shoulder beyond 90°. She sometimes experienced a dull ache around the shoulder. Internal and external rotation of the shoulder was normal. There was no cosmetic defect and the shoulder region had a normal contour. The body of the left scapula had normal borders and a normal position on the chest wall. It was level and symmetrical with the right scapula. The left trapezius muscle was mildly atrophic. The left elbow and hand and the right shoulder were normal. On palpation there was a hard structure in the anterolateral section of the deltoid muscle. It was attached to the acromion and did not have independent movement. There were no other musculoskeletal or visceral abnormalities. She remembered that during previous medical examinations in the childhood and adolescent periods she had been diagnosed as having birth palsy with involvement of the upper roots. Earlier radiographs were not available but she was not informed of any abnormal osseous structure around her left shoulder. Electromyography and nerve conduction showed no neurological deficit.

Plain radiography revealed a finger-like osseous structure projecting laterally from the acromion process (Figure 1). The precise location of the abnormal structure was demonstrated by three-dimensional computerized tomography (3D CT) imaging. A superior view of the shoulder showed that the scapular spinous process terminated in a bifid acromion process. The medial acromion process articulated with the left clavicle (Figure 2(a)). The posterior view revealed that the abnormal structure extended laterally from the lateral acromion process. The abnormal structure consisted of distal and intercalated osseous fragments with two intervals (Figure 2(b)). An anterolateral view of the shoulder demonstrated that the glenoid fossa was underdeveloped and the humeral head did not articulate with it. The coracoid process also had an abnormal shape (Figure 2(c)). The patient declined surgery to remove the structure. Therefore, we were not able to provide further information about the histopathology and a probable soft tissue abnormality.

Since the limb buds develop in a proximal-to-distal direction it might be interpreted that the intermediate osseous structure was a vestigial predecessor of an extra humerus, even though it did not articulate with the glenoid fossa, and the distal structure was a vestigial predecessor of an extra below-elbow structure. It could be that the abnormal osseous structure was a vestige of an extra upper-limb skeleton.

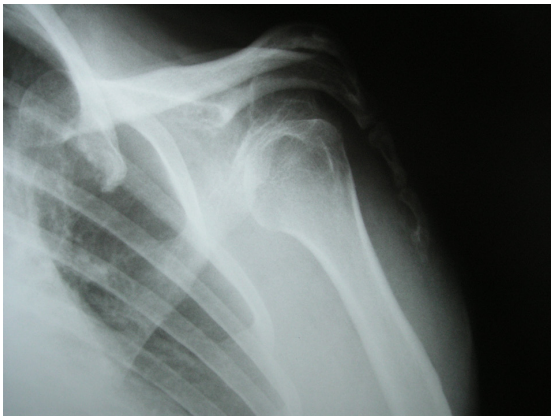

Figure 1. Plain radiography reveals a finger- like structure projecting laterally from the acromion process.

The limbs buds appear 26 days after fertilization. They are formed by an outgrowth of somatic and lateral plate mesoderm into the ectoderm. The lateral plate mesoderm forms bone and cartilage. The junction between the ventral and dorsal ectoderm is called the apical ectodermal ridge (AER). The first signal interactions in the limb bud, which stimulate the development of the limb bud from proximal to distal, occur between the AER and the underlying mesoderm. In early embryonic life when the root of a limb begins to form, the signals expressed from the AER to the underlying mesoderm stimulate formation of the underlying structures (Al-Qattan et al., 2009; Daluiski et al., 2001; Oberg et al., 2006). An embryological explanation of the current case might be that erroneous signals from the AER stimulated the lateral plate mesoderm to produce an extra upper-limb skeleton; however, the signals were limited and produced a vestigial predecessor of the upper-limb skeleton.

During the fifth week of the embryonic period the central core of the humerus begins to chondrify, but the precursor of the shoulder joint does not chondrify. During the sixth week the glenoid lip is discernable but cavitation of the joint has not yet occurred. During the seventh week the shoulder joint develops (O'Brien et al., 2004). It might be assumed that the embryological error in the current case coincided with the time that the shoulder joint began to form. The structures around the shoulder are not completely ossified and are not recognizable by radiography before puberty. The ossification centres of the acromion appear during puberty and fuse at around 22 years (O'Brien et al., 2004). It is likely that, in the current case, the abnormal structure ossified and matured in adulthood, although it was cartilaginous at the time of earlier medical examinations in

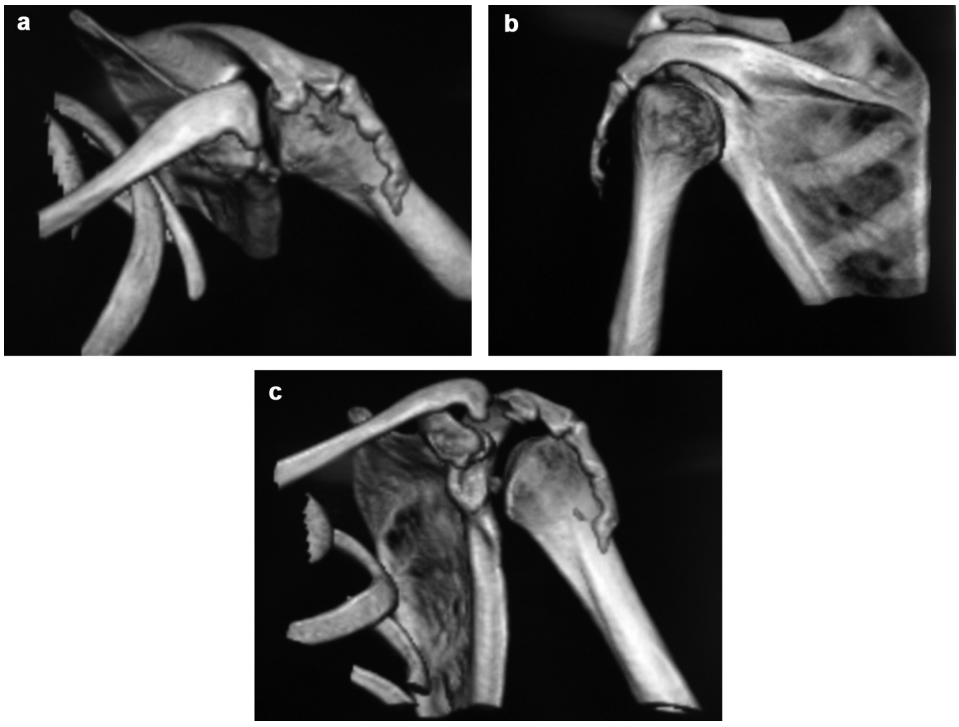

Figure 2. (a) Superior 3D CT view of the shoulder shows a bifid acromion process. (b) Posterior view demonstrates that the abnormal osseous structure extends from the spinous process of the scapula and has distal and intercalated osseous structures. (c) Lateral view shows that the glenoid fossa is underdeveloped and the humeral head does not articulate normally with it. The coracoid process has an abnormal shape.

childhood. Therefore, the nature and size of the abnormality was not discernable earlier and the condition was misdiagnosed as a case of birth palsy with the involvement of the upper roots.

Conflict of interests

None declared.

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Traction and fixation of four neglected fracture-subluxations of the distal interphalangeal joint

Dear Sir,

The treatment of a chronic displaced distal interphalangeal (DIP) intra-articular fracture can be extremely difficult. We report four DIP intra-articular