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CASE REPORT

Accessory lower limb associated with spina bifida: case report

Yaşar Bayri · Bahattin Tanrıkulu · Murat Şakir Ekşi · Adnan Dağçınar

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Abstract
Purpose Heterotopic redundancies, such as an accessory limb associated with spina bifida, are extremely rare anomalies. There are 12 cases of accessory limb associated with spinal bifida in literature. This report aims a detailed description of the additional case and an analysis of the findings in light.
Methods A male baby was born at 40 weeks of gestation and was referred to the neurosurgery clinic with a diagnosis of accessory lower limb. On physical examination, the dorsal meningocele was located at the lumbosacral region and there was accessory lower limb on it. There was no open neural placode.
Results The accessory limb was excised on postnatal day 3.
Conclusıons Dysraphic appendages are rare and complicated anomalies. They should be investigated carefully, and all of the lesions must be repaired for babies' quality of life.

Keywords Accessory limb · Spina bifida · Heterotopic redundancy · Dysraphic appendage

Introduction

Heterotopic redundancies, such as an accessory limb associated with spina bifida, are extremely rare anomalies. An accessory limb associated with spinal bifida was first reported by Jones in 1889 [4]. Since then, only 12 cases have been reported in the literature [2, 3, 5–11] (Table 1). This report aims a detailed description of the additional case and an analysis of the findings in light of the previous findings and current theories.

Y. Bayri (✉) · B. Tanrıkulu · A. Dağçınar
Faculty of Medicine, Department of Neurosurgery, Marmara University, Marmara Üniversitesi Tıp Fakültesi, Selimiye M. Tıbbiye C. No:38, 34668, Haydarpaşa Istanbul, Turkey
e-mail: ybayri2000@yahoo.com

M. Ş. Ekşi
Department of Orthopaedic Surgery, University of California, 521 Parnassus Ave., San Francisco, CA 94117, USA

Case report

A baby boy was delivered by Cesarean section at 40 weeks of gestation and was referred to the neurosurgery clinic with a diagnosis of accessory limb associated with spina bifida. The mother had undergone routine pregnancy follow-up, and a diagnosis of spina bifida was made at 20 weeks of gestation. On physical examination, the dorsal meningocele was located at the lumbosacral region and the accessory limb was seated on it, pointing cranially (Fig. 1). No open neural placode was noted. Spinal reflexes, such as withdrawal to pain, were present in the accessory limb. The limb resembled a normal leg, ending with a rudimentary foot containing only one toe. The normal legs of the patient were morphologically, functionally, and neurologically intact. The baby boy had normal looking external genitalia, anus, abdominal and thoracal walls. The head circumference was 35.5 cm (60th percentile) with normal appearance.

Radiological findings were as follows: On plain x-ray and computed tomography, unfused posterior vertebral arches were detected between L1-S1 segments. The accessory limb was attached to a rudimentary posterior vertebral arch by rudimentary hemi-pelvis at L5-S1 level (Fig. 2). The leg was arising from lumbosacral region in the midline and containing two long bones resembling femur, tibia, and rudimentary foot bones. Magnetic resonance examination of the spine disclosed a lumbosacral meningocele with a low-lying conus (Fig. 3). The cranial magnetic resonance investigation was within normal limits with no associated cerebral anomalies.

Table 1 Reported cases of dysraphic appendages

Reference	*n*	Gender	Resembling. lower or upper extremity	Localization of appendage	Spina bifida morphology and localization	Accompanied anomaly
Jones and Larkin [4]	1	F	U	Thoracal	Meningocele (thoracolumbar)	–
Krishra et al. [6]	1	F	L	Lumbosacral	Meningocele (sacral)	Rudimental phallus and scrotal skin
Sharma et al. [11]	1	F	U	Thoracal	Meningocele (lumbar)	–
	1	F	U	Thoracal	Lipomyelocele (lumbar)	–
Parkinson [10]	1	M	L	Lumbosacral	Meningocele	Rudimentary external genitalia Rudimentary bowel
Humphreys et al. [3]	1	F	U	Cervical	Lipoma	–
Nanni et al. [9]	1	F	L	Sacral	Myelomeningocele (thoracolumbosacral)	Anorectal agenesis and rectovestibular fistula
Krishna and Lal [5]	1	F	U	Thoracal	Lipoma (sacral)	–
	1	F	L	Lumbar	Lipomyelomeningocele (lumbosacral)	–
	1	F	L	Lumbar	Lipomyelomeningocele (lumbosacral)	Rudimentary external genitalia Rudimentary bowel on the surface of the limb
Gamanagatti et al. [2]	1	?	L	Sacral	Lipomyelocele (sacral)	–
Lende et al. [8]	1	F	L	Thoracal	Myelomeningocele (lumbar)	Isolated intestinal ectopic loop
Kumar [7]	1	M	L	Thoracolumbar	Lipomyelomeningocele Split cord malformation type 1	Rudimentary external genitalia Multiple vertebral anomalies Chiari 1 malformation
Bayri et al. (current case)	1	M	L	Lumbosacral	Meningocele (lumbar)	–

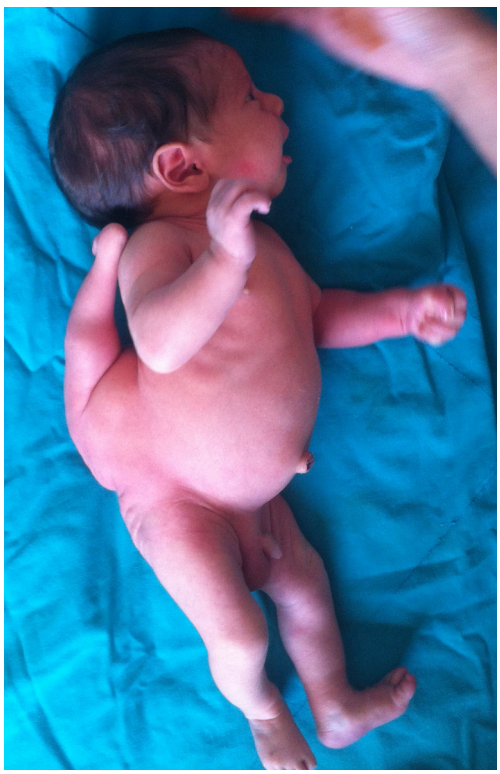

Fig. 1 The dorsal meningocele was located at the lumbosacral region and the accessory limb was seated on it, pointing cranially

The patient was operated on postnatal day 3. A circumferential skin incision was made around the skin plica created by the

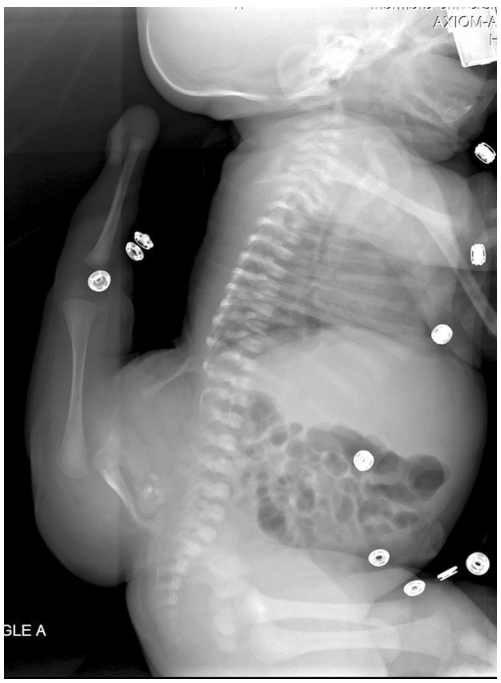

Fig. 2 An x-ray image showing the accessory limb which was attached to a rudimentary posterior vertebral arch by rudimentary hemi-pelvis at L5-S1 level

accessory limb. Incision was deepened until the attachment point of rudimentary pelvis was revealed. A thick peripheral nerve arising from the vertebral column was entering and innervating the accessory limb. A pair of arteries was accompanying the nerve. This neurovascular bundle was cut. The accessory limb was separated from its attachment side and excised. The dorsolumbar fascia was found to be defective in the right paravertebral region and repaired primarily. Open posterior vertebral arches were observed between L1-S1 levels when the vertebral column was checked. Dura mater was anatomically intact. Dura was opened and some attachments between cord and dura were separated, and the tethered filum was divided.

Pathological examination of the limb revealed that the limb contained grossly normal looking muscular layers and bones resembling normal anatomy (Fig. 4).

The patient was discharged in the seventh postoperative day without neurological deficit and well-healing incision line. In his routine controls, he was found to have normal growth and development.

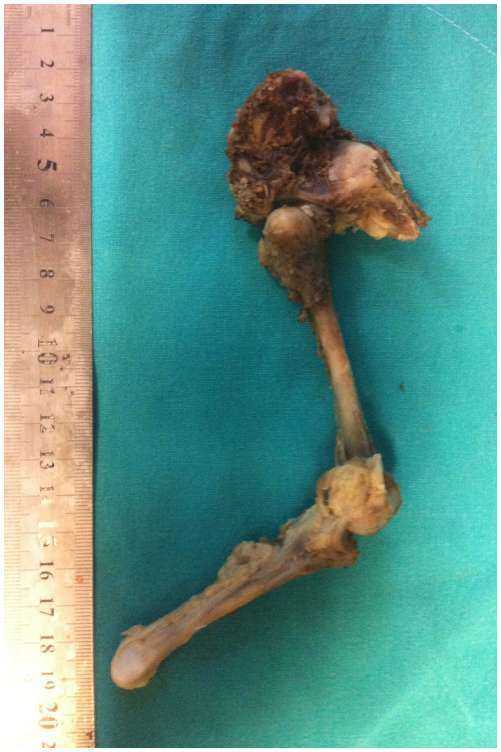

Fig. 4 Gross pathological examination: the limb contained grossly normal looking muscular layers and bones resembling normal anatomy

Discussion

The association of developmental spinal anomalies with other developmental systemic anomalies has been extensively studied [1]. Disrupted development of the caudal cell mass may be associated with lesions of lesions of close proximity such as the urogenital tract, anorectal structures. Alternatively; disordered notochord development may be associated with anomalies of the thoracic and abdominal viscera as well as the neural tube. However, for some developmental spinal anomalies, even a theoretical explanation of the embryogenesis is not possible: Heterotopic redundancies, such as an accessory-limb associated with spina-bifida, are such an anomaly. This is an extremely rare anomaly, and only 13 cases have been reported so far [2–11]. The co-occurrence of the accessory limb together with spina bifida strongly hints to a common pathogenetic mechanism, which is currently unknown. This report aims to add to the existing body of knowledge of this extremely rare anomaly.

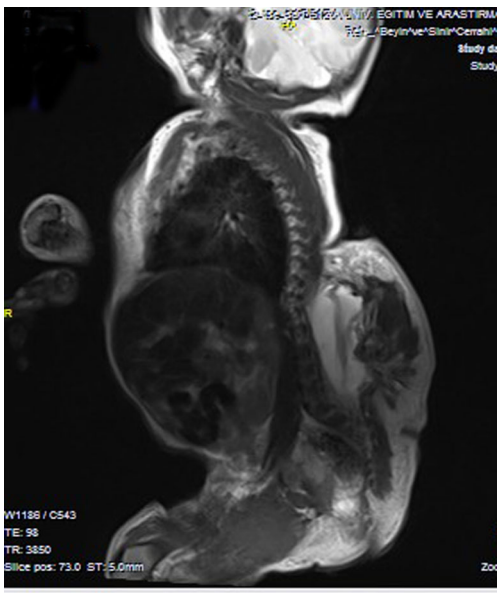

Fig. 3 The magnetic resonance image of the spine showing a lumbosacral meningocele with a low-lying conus

Several names have been offered to these anomalies; tripedus, aborted twinning, teratoma, and heterotopic redundancies [5] are used but not appropriate. The term ‘dysraphic appendage’ was used by Humphreys et al. [3] and was found to be most descriptive by Krishna [5] who has the largest series. We also agree with him that it appeals the malformations in both contents.

An analysis of the cases (including the present one) has revealed the following characteristics: Most of the babies born with this anomaly are female (11/14). Except for one case with oral contraceptive usage in the first trimester no gestational insult has been reported in association with this anomaly [8]. The appendages were located in the cervical region in one (7 %), thoracic-interscapular in five (35 %), lumbar in three (22 %), lumbosacral in three (22 %), and sacral in two (14 %) cases. Four of the thoracic lesions resembled rudimentary upper extremities and the remaining resembled lower extremities. The reported lesions contained bone, muscle, cartilage

tissues, and formed organs with close but incomplete resemblance of normal anatomy.

Neural tube defects associated so far with accessory limbs can take three forms: Five (36 %) of the lesions were meningoceles, two (14 %) myelomeningoceles, and seven (50 %) spinal lipomas (lipomyeloce, lipomyelomeningocele, lipoma). Both myelomeningoceles and lipomas of the spinal cord are hypothesized to arise as a result of faulty neurulation. In patients with lipomas, premature separation of the cutaneous ectoderm from neural ectoderm is hypothesized to cause interaction of the surrounding mesenchyma with interior of the cord, namely the central canal. Existence of mesenchymal tissue prevents the neural placode fusing in the dorsal midline and mesenchyma itself differentiates to fat. Barkovich suggested that the interaction of the same mesenchyme with the exterior of the cord would result in abnormal meningeal, bone and muscle tissue, which may be the cause of the ectopic appendages [1]. Krishna et al. [5] suggested that the process may be explained with Gardner's theory: namely, an overexpansion and resultant rupture of the neural tube due to oversecretion of neural tube fluid. After closure of the midline ectoderm, ruptured neural tube would lead to infiltration of the proteinaceous neural tube fluid to the mesoderm. Dislocation of mesenchymal cells and interaction with neural tube fluid would results in abnormal heterotopic appendages. Such appendages contain muscle, fat, and bone tissue differentiated from mesenchymal cells. The theory also is valuable when we try to explain of dysraphic lesions accompanying ectopic appendages. In the case of lipomas, interaction of neural tube fluid with mesenchymal cells lead these cells to differentiate to lipomatous tissue. In fact, both of the theories try to explain how the mesenchyme and neural ectoderm interacts, but the final pathway is same.

If rupture of the neural tube occurs before the dysjunction or the separation of cutaneous and neural ectoderm cannot be accomplished, central canal and mesenchymal tissue cannot interact and myelomeningocele may take place.

It is more hard to explain of occurrence of meningocele lesions because of their origin cannot be easily defined. But in most of the cases described, meningocele may not be the real one and can be accepted as an enlargement of dural sac secondary to enlarged spinal canal caused by absent posterior elements, just like in our case.

Other systemic anomalies were also common in the reported cases: Rudimentary external genitalia were accompanying lesions in four (28 %) babies [5, 7, 10]. Ectopic intestinal loops could be identified under the skin around the extra limb in three (22 %) patients [5, 8, 10]. Anorectal agenesis and rectovestibular fistula were observed in one case (7 %) [9, 10]. In another baby Chiari type 1 malformation and multiple vertebral anomalies were detected[7].

Conclusion

Dysraphic appendages are very rare, complicated anomalies. They should be investigated carefully and all of the lesions must be repaired for babies' quality of life.

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