Impact Case Reports

Case Report

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Congenital unilateral absence of the clavicle and scapula with preserved humerus: incidental finding in empyema thoracis

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Abstract

Introduction: Congenital musculoskeletal anomalies are among the commonest birth defects described in the literature, with many patterns recognised and others probably unrecorded. Partial absence of the shoulder girdle or isolated clavicular aplasia are documented; complete absence of the scapula is rare and usually described with absence of the limb. We report an adult with empyema thoracis in whom complete absence of the left pectoral girdle was found incidentally.

Case report: An adult man presented with eight months of recurrent discharge from the left chest. The illness began after a closed chest injury from a fall, managed initially with an intercostal drain for eight days at a peripheral hospital. Four weeks later he developed recurrent drainage and sought further care. He had no known comorbidities. On examination he was haemodynamically stable, respiratory rate 22 breaths per minute, with an actively discharging sinus at the left fifth intercostal space (mid-axillary line) and two healed drain sites. Chest expansion and air entry were reduced over the left mid and lower zones, with dullness to percussion. The right upper limb was normal. On the left, the shoulder outline was normal; the arm rested at about 30° abduction and active abduction reached about 70°. Elbow, forearm and hand were normal. Chest radiographs showed empyema thoracis and complete absence of the left clavicle and scapula with a normally positioned humerus. MRI was requested but the patient underwent decortication elsewhere and died a few days post-operatively. Consent for publication was obtained from the next of kin.

Discussion: Complete absence of the left pectoral girdle was found incidentally during assessment for empyema thoracis. With an intact humerus and no history of resection or major trauma, the features support a congenital origin and remind clinicians to check for musculoskeletal anomalies when chest imaging is atypical.

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Introduction

Congenital musculoskeletal abnormalities are among the commonest birth anomalies reported in the literature,[1,2,3] with many forms recognised and others likely unrecognised. Partial deficiency of the shoulder girdle or complete absence of the clavicle have been described; however, complete absence of the scapula is rare and is usually associated with absence of the arm[4]. We report a patient with empyema thoracis and an incidental finding of complete absence of the left pectoral girdle.

Case report

An adult man was reviewed after eight months of recurrent discharge from the left chest. The illness began after a closed chest injury from a fall from a tree, managed initially at a peripheral hospital with an intercostal chest

drain for eight days. About four weeks later he developed recurrent drainage and presented to a tertiary centre for further care. He had no known comorbidities.

On examination he was well preserved and haemodynamically stable. Respiratory rate was 22 breaths per minute. There was an actively discharging sinus on the left hemithorax at the fifth intercostal space, mid-axillary line, and two healed chest-drain sites. Chest expansion and air entry were reduced over the left mid and lower zones, with a dull percussion note.

Musculoskeletal examination showed a normal right upper limb. On the left, the shoulder contour was normal but the arm was held at about 30° of abduction (attributed to prolonged chest discharge) and active abduction was reduced to about 70°. The elbow, forearm and hand were normal in appearance and function.

Chest radiographs provided by the patient (figures 1 and 2) showed features of empyema thoracis and complete absence of the left pectoral girdle (clavicle and scapula) with a normally positioned humerus. MRI was requested to define the anomaly further, but the patient subsequently underwent decortication surgery elsewhere and died a few days post-operatively. No additional requested investigations were completed. Written consent for publication was obtained from the next of kin.

Discussion

Although the evaluation was incomplete because of the patient's death, this is a rare clinical finding. The absent left pectoral girdle is unlikely to be secondary to infection: adjacent ribs were intact and there was no history suggestive of surgical resection or traumatic loss. A congenital origin is therefore most plausible and has been proposed previously by Samilson[4]. Hawkes and Wells described absence of the left upper limb and pectoral girdle in an Anglo-Saxon skeleton, supporting the possibility of congenital agenesis[5].

Differential considerations include Poland syndrome and cleidocranial dysplasia. Poland syndrome typically involves absence or hypoplasia of the pectoralis muscles with associated chest-wall defects (for example, absent segments of sternum, rib or costal-cartilage anomalies) and characteristic hand malformations, none of which was present here[6]. Cleidocranial dysplasia features clavicular aplasia or hypoplasia with craniofacial and dental anomalies; these were not identified in this patient[7].

In summary, the features are most consistent with congenital absence of the left pectoral girdle with a normally formed humerus, discovered incidentally during assessment of empyema thoracis.

Consent: The patient's next of kin granted written informed consent for the publication of this case report which is held by the authors.

Competing interests: None declared.

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