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A Rare Case "Human Tail" Associated with Lipomeningocele and Tethered Cord

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Abstract

Introduction: The human tail is perhaps the most exciting sign of a neural tube defect in the skin. From the appendix until creating a 20 cm long tail-like lesions are reported in this literature. They may occur associated with an underlying pathology condition such as a lipoma or teratoma, but mostly, they hide the underlying spinal dissrafism.

Case Report: A 5 years girl came with complaints of mass 2x1x2 cm soft and skin covered with fluctuations in the lumbosacral area and tail 18 cm long extending from this mass. The neurological examination does not indicate a deficit. Power movement shows no decrease in motor skills in both limbs. The Cranial MRI image of the patient is standing. Spinal MRI shows fusion defect posteriorly at S3 and S4 levels and the meningocele sac extending to the deck's right. Sequence S1 revealed that the lipomatous tissue extends through the bone canal at the S3 level. Bone marrow ends at the S3 level. Excision of lipomeningocele is planned for this patient.

Discussion: Many classifications about the human tail have been suggested in history. However, regardless of the type, the primary approach to this lesson is always the same. They investigated the possibility of spinal dysraphism with pathology that occurs concurrently and planned treatment based on the patient. In neurosurgery, our goal is to determine the pathology that co-occurred accurately, fixes defects, and correct aesthetics so that the patient can be more confident and for the follow-up on pediatric patients regarding their development.

Conclusion: The clinicians must do the human tail case in clinical and radiological examination very carefully. There are many classifications based on embryology and aetiology; however, we should plan the treatment specifically for patients with clinical findings and radiological findings despite this classification. In neurosurgery, our goal is to determine the pathology that co-occurred accurately, fixes defects, and correct aesthetics so that the patient can be more confident and for the follow-up on pediatric patients regarding their development.

Keyword: Human tail, spina bifida, lipomeningocele, tethered cord

Introduction

The human tail may be the most prominent skin sign of spine dysraphism. This skin fold is mainly seen in the lumbosacral region. There are many theories and classifications about the embryological background.[1] Management must include a complete neurology history and examination also magnetic resonance imaging or computed tomography scan. After diagnosis, we should perform microsurgery with intraspinal components to avoid damage and neurological deficit. [2]

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Case Report

A 5 years girl came with complaints of mass 2x1x2 cm soft and skin covered with fluctuations in the lumbosacral area and tail 18 cm long extending from this mass (Figure 1). The neurological examination does not indicate a deficit. Power movement shows no decrease in motor skills in both limbs. The Cranial MRI image of the patient is standing. Spinal MRI shows fusion defect posteriorly at S3 and S4 levels and the meningocele sac extending to the deck's right (Figure 2). Sequence S1 revealed that the lipomatous tissue extends through the bone canal at the S3 level. Bone marrow ends at the S3 level (Figure 3). Excision of lipomeningocele is planned for this patient.

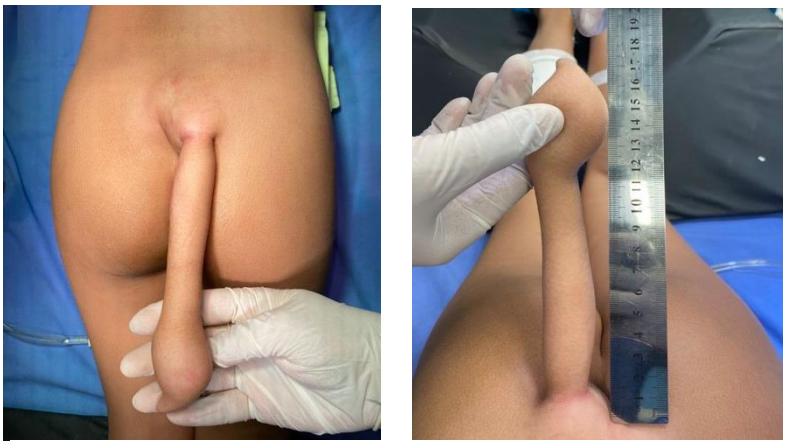

Figure 1. Soft tissue mass 2x1x2 cm covered by skin with fluctuation in paramedian right lumbosacral area and skin appendix extending 18 cm from this mass.

The patient was operated on by neuromonitoring. The arch incision adjusted to the position of the mass, which consists of both sides of the base of the tail, performed at the level S3, solid dura mater achieved and its layers that pass through the defect zone removed. In zone fusion of sac and posterior defect, tissue lipomatosis extending from the tail was observed. Dura mater incision on the tail and pockets opened. Nerve roots are kept in the inner wall of the pouch (Figure 4). Under the guidance of neuro monitorization, pocket wall, lipomatous tissue and tail are removed (Figure 5). Laminectomy was performed until S1 level to explore lipomatous at the sacral level. Fatty phylum terminal seen and cut. Pathological evaluation only reveals skin and subcutaneous tissue, and no neural networks were reported.

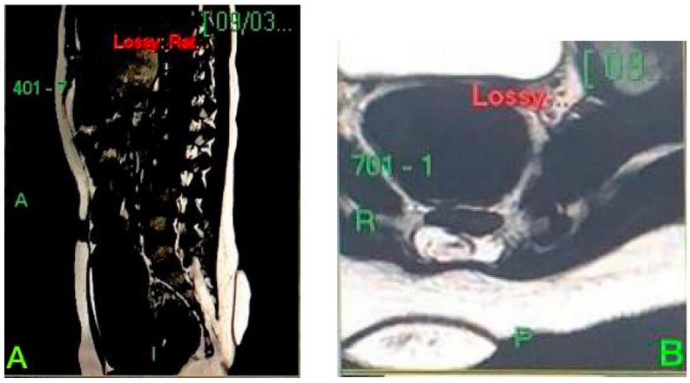

Figure 2. Lesion at S3 and S4 level and meningocele sac to the right from the lesion. A) Sagittal field and B) axial field at S3 vertebrae level

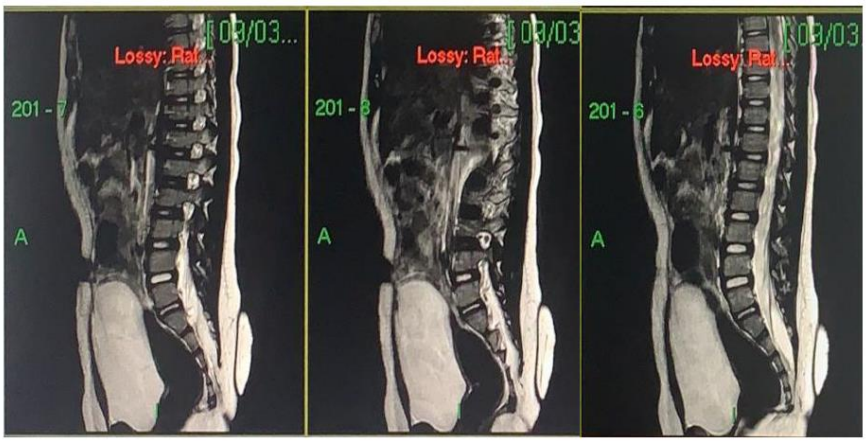

Figure 3. Lipoma extends through the spinal canal at the S1 level. Fatty phylum tip thick at the tail

Discussions

The human tail is a general for the tail-like folds of skin lesions that usually occur at the lumbosacral and bony tail levels. Because of dysraphismspine, when faced with extending of the tail was observed. Dura mater incision on the tail and pockets opened. Nerve roots are kept in the inner wall of the pouch (Figure 4). Under the guidance of neuro monitorization, pocket wall, lipomatous tissue and tail are removed (Figure 5). Laminectomy was performed until S1 level to explore lipomatous at the sacral level. Fatty phylum terminal seen and cut. Pathological evaluation only reveals skin and subcutaneous tissue, and no neural networks were reported lesions like that. We must always consider the possibility of occult spina bifida. Although the human tail is a rare congenital anomaly, it is essential to know because sometimes the bias accompanies central nervous system anomalies. [3]

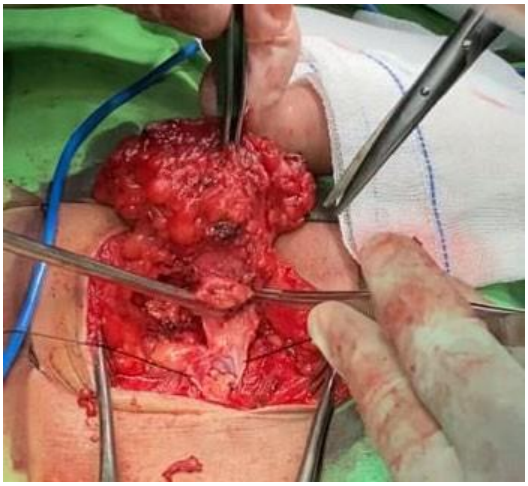

Figure 4. Nerve roots in the inner wall of the sac

Figure 5. Tail cut and measured

In the fourth week of life, embryologically, sclerotomes are developed from somites, which start to wrap around the tube nerve. In the fourth until sixth week, during spine formation, the embryo has a tail formed by 10-12 coccyx. At the end of the sixth week, the spine becomes the primary support for the source on the plan axial. After this week, the coccyx bone in the tail mentioned above started dissolved by phagocytes. Everything disappears at the end of the eighth week, except for a few tail segments proximally. Vertebrae proximal remaining from the tail level regressed, and after birth, they join and form bones tails. If this fusion is to be stopped, the deficit will appear as a "human tail" formation. It's the leading theory described in the 1900s.[1] In 1989, Gaskill and Marlin stated that it could develop these lesions from neuroectoderm. This may form by the ducts of the dermal sinuses, which expand out of the skin, as an alternative theory. [4]

Studies about "dermal sinus" and "myeloschisis limited" by Pang et al. in 2013 also supports this theory.[5] This theory is consistent with high-level spinal dysraphism (50%) and umbilical cord (81%).[1][5] Considering the number and variety of accompanying anomalies (lipomielomeningocele, myelocyte, umbilical cord tethered, lipoma, anal atresia, poultice kidney horses, congenital heart disease, teratoma), it is suggested that there may be some

pathology as causes of these forms.[6][7] In the accompanying case, lipomeningocele, as presented before, there is primary neurulation. Most likely, that stage is responsible for the formation of the tail.

Bartel made the first classification of the human tail in 1884. Bartel classified this appendix into four categories according to form and bone tissue retention. [8] In 1984, Dao and Netsky sorted humans' tails into two categories as original and pseudotails according to their origins embryological. [9] According to this classification, the actual tail is formed by embryological remains and includes muscle, adipose, and tissue but no spine in the structure. Pseudotails are lesions that have an underlying disorder, such as lipoma or teratoma.[10] Despite the classification having embryological value, this classification is not significant from a clinical perspective. In 1998, Lu et al. suggested different criteria for accurate pseudotail-tail classification. He called benign coccygeal and gluteal lesions true tails, and he indicated that necessary excision is sufficient as the treatment. He described pseudotails as tail-like lesions accompanied by a spinal dysraphism, and he explained that this tail-like lesion has an induced ectodermal origin by spinal dysraphism.[10] Classification helpful in clinically that used today, and the primary approach to this case is looking for the spina bifida accompanying or other anomalies, analyzing, and decide how the best treatment after the examination.[10] In 2016, the new classification that divides the human tail into five groups — tail appendage soft tissue, coccyx, protruding bone tail, true tail and other tail appendages - recommended. [11] However, all of these classifications only have more value for embryology alone, not clinical practice.

Radiological evaluation is critical to do in the case of the human tail. These lesions often contain pathology of the spine, and apart from all studied classification, our target is determining this pathology and treating it one by one. MRI can see what tissue is present in the lesion on the case to determine the course of action it will take.

Conclusion

The clinicians must do the human tail case in clinical and radiological examination very carefully. There are many classifications based on embryology and aetiology; however, we should plan the treatment specifically for patients with clinical findings and radiological findings despite this classification. In neurosurgery, our goal is to determine the pathology that co-occurred accurately, fixes defects, and correct aesthetics so that the patient can be more confident and for the follow-up on pediatric patients regarding their development.

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