Accessory arm — dysraphism or disparity?

Case report

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The case of a 3-month-old infant with an accessory third arm is reported. The extra appendage was attached at the midcervical region and was associated with posterior cervical dysraphism and a cervical cord lipoma. Possible theories of origin are examined.

KEY WORDS • accessory limb • arm • spina bifida • neurulation

In tabulating the malformations which are representative of the “anomalous and curious, of exceptional and persistent fascination” to man, Gould and Pyle referred to earlier reports of human subjects with supernumerary limbs. They quoted Pare, who claimed that in January, 1529, there was living in Germany a male infant with four legs and four arms. Bardsley described a male child with one head, four arms, four legs, and double generative organs. Many of these earlier reports as well as more recent publications document cases of accessory lower extremities with or without spinal column and pelvic abnormalities.

There have been few instances of supernumerary arm formation and hardly any mention of associated spinal dysraphism. Tucker described a baby born in New York in October, 1894, who had a third leg hanging from a bony and fleshy union attached to the dorsal spine. Jones and Larkin mentioned the removal of a meningocele and supernumerary limb from a 4-month-old infant. The present report examines the case of an infant with a third arm arising from the posterior midline cervical region and associated with spinal dysraphism and a penetrating cervical cord lipoma.

Case Report

This baby girl was the first child delivered to a 26-year-old mother and was the product of a normal pregnancy and labor. The mother had been taking clomiphene citrate (Clomid) and medroxyprogesterone acetate (Provera) initially, and the father was receiving cortisone. There was no unusual family history. The infant weighed 6 lbs 13 oz. at birth and was sufficiently well to be released home at 5 days of age. She fed from the breast and was gaining weight when first reviewed in neurosurgical consultation at 4 weeks of age.

Examination. The presence of an accessory limb with characteristic features of an arm was obvious at birth (Fig. 1). The arm had a broad attachment to the midline spinal structures at the C3–6 level and extended horizontally across the posterior aspect of the left shoul-

Fig. 1. The accessory arm is located in the midcervical line, approximately covering the C3–6 vertebral segments.
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der. The cervical spine appeared shortened. There was a small central pit of skin at the level of the rudimentary elbow; the elbow was fixed and the hand had four digits. On palpation the limb was soft and fatty, but contained bony structures and a small amount of muscle tissue. The limb demonstrated transmitted movements from shoulder and chest musculature, but from time to time spontaneous flexion of the digits could be observed. The patient did not withdraw any muscle groups of the accessory limb from pinprick stimulation, but did so in response to deep pain. There was also the suggestion of coincident facial grimace to the painful stimuli.

The neurological examination was otherwise normal, with the patient showing a good suck response, palmar and plantar grasps, preserved Moro reflex, and stepping and placing responses. Neurological performance in the two regularly positioned arms and both legs was normal.

Plain spine radiographs and thorough computerized tomography (CT)-metrizamide myelography demonstrated spinal dysraphism at the C4-6 level. A rudimentary scapula, and normal glenohumeral articulation, humerus, ulna, and four digits were associated with the accessory arm which also contained recognizable bone components of ulna and digits. Fatty tissue which was present in the subcutaneous region of the accessory limb traversed the spina bifida, partially circumscribing the extradural sac in the cervical region on one side; the fatty tissue also penetrated the dura as a tongue of tissue extending directly to the intramedullary cord substance (Fig. 2). Electromyography performed in the region of the suspected triceps muscle of the accessory limb showed fibrillation compatible with denervation. The biceps region had fibrillation and good action potentials indicating innervation.

Obviously the limb had to be removed for nursing and cosmetic reasons, and the parents were counseled accordingly. The patient was operated on at 3 months of age; in the interval from birth the patients noted that the limb was continuing to grow relative to the infant's overall growth.

Operation. The surgery consisted first of disarticulation and excision of the accessory limb, followed by cervical laminectomy with release of the tethered spinal cord and debulking of the attached intramedullary lipoma. After reflection of appropriate skin flaps about the accessory limb, the musculature equivalent to the deltoid and biceps muscles was transected and the neurovascular bundles identified. The equivalents of a median nerve and a brachial artery and vein were identified and ligated. Subperiosteal dissection of the humerus down to the level of the shoulder joint was performed and the shoulder was disarticulated (Fig. 3). A rudimentary scapula, which had a well-formed glenoid and a fairly normal-appearing body, remained. This was lo-

![Fig. 2. Photograph of the excised accessory appendage revealing the features of a radial club hand, four digits, and the disarticulated humerus (arrow).](image)

![Fig. 3. Axial computerized tomography-metrizamide myelogram sections (dorsal structures at bottom). The accessory appendage contains the humerus which attempts formation of a glenohumeral joint. The posterior bone arch is incomplete and is invaded by fatty tissue entering from the left side and penetrating cord substance (arrow).](image)
Accessory arm in an infant

cated on the right side of the spine facing outward, and was associated with a horseshoe-shaped bone which may have represented a rudimentary clavicle. This extended across the top of the cervical spine to its left side. The neurovascular bundle was traced down to the level of the spinal dysraphic defect, and all accessory bones, including the scapula and clavicle, and the cartilage remnants were excised. The normal posterior vertebral elements rostral and caudal to the lesion at the levels of C-1 and C-6 were identified. The dorsal arches of C2-5 were dysraphic.

Where required, a posterior laminectomy was completed over the C1-4 levels. With that done, soft tissue and fatty and neurovascular structures were seen entering the dorsal dural sac just to the left of the midline. When the dura was opened and the cord examined with the operating microscope, the appearance was that of a classic dorsal type lipomyelomeningocele. The intradural extension of the nerve roots ran into and blended with fat. A major feeding artery, running along the left lateral gutter intradurally, penetrated the dura to run to the accessory limb. This vessel was coagulated and divided at its dural exit site. Therefore, the tethering was all to the left of the midline and was the extension of the extrudal neurovascular and fatty structures to the dorsal fatty lipoma. This tethering was released, the intramedullary fat substance was partially removed with a Cavitron surgical aspirator, and the dura was closed. The excess skin was trimmed and closed by plastic surgical techniques.

Pathological Examination. Detailed pathological examination of the submitted tissue showed that it had the form of a congenital radial club hand with absence of the radius and thumb, a median nerve with mild axonal degeneration, intact muscle, and neurovascular bundle. A histogram performed on the median nerve showed 66% type H fibers and 33% type I fibers. The histogram was calculated as within the normal range. No karyotyping was performed on the excised tissues. The cervical cord lipoma had no other unusual pathological features.

Postoperative Course. By the age of 4 years, the patient had been treated for persisting torticollis with a night collar. She showed normal spinal development otherwise, and neurological testing was intact except for minor impairment of pencil grasp and slight incoordination of the left arm for complex fine motor tasks.

Discussion

The presence of an accessory limb, especially an arm, is an extraordinary and rare occurrence. The curiosities reported by Gould and Pyle are startling and in most instances seem to represent a forme fruste of twinning. Pittman and Manwaring treated a 6-month-old baby girl born with a mass on the posterior thorax which was a "twin" with incomplete cleavage (HW Pittman and KH Manwaring, personal communication, 1989). There resulted a prominence of the posterior thorax with an appendage extending upward from the mass toward the occiput and containing what was clearly two digits and a thumb. Radiography of the appendage showed bones characteristic of an upper extremity and scapula. The vertebral bodies appeared to be rotated so that the bodies were mostly dorsal, with the posterior arches situated ventrally. There was but one spine and spinal cord, but the dural sac on CT-metrizamide myelography was ectatic and the spinal cord appeared to split into two, becoming airophic caudally. When the mass was operated on, the spinal canal and cord were not exposed for the anatomical reasons noted. Pittman and Manwaring concluded that their patient had a disparate twin of the conjoined type, a designation which others have chosen in the past because of the intimacy of certain shared organ structures.

The possibility of a twinning phenomenon was considered by the geneticists who examined our patient, in part because of the knowledge that the mother had been taking Clomid. The drug manufacturer's studies of birth defects associated with this product found 58 birth defects among 2339 completed pregnancies. These occurred in 39 of 1676 infants from single pregnancies. Our patient's accessory limb had no shared organs with the surrounding body. While the cervical spinal canal was dysraphic and the cord contained a lipoma, the only connecting structures to the accessory limb (in addition to the covering skin) were an anomalous nerve and artery exiting the dural sac and running directly into the limb. Hence, our geneticists concluded that, as this drug stimulates ovulation, twins (if conceived) would arise from two eggs, not from the division of one egg as would have to be postulated in the case reported here. Thus, the drug was not likely responsible for this child's birth defect.

Many earlier cases of accessory limbs have been characterized by duplication of all parts at and below the lumbar region, or by duplication of both legs. In those instances of a single accessory leg, there were often features of duplication of pelvic organs, external genitalia, and mammary glands. Our case, like others recently reported, seems to represent an anomalous duplication of a limb and perhaps other nearby associated tissues, but without intimate sharing of organs vital to the child. Hanley and Stanitski have postulated that in these circumstances limb duplication has arisen from division of the limb bud which in amphibians, experimentally at least, results in duplication of the limb. They admit that such a theory does not satisfactorily explain those circumstances of incomplete duplication of all components of the limb. Nor would it explain why in our case the duplicated arm was not juxtaposed to one of the normal arms but located instead midway between the shoulders over the cervical spine.

As in the case reported by Jones and Larkin a century ago, our patient had posterior cervical spinal dysraphism which was intimately involved with the accessory arm. The previous case was remarkably sim-
ilar to ours: a 4-month-old infant had a supernumerary limb containing three fingers which arose from the spinal axis at the cervicothoracic junction. At operation, spina bifida was confirmed from C6-T2, and a meningocele sac and cerebrospinal fluid were encountered. The spinal cord “bifurcated” and there was a thick “nervous cord” exiting from the accessory limb. The patient died on the 3rd postoperative day. One might postulate that in their patient and in ours the accessory limb arose as a consequence of aberrant neurulation, perhaps with splitting of neural crest cells. Spinal dysraphism is associated with a variety of anomalies of non-neural organs, including mesodermal heterotopias (lipomatous masses and nephrotic tubules) in the spina bifida lesion and duplications (double vertebrae, double ribs, and duplication of ureters and kidneys). In support of his overdistention theory of the neural tube, Gardner proposed that if the distending neural tube ruptures beneath intact cutaneous ectoderm, the escaping neural tube fluid will infiltrate mesoderm. The resulting dislocation of cells and their possible injury by the extraneous protein may damage the as yet undentifiable anlagen of mesodermal organs. He further proposed that overdistention could result in duplication of external structures at either end of the embryo, supporting his theory with reported cases of anencephalics with cranial base and perineal bony soft tissue abnormalities. None of the data he presented, however, defines the formation of a “mature” appendage containing anatomically recognizable tissues.

**Conclusions**

The case of an accessory third arm reported here is unusual in its location on the posterior cervical spine and in its being distinct from the normal right and left arms. As such, it does not appear to have arisen as the result of a disparate twinning process or simple limb duplication. It is intriguing that the appendage was associated with posterior spinal dysraphism and cervical cord lipoma, from which it gained its legitimate neurovascular supply. That it should arise as part of the heterotopic or duplicating processes associated with spina bifida is a moot point.

**References**


Manuscript received April 11, 1990.
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