Multidisciplinary management of human tail and associated spinal dysraphism.

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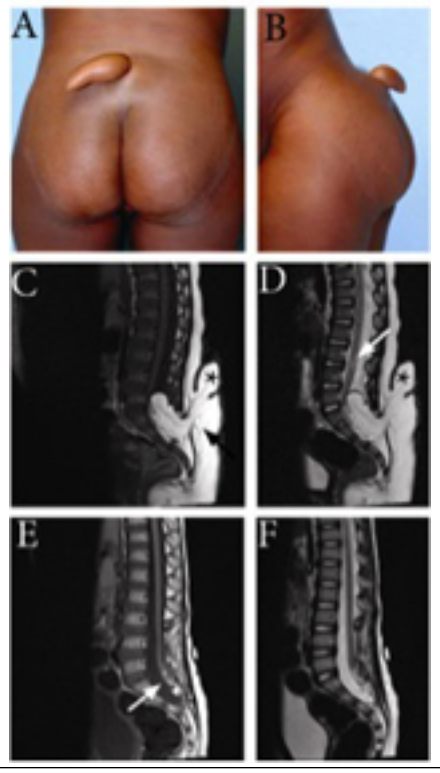
Introduction

The dorsal cutaneous appendage, aka human tail, is a rare congenital lesion defined as "a protruding abnormality from the lumbosacral region". The soft tissue tail may occur in isolation or may be a marker for underlying "occult" spinal dysraphism. Human embryos typically have a tail at the 6th week of gestation, but this typically regresses by the end of the 8th week. Failure of regression is a rare event, and in the newborn, it usually presents with a skin-covered lumbosacral appendage containing adipose and connective tissue, muscle, vessels, and nerves, but lacking vertebra, cartilage, notochord, and spinal cord.

Patients: We present four patients, treated between 2011 and 2014. #1 A 1 yo with a large tail had spinal dysraphism in continuity with a lipomyelomeningocele;progressive left lower extremity weakness and foot drop, which improved within 6 months after microsurgical detethering. #2 A 10 months old with a smaller tail isolated to the skin and subcutaneous tissue associated with a dorsal intradural lipoma and presented with frequent urinary tract infections (UTI). An intradural lipoma was microsurgically detethered. No further UTIs were observed in the 2 years postop follow-up. #3 A 22 year female, presented with a small tail isolated to the skin in the sacral area combined with multiple thoracic and lumbar anomalies.

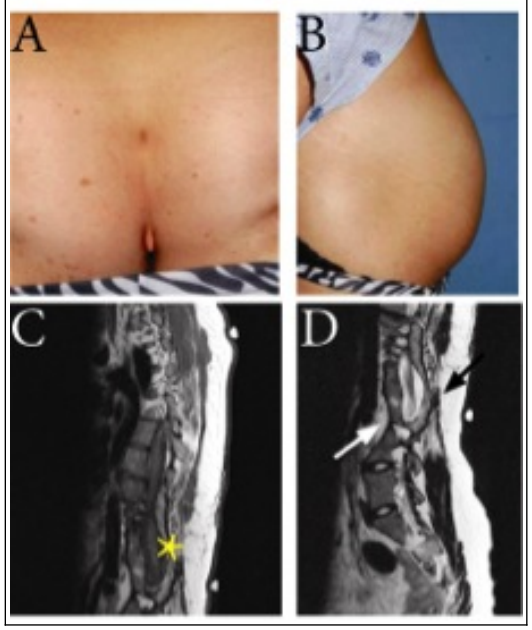
She was born with a thoracic level myelomeningocoele and clubfoot treated in infancy. The 22 year old female had cosmetic repair of the isolated skin appendage. #4 A newborn presented with a large tail and MRI of his spine and brain revealed a lipomyelomeningocele with thoracic syringomyelia. The patient had decreased tone in the lower extremities on follow-up and also dilated ureter on an pelvic ultrasound that warranted microsurgical detethering and repair of the skin appendage at 3 months of age.

Case #1



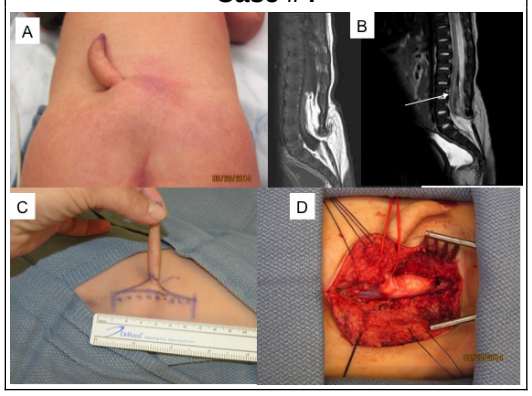
Prominent lumbar cutaneous appendage at 1 year of age. Asterisk: exophytic lobulated fatty mass, white arrow: syrinx (E,F) 2 ys post repair.

Case #3



Sacrococcygeal appendage in an adult. No continuity with the spinal canal (asterisk). Significant scoliosis and vertebral anomalies (arrows).

Case #4



T2 shows the placode with edema (arrow). D:myelomeningocele connected to the tail through a lipomatous and fibrovasular stalk.

Conclusions

If neurologic compromise is progressive as evidenced in one of our patient youngest patient, surgical repair might be warranted sooner within the first year of age. Traditionally, occult spinal disraphism is not repaired before 6 months of age. The fact that deterioration can occur in an instant is shown by one of our patient that developed a foot drop within her first year of life. Both patients with progressive neurological deterioation did show most prominent skin appendages. We emphasize the importance of a collaborative and interdisciplinary surgical approach between pediatric, plastic and neurosurgeons for best functional results.

References

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