

Vestigial Accessory Limbs with Spina Bifida: Our 5 – Year Experience

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ABSTRACT

Background: Accessory limb on the back of trunk with spina bifida is an exceedingly rare entity which has perplexing morphogenesis. We are sharing our 5 – years' experience of managing 5 patients with accessory vestigial limbs on the back associated with spina bifida.

Materials and Methods: The medical record of 5 patients with accessory limbs on the back of the trunk was reviewed for mode of presentation, investigations, management, and outcome.

Results: There were 5 infants, one was male while rest were females (M: F = 1: 4). Ages ranged from 1.5month to 1 year (mean age 6.3 months). All patients presented with a vestigial limb attached to the lumbosacral region. One patient had anal pit like depression on the vestigial accessory limb. Every patient had associated spina bifida. One patient had associated lipomenigocele. Another patient had associated right clubfoot. All patients were investigated with X-rays and MRI and underwent excision of the accessory limbs. One patient developed wound dehiscence and CSF leak post-operatively which settled on conservative management. Still another patient had repeated wound infections and is booked for release of tethered spinal cord. All patients survived. Two patients required physiotherapy for lower limb weakness.

Conclusion: Accessory limbs on the back are quite rare anomalies and are associated with a number of other anomalies. It is more common in females. The anomaly is a psychosocial dilemma for the parents. Early excision is necessary to allay the anxiety of the parents.

Keyword: *Rachypagus, Partial twinning, Accessory limb, Spina bifida.*

INTRODUCTION

Accessory limbs on the back of trunk also known as *rachypagus* are rare which are associated with spina bifida. It was first described by Deslongchamps in 1851.¹ Very few case reports have been published so far. It has a perplexing morphogenesis and literature revolves around its differentiation from Teratoma or Fetus-in-Fetus.¹⁻⁴ We herein describe an experience with 5 cases of lumbosacral rachypagy associated with spina bifida.

MATERIALS AND METHODS

It is a retrospective case series performed in the depar-

ment of Pediatric Neurosurgery at The Children's Hospital and the Institute of Child Health, Lahore, Pakistan between Jan-2011 to Jan 2016. The medical record of infants with accessory limbs on the back, managed in our department, was reviewed for the mode of presentation, investigations, management, and outcome.

RESULTS

There were 5 infants, one was male while rest were females (M : F = 1 : 4). Ages ranged from 1.5month to 1 year (Median age 6.3 months). All patients presented with a vestigial limb attached to the lumbosacral reg-

Table 1: Summary of study population.

No. of Patients	Age / Sex	Presentation	Type of Anomaly	Complications	Outcome
1.	12 m/M	Accessory limb attached to lumbosacral area + clubfoot	Accessory limb with spina bifida+ scrotum and phallus like structures	Nil	Survived
2.	8 m/F	Accessory limb attached to lumbosacral area + incontinence + B/L weakness of lower limbs	Accessory limb with spina bifida	Nil	Survived
3.	6 m/F	Accessory limb attached to lumbosacral area	Accessory limb with spina bifida+ lipomeningocele	Wound dehiscence with CSF leak	Survived
4.	4.5 m/F	Accessory limb attached to lumbosacral area + left leg weakness +urinary incontinence	Accessory limb with spina bifida+gut like mucosal tissue	Increased weakness of left lower limb and incontinence	Booked for untethering
5.	1.5 m/F	Accessory limb attached to lumbosacral area	Accessory limb with spina bifida	Nil	Survived

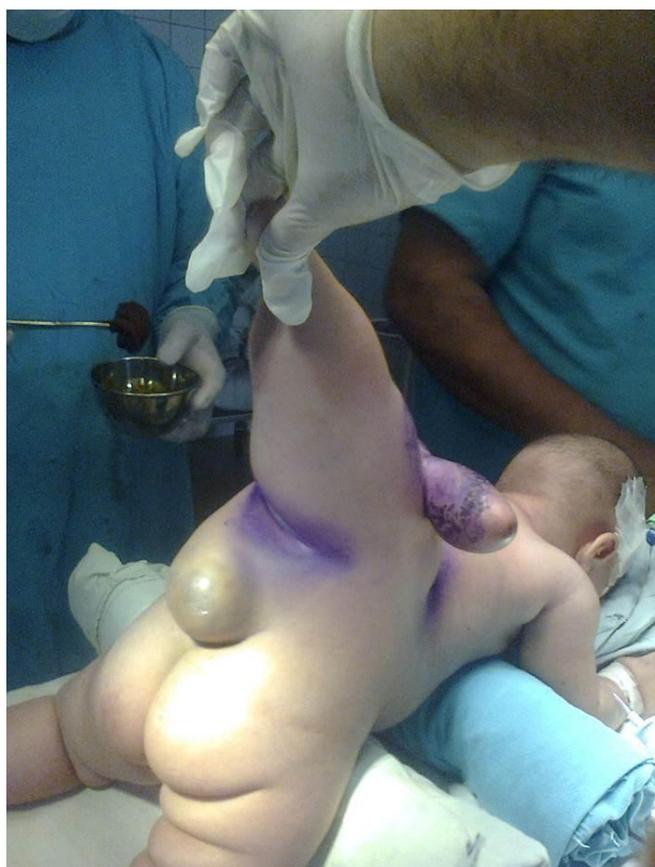


Fig. 1: Accessory limb: Patient had associated lipomeningocele.

ion (Fig. 1 – 3). One patient had anal pit like depression on the vestigial accessory limb. Another patient had scrotum and phallus – like tissue attached to its lower side (Fig. 4). Still another patient had gut – like blind ending mucosal tube attached to the accessory limb (Fig. 5). Every patient had associated spina bifida. One patient had associated additional lipomeningocele, while one patient had associated right clubfoot. Accessory limb had no movements, pulsations in each case. One patient was incontinent with bilateral lower limb weakness while one other patient had urinary incontinence with left lower limb weakness. Lower limb movements in the remaining three patients was intact.

All patients were investigated with X-rays and MRI and underwent excision of the accessory limbs and untethering of nervous tissue, along with repair of lipomeningocele in one patient. One patient developed wound dehiscence and CSF leak post-operatively which settled on conservative management. Another patient had repeated wound infections and is booked for release of tethered spinal cord. All patients survived. Two patients required physiotherapy for lower limb weakness. Table 1 shows summary of study population.



Fig. 2: Accessory limb.



Fig. 4: Accessory limb with scrotum and phallus like tissue on lower side.



Fig. 3: Excised accessory limb in another patient.



Fig. 5: Gut mucosal blind ending tube attached to accessory limb.

DISCUSSION

The etio-pathogenesis of abortive rachypagus or accessory limb attached to the back of trunk may be attributed to modified aborted process in twinning. However, the exact morphogenesis is unknown. As the limb buds develop from mesoderm adjacent to paraxial mesoderm and a very early splitting of one limb bud may result in this anomaly. The migrated mass of tissue formed at very early stage may cause mechanical interference with closure of neural tube which results in associated spina bifida.²⁻⁵ A case of accessory limb attached to thigh has been reported and the authors have hypothesized about late splitting of the limb bud.⁶ Few authors are in favor calling it a teratoma or fetus-in-fetus, but this assumption could not gain much popularity.⁷

The accessory limb can be attached to any part of the back ranging from inter-scapular region to sacrum. In our series, all patients had accessory limb attached to lumbosacral region. Our all patients also exhibit spina bifida as documented on x-rays spine and MRI. MRI also depicted tethering of spinal cord in all patients and lipomenigocele in one patient. X-rays depicted various malformed bones in all cases. Some resembled femur and tibia and others to unidentified bony tissues with malformed articulation. Variable numbers of phalanges were also present in our patients on accessory limb radiography.

Various other anomalies are associated with rachypagus. These include a case of teratoma in addition to accessory limb, lipoma, meningocele, meningomyelocele, ventricular septal defects, and club foot etc.⁴⁻¹¹ In our series we had club foot in one patient, lipomenigocele in one patient, and a gut mucosa – like blind ending tube in still another patient, which sometimes make it very hard to differentiate it from a teratoma. Few authors reported presence of anal pits, scrotum, and phallus within the accessory limb. In our series, we too had one patient with multiple anal pits, and one patient with scrotum and phallus attached to the accessory limb.

The attached limb never have movements in it but sensory activity has been reported in response to painful stimulus, in the form of crying, on pinching the accessory limbs.⁴⁻⁶ In our series too, none of the patients had movements in the accessory limbs. The patients may have associated incontinence and weakness of lower limbs as depicted by two of our patients in this series. This is attributed to associated tethering of spinal cord. Early untethering may help in improving these parameters. These patients are on combined fol-

low-up of physiotherapy, orthopedics and pediatric neurosurgery.

The ultimate management is surgical excision with untethering of nervous tissue and coverage of the defect with myocutaneous tissue.⁴⁻⁷ Removal of these accessory limbs is very similar to the excision of huge lipomenigocele. It may require a transfusion of 250 – 500 ml of blood during the procedure. The average time for the surgery was about 90 minutes. In all of our cases, no difficulty was encountered in excising these structures. Postoperatively various wound-related problems were encountered including dehiscence and CSF leakage. In still another patient, adhesions were formed between spinal cord and dura along the scar line as depicted on postoperative MRI performed for increasing weakness of leg and urinary incontinence in the postoperative period.

CONCLUSION

Accessory limb, attached to back of trunk, is a rare congenital anomaly with only a few cases reported so far. It is a strange anomaly with psychosocial issues for the family. Early intervention is required to relieve anxiety of parents. Multidisciplinary approach is required to deal with associated problems. As these vestigial limbs can be removed very successfully without causing harm to the useful limbs, so there is need to create awareness amongst masses that these children should not be handed over for exhibition in circuses to amuse other people.

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