

Neuroectodermal Appendage: A Case Report and Review

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Abstract : The human tail has long been an object of curiosity. Sporadic cases have been reported. We present a case of a tail-like structure in an adult associated with an intradural dermoid tumour at L1-L2. The dermoid tumour attached to the dorsal surface of the conus medullaris and cauda equina was excised and the filum terminale extending to the sacral appendage was

dissected. These findings support the hypothesis that neuroectodermal appendages may be a superficial extension of a dermal sinus tract.

Key Words : Dermal sinus tract, Human tail, Neuroectodermal appendage.

INTRODUCTION

The human tail has long been an object of curiosity (2,14). It was an example to early scientists of the recapitulation involving phylogeny and ontogeny or reversion to lower species (7,13). Neuroectodermal appendages are persistent vestigial appendages localized in or near the midline posteriorly and extending to the spinal canal. Sporadic cases have been reported (1,4,5,9). More rational approaches regarding the aetiology and surgical treatment have been made recently (6). We present a case of neuroectodermal appendage associated with intradural dermoid tumour.

CASE REPORT

The patient was a 21yearold man, a recruit in the Turkish Army. He was a product of a normal pregnancy and delivery. His parents had refused surgery for the skin appendage at the age of 5 months. A deformity in the right foot developed after the age of 15 years. He started to have weakness and numbness in the right leg and foot after joining the army and was admitted to the department of neurosurgery. Physical examination was unremarkable except for right pes cavus deformity

and a sacral appendage 3 cm. in length tapering at the distal end (Fig.1). On neurological examination, hypoaesthesia in right L4-L5 and left L5, bilateral sacral dermatomes, and paresis in the right foot were found, and bilateral ankle jerks were absent. He was continent and did not complain of sexual dysfunction. X-ray of the lumbar-sacral vertebrae showed an increased interpedicular distance at L1 and L2, and S1 spina bifida. Contrast material did not pass above L3 in myelography. An intradural extramedullary mass, located at L1 and L2 was detected on MRI scans (Fig. 2). At surgery, L1 and L2 total laminectomies were done and an intradural dermoid tumour attached to the dorsal surface of the conus medullaris and cauda equina was excised totally under an operating microscope. The conus extended to the level of L2-L3. The neuroectodermal appendage was seen to pierce the dural sac. S1 total laminectomy was done and a thickened fibrous filum terminale, connected to the tail, was sectioned and the neuroectodermal appendage removed. The postoperative course was uneventful, however preoperative neurological deficits persisted. Follow-up examination revealed no change in his neurological status at the tenth postoperative month. Microscopy disclosed a dermoid tumour consisting of a stratified squamous epithelium and hair follicles. The skin

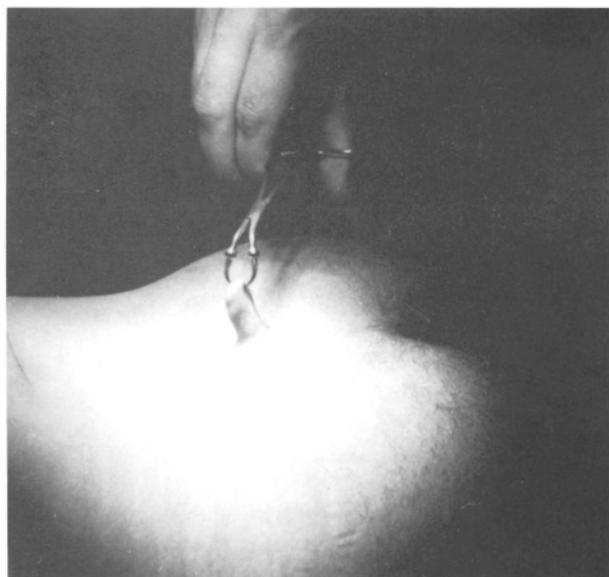


Fig. 1 : Photo taken on the operating table shows sacral neuroectodermal appendage.

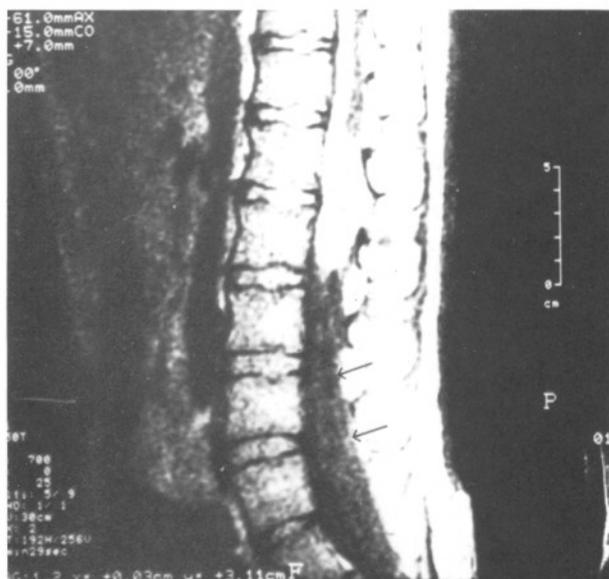


Fig. 2 : MRI scans showing the dermoid tumour at L1 and L2, and the filum terminale (arrows)

appendage when examined microscopically, was composed of adipose tissue and fibrous bands.

DISCUSSION

During the sixth week of gestation (14 to 16mm.), the human embryo has a tail, virtually indistinguishable from the embryonic tails of tailed species (11). In the seventh and eighth weeks of gestation, the human tail regresses. As the number of

caudal vertebrae reduces, the projecting portion of the tail disappears as a result of the growth of other caudal structures (8). Tailless animals and humans do not have caudal spinal roots. The filum terminale containing glial fibres and ependyma is the remains of these roots.

Dao and Netsky (4) reviewed a total of 32 cases of skin appendages published from 1859 to 1982, and classified human tails as true or pseudotails. True tails contain adipose and connective tissue, central bundles of striated muscles, blood vessels and nerves, and are covered by skin. Bone, cartilage, notochord, spinal cord are lacking in true tails. Pseudotails are protrusions such as lumbosacral myelomeningocele, lipoma or anomalous prolongations of coccygeal vertebrae. In the Dao and Netsky review, only two patients were adults. Other reported cases are in the paediatric age group (1,4,6,9,10,12,14,17). Our case was 21 years old and developed the findings of cauda equina and conus medullaris involvement. Tails are localized mostly in the sacrococcygeal (1,3,4,5,9,10,12,14,17) and lumbar regions (3,6,16). However, Gaskill and Marlin (6) reported a case of a neuroectodermal appendage in the thoracocervical region in which a tract was extending intradurally. It was reported that movement or contraction of the tail was noted in some cases (4,10,15,17). But we did not see any movement of the taillike protrusion in our case. Microscopically neuroectodermal appendages may contain microscopically some or all of the following structures; adipose and fibrous tissue, small blood vessels, nerve fibres. They are covered by skin with appendages such as sweat glands and hair follicles. No bone or cartilage is found in true tails (4,5,9,10,12,14,17). Lipoma, spina bifida, club foot, cleft palate, syndactyly, small toe were among the associated anomalies in cases of human tail reviewed by Dao and Netsky (4), and Dubrow et al. (5). In all cases reported by Gaskill and Marlin (6) the skin appendages extended into the spinal canal and attached to the spinal cord and were associated with intradural lipoma in two cases. Talwaker (15) and Belzberg et al. (3) also mentioned the intradural extension of the appendage in their cases.

Gaskill and Marlin (6) discussing the aetiology of human tails in their paper, proposed that neuroectodermal appendages represented the superficial extension of a dermal sinus tract, and the simultaneous

epithelization of this tract in both outward and inward directions to form a neuroectodermal appendage. The dermoid tumour and intradural extension of the tail in our patient support this hypothesis. Most cases reported in the literature lacked extensive neurological examination and work-up. The patients were in the very early years of life and we believe that the skin appendages extended intradurally in many cases. If those patients had been seen at older ages, signs of a tethered cord syndrome could have been noticed.

In conclusion, tail-like structures or neuroectodermal appendages need meticulous neurological examination and extensive workup. Intradural extension of these lesions and associated spinal pathologies should always be kept in mind. Surgical treatment is necessary and it is wise to make sure that the cord tethering is released.

Acknowledgement : The authors thank Dr. David G. McLone for reviewing the manuscript.

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