Case Report

Perianal Human Tail with Lower Imperforate Anus: A Case Report

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Abstract

The human tail is usually attached to the sacrococcygeal skin. However, few reports of the human tail at the perineal or perianal region have been described. Moreover, the tails are rarely accompanied with anorectal malformations (ARMs). Although the exact relationship between the human tail at the perineal or perianal region and ARM remains obscure, we consider that the human tail at the sacrococcygeal region and the perineal or perianal region may have a slightly different mechanism of development.

Keywords: Anorectal malformation, human tail, imperforate anus, perianal

Introduction

The congenital anomaly referred to as 'human tail', i.e., the occurrence of a tail-like structure has been reported and was thought to represent a vestige of the embryonic tail; however, this hypothesis has not been verified.[1-4] Human tail is usually attached to the sacrococcygeal skin. However, few reports of the human tail at the perineal or perianal region have been described. Anorectal malformations (ARMs) are also relatively uncommon congenital defects that often occur in conjunction with syndromes or other congenital abnormalities. It is more interesting that the tails at the perineal or perianal region are rarely accompanied with ARM in our case as well.[5-9] Therefore, we consider that the human tail at the sacrococcygeal region and the perineal or perianal region may have a slightly different mechanism of development. We report an extremely rare case of perianal human tail with lower imperforate anus.

Case Report

Prenatal ultrasonography had revealed a normal amount of amniotic fluid and no abnormality. The patient was born at 39 weeks and 0 days gestational age by spontaneous vaginal delivery. Birth weight was 2884 g, and Apgar scores at 1 and 5 min after birth were 10 and 10, respectively. A female newborn presented with a perineal mass measuring 3 cm in length located just to the right of an anal dimple but with no anal opening. There was a perineal fistula [Figure 1a]. The mass was soft and was completely covered by skin. The neurological examination was normal, as was the rest of the physical examination. A detailed examination revealed no other associated anomalies. A diagnosis of lower imperforate anus accompanied by human tail was made. Air and contrast enemas were performed through the perineal fistula and revealed a narrow fistulous tract and distended rectum above the expected location of the anus [Figure 1b]. Magnetic resonance imaging (MRI) of the spine showed no evidence of spinal dysraphism. MRI revealed a pedunculated mass with predominantly fat intensity and a single central vessel and confirmed the diagnosis of a true human tail [Figure 1c and d]. Preoperatively, Hegar dilatation and a glycerine enema were used for initial management through the fistula. Because defecation control was relatively good after oral feeding was initiated, excision of the mass and anterior sagittal anorectoplasty were performed at 14 months of age. The fibromuscular band was found to extend toward the external anal sphincter, and it was transected. Histopathological findings revealed the tail consisted of adipose tissue and blood vessels with striated muscular fibres [Figure 1e]. The post-operative course was uneventful. At 8-year follow-up, the wound had healed well and no complications such as constipation or anal stenosis occurred.

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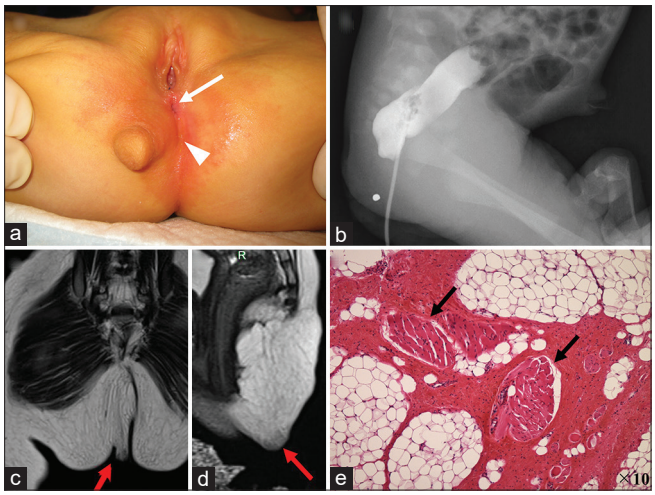


Figure 1: (a) Clinical photograph of a female newborn showing a mass extending from the perineal region. A 3 cm perineal mass located in the right at an anal dimple. (Arrow: perineal fistula, arrow head: anal dimple). (b) Air and contrast enemas revealed the narrow fistulous tract and distended rectum above the expected location of the anus. (c) Coronal T1-weighted MRI scan through the sacrum showing the fat containing a true human tail (arrow). (d) Sagittal T2-weighted MRI scan showing no evidence of spinal dysraphism, and dilated rectum (R) in this patient with known imperforate anus. The perianal mass (arrow) indicating a signal intensity consistent with fat located close to anal dimple. (e) Hisopathological findings revealed the tail consisted of adipose tissue, blood vessels with striated muscular fibres. (Arrows were striated muscular fibres) Haematoxylin and eosin (original magnification ×10). MRI: Magnetic resonance imaging

Discussion

A human tail is a benign congenital anomaly resulting in a vestigial lumbosacral dorsal cutaneous appendage which can be categorised as either a 'true tail' or 'pseudo tail' by clinical, radiological and histopathological examination.[1-4] Human tails may occur alone or are associated with other anomalies. The most common abnormality associated with a tail is spinal dysraphism, which occurs in 49% of cases and can be diagnosed as meningocele, myelomeningocele or spina bifida. Other associated anomalies include lipoma (27%), tethered spinal cord (20%), coccygeal vertebrae (12%), syndactyly, hemangioma, cleft palate, Crouzon syndrome, clubfoot, omphalocele, congenital tracheal stenosis, Von Recklinghausen disease, digit hypoplasia and tetralogy of Fallot.[2] However, few reports of the human tail at the perineal or perianal region have been described. Moreover, the tails are rarely accompanied with ARM.[5-9]

Human embryos normally have a prenatal tail that measures about one-sixth of the size of the embryo itself. At between 5 and 6 weeks of age, the normal human embryo has 10–12 developing tail vertebrae. By the 7–8th week of gestation, the 6th to 12th tail vertebrae have disappeared through cell death, and likewise, the associated tail tissues also undergo cell death and regress. Harrison suggested that the true tail of humans results from incomplete regression of the most distal end of the normal embryonic tail found in the developing human foetus.[1] However, this hypothesis cannot explain all cases reported as so-called 'human tail' and this has not been verified.

The aetiology of ARM remains unknown. The common theories in ARM lie with the premise of incomplete separation of the urogenital cavity and anorectal cavity during their normal septation of these cavities promotes the formation of abnormal external openings and maintained fistulous connections with the break down of the cloacal membrane at 7th week of gestation.[10] Given that the timing of 'the regression of the embryonic tail' and 'the formation of ARM', though the exact relationship between the human tail at the perineal or perianal region and ARM remains obscure, it is also possible that both processes of these simultaneous events in the same developmental area have interfered with each other.

In fact, in our case as well, the tail was connected to the external anal sphincter muscle, and it was also associated with the lower imperforate anus. Although perineal or perianal region cases also may be included in the general meaning of the human tail, it is considered one of the possibilities that for perineal human tails including the striated muscle, a mechanism such as traction or rotation of the tail was added with the development of the external anal sphincter.

Although a process at the perineal or perianal region could be considered a kind of human tail, we consider that its genesis was considered different from that of sacrococcygeal origin. From the embryological point of view, we suggest that the human tail at the perineal or perianal region is thought to be related to the ARM.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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