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ORIGINAL ARTICLE

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Accessory limbs associated with spina bifida – a second look

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Abstract An accessory limb associated with spina bifida was already reported by the authors. We had then described it as a result of a very early splitting of the limb bud arising from the paraxial mesoderm. We have subsequently seen three other such cases, which are described in this report as well as a review of five other cases in the literature. It is proposed that the growth of the accessory limb occurs from a mesodermal blastema that is a result of de-differentiation from Schwann cells.

Key words Accessory limb · Spina bifida · Lipomyelomeningocele

Introduction

The association of accessory limbs with spinal dysraphism was reported by the senior author (AK) in 1989, and we had claimed this to be the first such case [9]. Subsequently, we discovered another similar association described by Jones a century earlier in 1889 [8]. In our earlier paper, we had suggested that this anomaly results from a spontaneous aberration of morphogenesis. Since the limb bud develops from the mesoderm adjacent to the paraxial mesoderm, which gives rise to the dorsal vertebral arches, we proposed that very early splitting of one limb bud may result in an accessory limb with associated spinal dysraphism.

Since 1989, we have seen three more cases of a similar association. The experience with these cases as well as evaluation of five similar cases reported in the literature [7, 10, 12, 14] since our initial paper has prompted us to reconsider the possible mechanism that may result in such an anomaly.

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Case Reports

Case 1

A 10-month-old female born to a secundigravida was admitted with three spinal lesions: a small occipital meningocele, a sacral lipoma, and an aborted accessory limb in the mid-thoracic region (Fig. 1). The infant had normal developmental milestones. There was no neurological deficit or evidence of hydrocephalus. At operation, the accessory limb was composed of a lipomatous mass containing a long bone articulating with a scapula-like flat bone. The flat bone was astride the vertebral spines like a shield without any definite articulation. To the skeleton core of the limb muscles were attached, supplied by a neurovascular bundle. The accessory limb was removed and the lipoma was excised, taking care to preserve the spinal-cord tissue.

Case 2

An 18-month-old girl had a skin-covered lipomyelomeningocele overlying the lumbar spine. An outgrowth from this mass resembled an aborted hind limb. No other congenital anomalies were seen. The baby was neurologically normal. Radiologic work-up revealed a lipomyelomeningocele with intraspinal extension. The lipomatous mass was seen to contain a long bone (Fig. 2). The appendage was excised and the spinal lipoma was removed, taking care to avoid injury to the spinal cord. The mass contained a long bone with attached muscles and a neurovascular bundle.

Case 3

A 12-day-old female presented with a large, skin-covered mass overlying the lumbosacral region that was essentially lipomatous in consistency, but had a limblike structure growing out of it (Fig. 3). Also seen on its surface was a patch of opened-out bowel with glistening mucosa. At the center of the mass, close to where the limb was attached, a small protrusion with hyperpigmented skin resembled an aborted phallus (Fig. 4). The baby had no neurological deficit or evidence of any other congenital anomaly. Radiologic work-up confirmed a spina bifida lesion with intraspinal extension of the lipoma. The entire mass was excised with careful removal of the intraspinal lipoma. The dura was closed with a fascial graft. The excised mass contained a flat bone in articulation

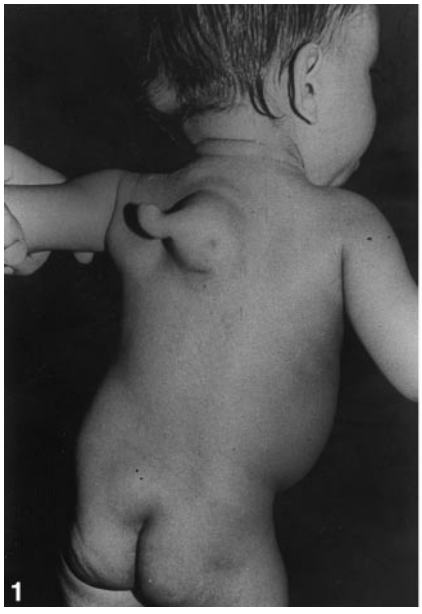

Fig. 1 Photograph of aborted accessory limb in interscapular region. Fully skin-covered lipoma has a skin pit. Note also small occipital meningocele and sacral lipoma

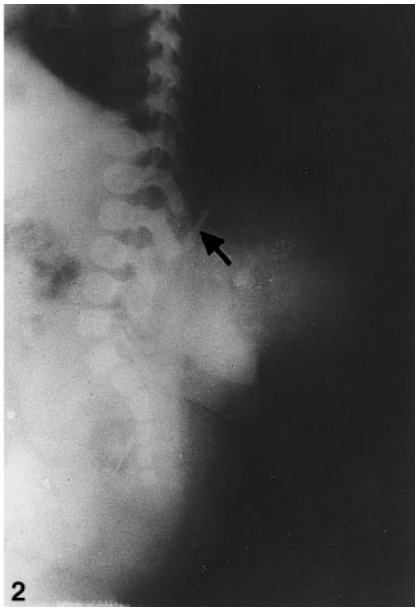

Fig. 2 Radiograph showing soft-tissue mass over lumbar spine containing a long bone (*arrow*)

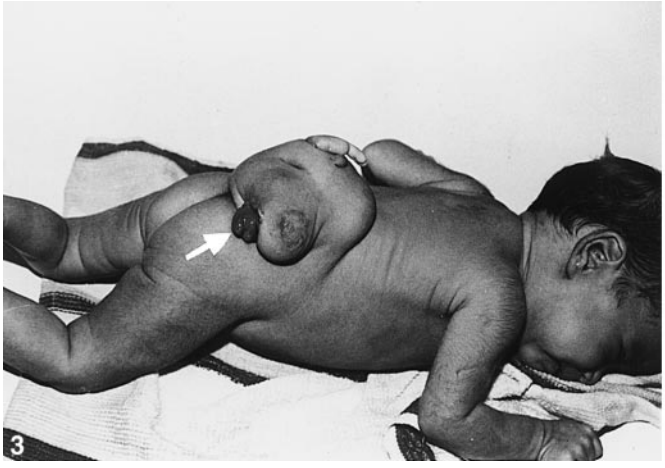

Fig. 3 Infant with large mass overlying lumbosacral spine. Note aborted limb at summit of mass and patch of colonic tissue (*arrow*)

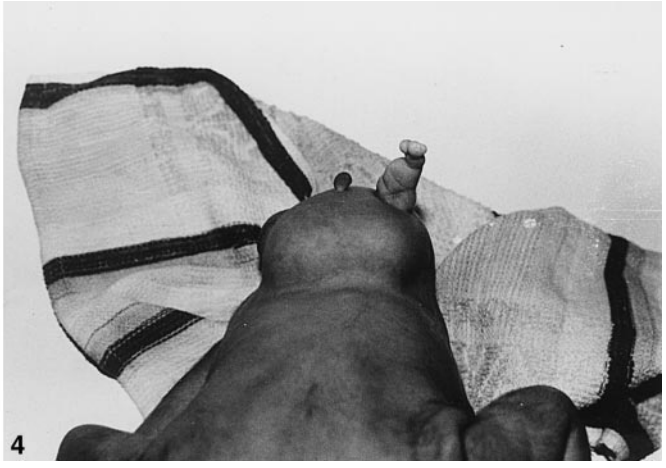

Fig. 4 Another view of aborted limb. Small central outgrowth close to origin of limb resembles a phallus

with a long bone that showed evidence of hemopoiesis. The phallus-like nubbin did not contain any identifiable urothelial structures. The isolated patch of bowel showed histologic features of a full-thickness segment of colon.

Discussion

Patients with a third leg have been frequently described in the literature [2, 6, 15, 16]. However, the association of accessory limbs with spinal dysraphism is rare. The first such association was reported in the last century [8]. In 1989, we described a similar case. Several names have been given to these limbs; intra-individual ipsilateral dipodia, heterotopic redundancies [11], aborted twinning, and teratoma are all inappropriate. Our own earlier term "tripedus" [9] was also incorrect, since it presumed that these accessory limbs could only occur in the region of the hind limb. The term "dysraphic appendage" used by Humphreys and Manwaring is descriptive and appealing [7].

Our evaluation of nine cases (including the present three) reported in this century [7, 9, 10, 12, 14] revealed that no antenatal insult has been clearly demonstrated as an etiologic factor. Interestingly, all but one [12] case were in female babies. One baby had an associated anorectal agenesis with a rectovestibular fistula [10]. The babies were born at term, mostly after a normal vaginal delivery except for one cesarean section [10]. Four infants had a second spinal lesion in addition to the dysraphic appendage. One of our patients (case 1) had two other neural-tube lesions. While in three babies the appendage was located in the interscapular region, in the other six it was lumbosacral. These accessory limbs lacked function except in one case, which showed involuntary flexion of the digits [7]. In three cases (including the present case 3) rudimentary external genitalia were seen [9, 12]. In one baby this was

connected to a urinary bladder within the accessory appendage [12]. Case 3 had an isolated segment of opened-out bowel histologically resembling colon on the surface of the accessory limb. In an earlier paper the presence of small bowel was also described [12].

All the dysraphic appendages were fully skin-covered with a good component of lipomatous tissue, often extending intradurally. It seems that accessory limbs develop in an association with a lipomyelomeningocele, since their association has not been seen with the usual variety of spina bifida cystica. In the four cases where other spinal lesions were also present, the accessory limb developed only in association with a fully skin-covered spinal lesion.

Our attempt to rationally explain the occurrence of this anomaly and the various observations that have been made with nine cases led us to the hypotheses proposed by Gardner [4] and Egar [3]. Gardner differentiated between primary and secondary neural-tube defects and suggested that while the former resulted from failure of the neural tube to close, the latter resulted from its rupture after closure due to oversecretion of neural-tube fluid. When this overexpanded neural tube ruptures beneath an intact cutaneous ectoderm, the proteinaceous neural-tube fluid infiltrates the mesoderm. Dislocation of cells occurs and is compounded by the injury caused by extraneous protein to the as yet unidentifiable anlagen of mesodermal organs. This interesting concept for secondary neural-tube defects may satisfactorily explain why most of our cases had a complete skin cover and why accessory limbs do not occur with the common variety of spina bifida cystica, which may be a primary neural-tube defect. The overdistension theory may also explain the very high occurrence (4/9) of more than one neural tube defect in the same baby.

In an animal experiment, Egar [3] produced accessory limbs in *Ambystoma mexicanum* (axolotls) by deflecting a large limb nerve to a proximal skin wound. The accessory limb grew only where the nerve exited between muscles and under the wound epidermis, and not where the cut end of the nerve was placed. The skeletal content of the mature accessory limb was variable, but in no instance was there evidence of attachment to the undisturbed skeleton of the normal limb. The muscles and nerves were present in the limbs in what appeared to be the normal arrangement. Quoting several other works, Egar suggested that "de-differentiation" of Schwann cells is the earliest source that gives rise to the mesenchymal core of the blastema. These blastemal cells accumulate under the dermis-free wound epidermis and grow into the accessory limb. The cut nerve seems to be the source of these Schwann cells.

Spinal lipomas, of which over 80% are lipomyelomeningoceles, are tumors comprised primarily of fat and a variety of other tissues (connective, cartilage, bone, neural) [13]. In our experience, cartilagenous pieces and bone spicules are often encountered in the lipomatous mass. These features, along with the fact that they are always skin-covered, put them in the category of occult spinal dysraphism, as distinct from spina bifida cystica.

Reviewing the hypotheses of Gardner [4] and Egar [3], it is plausible that lipomyelomeningoceles are secondary neural-tube defects that occur due to rupture of the neural tube under intact ectoderm. The leakage of the proteinaceous neural tube fluid acts as an abundant source of Schwann cells, which collect under the skin and de-differentiate. The majority of de-differentiated cells develop into a lipomatous mass that may extend intradurally. Sometimes cartilage, bone, muscles, and nerves may also develop in this lipomatous mass. On rare occasions, these components may grow in an organized manner and develop into an accessory limb. The stimulus for this organized growth remains uncertain. Even more baffling is the fact that this anomaly has such a predominant female predilection. Also, the presence of colon-like tissue in one of our cases is unusual, although ectopic metanephric tissue has been seen in association with diastematomyelia [5] and with a lumbosacral lipoma with intradural extension [1]. These cases may represent an extreme degree of de-differentiation of the blastemal cells.

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