An unusual case of lipomeningocoele with pseudo tail as cutaneous marker

Charu Tiwari[1], Hemanshi Shah[2], Jyoti Bothra[3], Gursev Sandlas[4], Mukta Waghmare[5]
[1] Registrar, [2] Professor and Head, [3] Registrar, [4] Assistant Professor, [5] Registrar, Dept of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai, Maharashtra. India. Pin: 400008

ABSTRACT

Tail-like caudal appendages are rare and have been reported to be associated with spinal dysraphism, especially with spinal lipomas or lipomyelomeningoceles. We present the case of an 11 year-old girl with human tail and lipomeningocoele.

Key words: Human tail, Lipomeningocoele, Tethering, Dysraphism

Corresponding author address: Hemanshi Shah, Professor and Head, Dept of Paediatric Surgery, TNMC and BYL Nair Hospital, Mumbai, Maharashtra. India. Pin: 400012.
Phone: 09004764291, **M:** 900476429 **E-Mail:** drcharusharma18@gmail.com

Conflict of interest: No

Case report is Original: YES

Whether case report publishes any where? NO

INTRODUCTION

A human tail or caudal appendage is a rare and peculiar condition which is frequently considered to be the cutaneous marker of occult spinal dysraphisms [1, 2]. Several studies have reported human tails causing tethered cord syndrome [1, 3, 4]. A human tail was initially thought to be an evidence of man's descent from or relation to other animals. It has even been made a subject of superstitious beliefs, especially in Asia [1]. We present a case of human tail in an 11 year old girl. The embryology, modes of presentation and management strategies has been highlighted in the discussion.

CASE REPORT

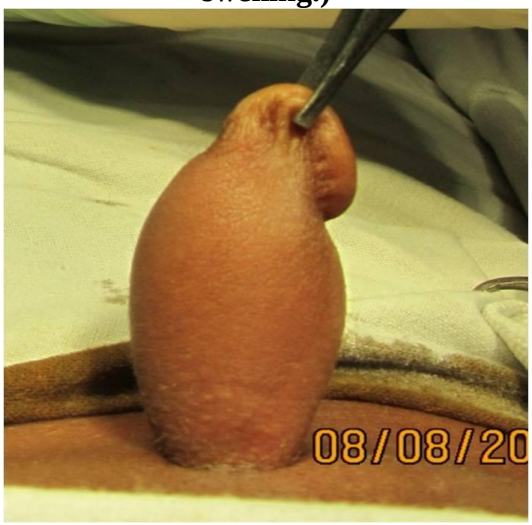
An 11year-old girl presented with swelling in the lower back since birth. There were no neurological complaints.

A single, elongated, soft skin covered swelling measuring 6x3cms protruding from the sacral regionand tapering distally was seen. Overlying and surrounding skin was normal. There was no neurological deficit.

MRI (Magnetic Resonance Imaging) showed lipomatous swelling at level of L5 – S1 with few nerve fibres entering in the lateral aspect suggesting lipomeningocele.

The swelling was excised taking an elliptical incision around its base. There was no communication with the dura. The patient is asymptomatic on follow-up.

Figure 1 (a): Clinical photograph of the swelling.)



(Figure 1 (b): Clinical photograph of the swelling.)

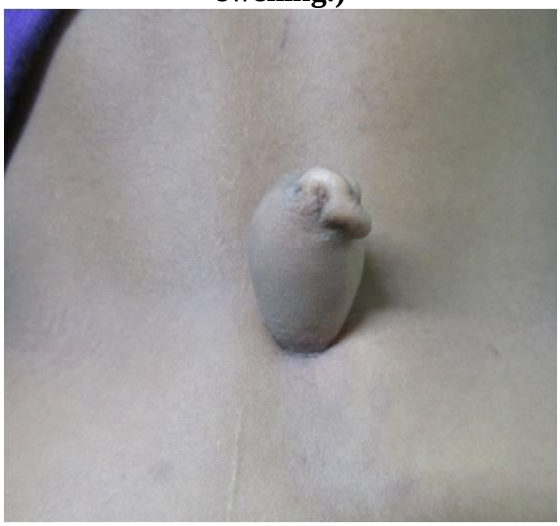
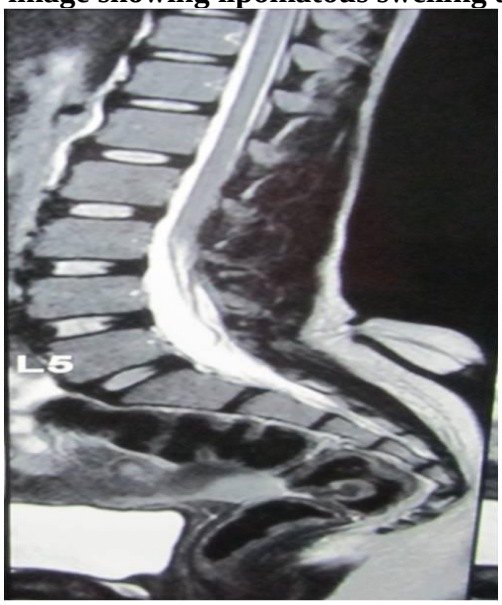


Figure 2: MRI image showing lipomatous swelling at L5-S1 level.)



DISCUSSION

A human tail is a remnant of an embryological structure [5]. At 5-6 weeks of gestation, the human embryo has a conspicuous tail with 10-12 vertebrae protruding from the trunk [1]. This tail regresses by fusion of vertebrae and disappears by the 8th week, leaving the vestigial coccyx [1]. The vestigial human tail probably arises from the distal, non-vertebrate portion of the embryonic tail found at this stage of gestation [5, 6].

However, the current accepted concept of embryology of this malformation states that the tail is a form of congenital dermal sinus tract which results from premature or incomplete

disjunction of the somatic and neuroectoderm during primary neurulation [1, 7]. Another theory states that it might be caused due to disorder of secondary neurulation [1]. The secondary neurulation with formation of tail bud begins prior to completion of primary neurulation; hence a disorder of the secondary neural tube or notochord might possibly affect the closure of neuropore resulting in spinal cord lipoma and abnormal tail bud regression [1]. An early disorder usually results in spinal lipoma and tethering and later disorders cause persistence of tail bud without spinal anomalies [1, 8]. The human tail associated with lipomeningomyelocele has its origin in secondary neurulation while the lipomeningomyelocele develops from defective primary neurulation [1].

Bartels have classified human tail into five types - First three are simple variations of the soft tail without any vertebral components [9]. The fourth type is bony hypertrophy of the sacrococcygeal vertebrae [9]. The fifth is a true animal tail containing additional vertebrae [9].Dao and Netsky classified them into true tail and pseudo tail [10]. True human tail contains adipose tissue, connective tissue, striated muscle, blood vessels and nerves and is covered by skin [5]. There is no element of bone (unlike animal tails), cartilage, notochord or spinal cord [5]. It arises from the most distal remnant of the embryonic tailcan move and contract [5]. Males are affected twice as females [5]. A true tail requires simple surgical excision [5].Pseudo tail resembles superficially to true tail and is an anomalous prolongation of the coccygeal vertebrae [5]. Additional lesions like lipomas, teratomas, chondromegaly and gliomas, and parasitic fetus may be present with pseudo tails [5, 10]. However, this classification system has embryological importance without clinical significance [1]. A more practical and clinically relevant classification of human tail was given by Lu et al based on the association of human tail with tethered cord [1, 3]. A true tail is a benign condition and a prolongation beyond the coccygeal or midgluteal region, and not associated with any underlying neural malformation [1]. A pseudotail occurs in association with spina bifida occulta or spinal dysraphism [1].Thus, a pseudotail is only a cutaneous sign of underlying spinal dysraphism [1].

Human tail, though usually occurs in the lumbosacral region, has also been reported in the cervical region[5, 11]. Lumbosacral and sacrococcygeal teratomas have also been reported in the tail [5, 12].The lengths of caudal appendage reported range from 1 to 20 cm [1, 8]. Associated cutaneous abnormalities such as hypertrichosis, hyperpigmentation, dermal sinus tracts and hemangiomas have also been reported [5, 13]. Rarely, the cutaneous appendages itself may have a continuity with intraspinal lipoma causing tethering of the cord [1, 3, 4, 11]. A case of bony human tail associated with tethered cord has also been reported [1, 14]. Hence, presence of tethering should always be kept in mind before considering surgery [1].

The preoperative assessment should include a complete clinical examination - neurology, plain radiographs of the spine and computed tomography or magnetic resonance imaging to rule out occult spinal dysraphism, intraspinal lesions, syringomyelia, lipomeningocele, and tethered cord in these cases. Occasionally, associated malformations like congenital heart disease, anal and vaginal atresia, and horseshoe kidney have also been reported to be present with such disorders and so appropriate preoperative evaluation is required [1, 15].

Early prophylactic un-tethering of spinal cord if present is recommended to avoid neurological deficit. Microsurgical treatment then becomes necessary to prevent tethered cord syndrome, even if there are no preoperative neurological signs and symptoms [1]. Intra-operative stimulation of sphincter muscles is helpful.

CONCLUSION:

Human tail or caudal appendage is a rare condition which is usually associated with spinal dysraphism. Neuroradiological evaluation and appropriate management is required for better outcome.

REFERENCES

1. Sadashiva N, Beniwal M, Shukla D, Srinivas D. A tale of two "tails:" A curiosity revisited. J PediatrNeurosci. 2016 Apr-Jun; 11(2): 153–155. doi: 10.4103/1817-1745.187647.
2. Humphreys RP. Clinical evaluation of cutaneous lesions of the back: Spinal signatures that do not go away. ClinNeurosurg. 1996;43:175–87.
3. Lu FL, Wang PJ, Teng RJ, Yau KI. The human tail. Pediatr Neurol. 1998;19:230–3.
4. Yamada S, Mandybur GT, Thompson JR. Dorsal midline proboscis associated with diastematomyelia and tethered cord syndrome. Case report. J Neurosurg. 1996;85:709–12.
5. Mukhopadhyay B, Shukla RM, Mukhopadhyay M, Mandal KC, Haldar P, Benare A. Spectrum of human tails: A report of six cases. J Indian AssocPediatr Surg. 2012 Jan-Mar; 17(1): 23–25. doi: 10.4103/0971-9261.91082.
6. Zimmer EZ, Bronshtein M. Early sonographic findings suggestive of human fetal tail. PrenatDiagn. 1996;16:360–2.
7. James HE, Canty TG. Human tails and associated spinal anomalies. ClinPediatr (Phila) 1995;34:286–8
8. Donovan DJ, Pedersen RC. Human tail with noncontiguous intraspinal lipoma and spinal cord tethering: Case report and embryologic discussion. PediatrNeurosurg. 2005;41:35–40.
9. Baruchin AM, Mahler D, Hauben DJ, Rosenberg L. The human caudal appendage (human tail). Br J Plastic Surg 1983;36:120-123.
10. Dao AH, Netsky MG. Human tails and pseudo tails. Hum Pathol. 1984;15:449–53.
11. Mohindra S. The 'human tail' causing tethered cervical cord. Spinal Cord. 2007;45:583–5.
12. Mc Lone DG, Naidich TP. Terminal myelocystocele. Neurosurgery. 1985;16:36–43.
13. Park SH, Huh JS, Cho KH, Shin YS, Kim SH, Ahn YH, et al. Teratoma in human tail lipoma. PediatrNeurosurg. 2005;41:158–61.
14. Muthukumar N. A bony human tail causing tethered cord syndrome: Case report. Childs Nerv Syst.2014;30:703–7
15. Alashari M, Torakawa J. True tail in a newborn. PediatrDermatol. 1995;12:263–6.