Longitudinal Dysplasia with humerus buried within soft tissue of the chest wall

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Abstract We describe a patient presenting with upper-extremity longitudinal dysplasia in whom the humerus was buried within the tissues of the chest wall. Surgery was performed to elevate the left humerus from the upper chest and place it within the empty soft tissue envelope, which extended distally from the shoulder. To our knowledge, this particular anomaly has not been previously described in the literature.

Keywords Buried Humerus · Longitudinal Dysplasia · Reconstruction

Introduction

Upper-extremity longitudinal dysplasia is a severe congenital malformation defined as a failure of formation of bone along its longitudinal axis [1]. According to the morphological classification of congenital anomalies of the upper extremity adopted by the International Federation of Societies for Surgery of the Hand (IFSSH), longitudinal arrest is a division under ‘Failure of Formation of Parts’, the first of IFSSH’s seven categories [1]. Longitudinal dysplasia and similar upper limb hypoplasias represent a challenge for both the patient as well as for the treating surgeon. The functional deficit is significant, and any gain in function through surgery, a prosthesis, or both can significantly improve the quality of life for an affected patient. This report describes an extremely unusual case of a patient with longitudinal deficiency in whom the humerus was buried within the tissues of the chest wall and the surgical treatment that was performed in infancy to provide better function and to allow for subsequent fitting and use of a prosthesis. This case is unique in that our patient’s humerus was located within the subcutaneous tissue of the upper chest wall and a soft-tissue envelope radiated out from the shoulder with no bone present.

Case description

An 8-month-old boy presented with hypoplasia of bilateral upper extremities, along with multiple other birth defects. He was born to a 19-year-old G₁P₁ Caucasian female whose pregnancy had been complicated by gestational diabetes, herpes infection, toxemia of pregnancy, and placenta previa with multiple episodes of bleeding. The parents denied any family history of limb abnormalities, mental retardation or any other genetic defects.

The left upper extremity exhibited hypoplasia of the entire arm with only a single distal nubbin (Figs 1A–C)

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This single nubbin was attached to an empty soft tissue envelope extending distally from the shoulder, creating a very shallow pseudo-axilla. A bony structure, presumed to be the humerus, could be palpated within the soft tissue overlying the left chest wall in the mid axillary line, and there was palpable movement of the buried humerus preoperatively indicating the presence of functional muscle attachments. In particular, the infant appeared to move the buried humerus in efforts to maintain his balance while sitting or standing. Radiography confirmed the presence of a humerus within the soft tissues of the chest wall; there was no radiographic evidence of osseous structures distal to the left humerus (Fig. 2). Clinically and radiographically it appeared that there was a functional glenohumeral articulation. The right upper extremity was significant for distal humerus hypoplasia, as well as a hypoplastic radius and presence of a single carpal and phalangeal structure (Fig. 1A). The patient underwent an echocardiogram, which was normal with the exception of a mild patent ductus arteriosus.

Surgical treatment was planned to liberate the buried humerus from the chest wall and to place it into the empty soft tissue envelope extending from the shoulder. The purpose of this procedure was to allow the infant to use the humerus to aid in balance and to provide a stump for later prosthesis fitting. A longitudinal incision was

marked along the medial aspect of the empty soft tissue envelope and onto the chest wall (Fig. 3). Dissection was carried out through the subcutaneous tissue and fascial layers down to the level of the humerus. Upon inspection, the humerus was approximately 7 cm long and had a diameter of 1.5–2 cm. The humerus was then elevated from the chest wall, and a muscle slide was performed by dividing distal attachments of musculature along the length of the humerus and then elevating these attachments off with a Freer elevator (Fig. 4). Care was taken not to disrupt the periosteum during this process and to avoid completely detaching the muscles of the humerus. The humerus was then placed into the soft tissue envelope and held distally with a suture tied over a button on the distal aspect of the soft tissue envelope. The muscles that had undergone the muscle slide were then reapproximated. The intraoperative degree of movement was not tested in order to prevent disruption of the newly

Fig. 1 (A) Bilateral upper extremity initial presentation. (B) Close up view of left chest wall and empty redundant soft tissue at presentation. (C) Intraoperative view of traction on distal nubbin demonstrating extent of empty soft tissue envelope

Fig. 2 Radiograph demonstrating left humerus within soft tissues of chest wall

Fig. 3 Preoperative marking of incision and four flap z-plasty

placed sutures. A four-flap z-plasty was then planned in order to create an axilla of appropriate contour and depth. Each of these flaps was raised and inset in the usual fashion (Fig. 5). A compressive bolster-type dressing was applied. Recovery was uneventful.

At one-year status-post the left humerus elevation procedure, a slight bony prominence was present at the end of the left humerus stump; however, there was no skin breakdown (Figs 6A–B). The patient was using the liberated humerus quite well without complaint of pain. The child was now able to obtain 60° abduction of the humerus to help with balance during walking and demonstrated strong adduction of the limb. He was able to use this adduction strength to carry objects between the humerus and the chest wall (Fig. 6C). In the future, this stump will allow fitting of an upper extremity prosthesis.

Discussion

This case report describes an unusual congenital anomaly of the upper extremity involving a humerus buried within

the subcutaneous tissue of the chest wall. The surgical treatment of this rare problem is described to illustrate how function can be improved in such a case. To the best of our knowledge, this particular anomaly has not previously been described in the literature.

Only two articles have reported upper limb anomalies similar to the one presented in this case report. The first was published in the *Journal of Pediatric Surgery* by Banerjee et al. [2]. That report describes a patient with pulmonary agenesis and an intrathoracic upper limb. This article is similar to our case report in two ways. First, both patients presented with a mass where the upper limb should have developed. Secondly, both children did develop a humerus. Although there are similarities between the two cases, there are some notable differences. To begin with, in our patient, the humerus was located within the subcutaneous tissue of the upper chest wall, while in the other case, the upper limb penetrated into the thorax resulting in respiratory distress. Furthermore, the mass at the shoulder of the infant in Banerjee's report contained cystic and skeletal elements and was adherent to the chest, whereas in our patient an empty soft-tissue envelope intended for the humerus was present extending from the shoulder and separated from the chest wall. Also, our patient had only a single, rather than a duplicated, humerus. Finally, there were no structures present distal to the humerus in our patient, as opposed to

Fig. 4 Intraoperative view of humerus being elevated with a hook. Arrow demonstrates soft tissue attachments tethering humerus to chest wall

Fig. 5 (A) Immediate postoperative view demonstrating four-flap z-plasty inset and button holding humerus distally in soft tissue envelope. (B) Immediate postoperative anterior view of axilla with four flap z-plasty inset

Fig. 6 (A) One-year postoperative view demonstrating contour of axilla and active abduction of humerus in its soft tissue envelope. (B) One-year postoperative close up view demonstrating contour of axilla and healed four-flap z-plasty. (C) Demonstrating adduction and ability to hold objects

the deformed hand attached to the forearm within the thorax in the other case.

The second article was written by Gupta et al. [3] who described a case of congenital amputation of the right forearm. One notable similarity between the two cases was the ‘excess’ of soft tissue that was present in both patients’ upper limb anomalies. Such a soft-tissue envelope can be useful in surgical reconstruction, as demonstrated by our case. The major difference between the two cases lies in the fact that although our patient had a soft-tissue envelope that radiated out from the shoulder, no bone was present within it.

Longitudinal dysplasia results in a functional deficit for the patient. The goal of surgery is to improve the quality of life for an affected patient, and this is accomplished either through manipulation of tissue and/or a prosthesis. By performing the surgical treat-

ment in infancy, a better functional outlook may be possible.

The benefits of surgical treatment in our patient were seen at the one-year postoperative visit: (1) the presence of a supple, anatomic-appearing axilla; (2) the patient’s ability to actively flex, adduct and abduct the arm without pain; (3) the presence of stable soft-tissue coverage over the end of the humerus.

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