CASE REPOSITORY

Upper Limb Dimelia

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A patient with upper limb dimelia including a double scapula, humerus, radius, and ulna, 11 metacarpals and digits (5 on the superior side, 6 on the inferior side) was treated with a simple amputation of the inferior limb resulting in cosmetic improvement and maintenance of range of motion in the preserved limb. During the amputation, the 2 limbs were found to be anatomically separate except for the ulnar nerve, which, in the superior limb, bifurcated into the sensory branch of radial nerve in the inferior limb, and the brachial artery, which bifurcated into the radial artery. Each case of this rare anomaly requires its own individually carefully planned surgical procedure. *(J Hand Surg Am. 2017;■(■):1.e1-e5. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Upper limb dimelia, duplication, amniotic constriction, epidermoid cyst, congenital anomaly.

Online Exclusive

UPPER LIMB DIMELIA IS A VERY RARE congenital anomaly that involves the duplication of the entire upper limb. There are 3 case reports in the medical literature of children with extra upper limbs, all originating near the glenohumeral joint.[1–3]

There are no reports of 2 upper limbs on the same side united by skin alone. In this report, we present the clinical details of such a case and describe our surgical decision making. The surgery involved excision of the inferior limb and transfer of the inferior limb biceps to the superior limb.

CASE REPORT

A 3-month-old boy presented to our clinic with upper limb dimelia. The prenatal history was notable only for maternal preeclampsia. Prenatal ultrasound was reported as normal. The boy was born via normal spontaneous vaginal delivery at full term. At birth, he was noted to have a diaphragmatic hernia and an accessory ear on the left side. He had undergone surgery for the diaphragmatic hernia on the first day after birth. He required home oxygen therapy following discharge from intensive respiratory care in the neonatal intensive care unit. There was no family history of medical abnormalities.

On physical examination, he had 2 arms on the right side united only with skin (Fig. 1). The superior extremity had 5 digits with some active movement, and the inferior one had 6 digits with flexion contractures (Video A; available on the *Journal*'s Web site at www.jhandsurg.org). The elbow could be flexed to 90° and extended to −10° (Video B; available on the *Journal*'s Web site at www.jhandsurg.org). The proximal humerus of each limb articulated with a glenoid, with a 20° to 90° range of shoulder abduction. There was no independent movement between the superior extremity and the inferior one.

Plain radiographs showed a double scapula and humerus. The superior limb arose from the superior

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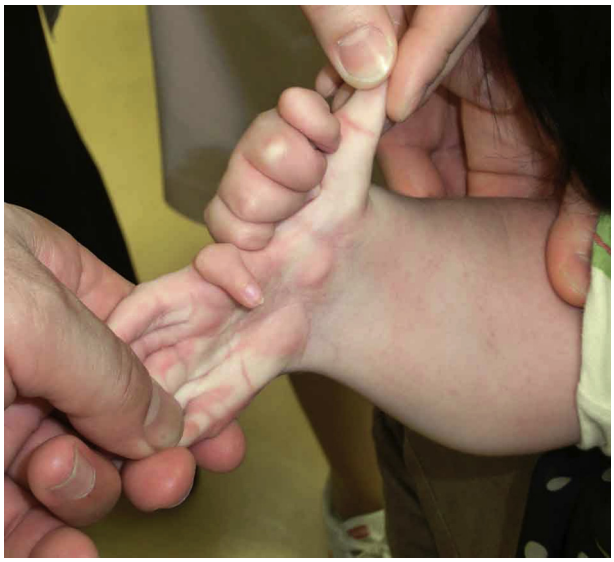

FIGURE 1: Two upper arms on the right side that were united with skin.

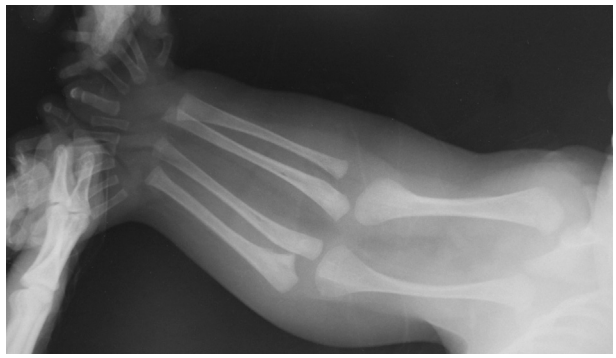

FIGURE 2: Standard radiographs showed a double scapula, humerus, radius, and ulna, with 11 metacarpals and phalanges (5 on the superior side, 6 on the inferior side) on the right side.

scapula, and the inferior limb originated from the smaller inferior scapula (Fig. 2).The superior limb had a radius and ulna. The forearm of the inferior side had ulnar dimelia, 2 identical ulnae without a radius that joined the lower end of the humerus and the wrist.[4,5] There were 11 digits (5 on the superior side, 6 on the inferior side).

Computed tomography angiography of the 2 limbs and the thorax demonstrated that a single large artery ran to each of the 2 limbs (Fig. 3). The superior artery ran in the typical position of the subclavian, axillary, and brachial arteries and bifurcated into the superior and inferior extremities. A single large vessel originating from the subclavian artery traveled posteroinferiorly along the left lateral chest wall to supply the inferior extremity. This pattern was similar to a previously reported case of upper limb duplication.[3]

The child's family strongly desired surgical reconstruction to create 1 extremity. A surgical plan was designed based on the clinical examination, radiographs, and computed tomography angiography. By 4 years of age, the child's respiratory condition had stabilized sufficiently for elective surgery, and he was beginning to become aware of his upper limb difference. The surgical team consisted of surgeons (A.N., A.S., and S.T.) and ancillary personnel well trained in congenital upper limb differences and microsurgery.

Surgical plan

Without a tourniquet, a longitudinal incision was made starting at the midline of the superior limb (Fig. 4). The 2 upper arms were easily separated because there was little soft tissue between them with the exception of a cartilage anlage connecting the medial humeral epicondyle of the superior extremity with the lateral humeral epicondyle of the inferior extremity. The ulnar nerve in the superior limb bifurcated into the sensory branch of radial nerve in the inferior limb; we divided the ulnar nerve at this bifurcation (Fig. 5). The brachial artery in the superior limb bifurcated into the radial artery in the inferior limb at the level of the elbow, as shown on computed tomography angiography (Fig. 3). We divided the artery at this bifurcation. After cutting the cartilage binding the epicondyles, the 2 limbs could then be separated. The inferior arm was divided at the midpoint of the humeral diaphysis and excised. The biceps muscle in the superior limb was hypoplastic; therefore, the biceps muscle from the inferior limb was transferred to the superior limb (Fig. 6). The proximal humerus from the inferior limb was then removed from the surrounding tissue above the periosteum. The inferior scapula was preserved owing to difficulty in excising it. The fingers on the superior side had severe flexion contractures and required volar soft tissue release over the proximal interphalangeal joints, which were temporarily fixed in each finger with a K-wire in maximum extension (Fig. 7). Full-thickness skin graft from the palm on the inferior side was used to close the skin defect left after the release of the proximal interphalangeal joints. The extensor carpi ulnaris was separated from the anconeus muscle, and the distal fibers of the anconeus were divided. Care was taken to protect the posterior interosseous nerve on the inferior side.

The surgical wounds healed uneventfully, and the patient began moving the limb soon after surgery. However, 2 weeks after surgery, we excised an epidermoid cyst on the proximal humerus. We removed the K-wires from the digits 6 weeks after