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Caudal cell mass developmental aberrations: an imaging approach

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ABSTRACT

The objective of this review is to describe antenatal and postnatal imaging criteria, which allow diagnosis and aid workup, prognostication and treatment of developmental anomalies of the caudal cell mass. The lower spinal cord (conus medullaris), filum terminale and inferior lumbar and sacral nerve roots develop from the caudal cell mass, a remnant of the embryologic primitive streak composed of undifferentiated pluripotential cells. Anomalous caudal cell mass development can manifest as tight filum terminale, caudal dysgenesis, terminal myelocystocele, anterior sacral meningocele or sacrococcygeal teratoma. Lower spinal cord development occurs simultaneously and in topological proximity to the developing lower gastrointestinal and genitourinary tracts, leading to coexistent malformations.

We review the embryology of the caudal cell mass, describe the role of antenatal and postnatal imaging for diagnosing, staging, prognosticating and guiding intranatal or postnatal intervention for developmental anomalies of this region and briefly discuss clinical manifestations and treatment goals and strategies. An overview of antenatal imaging diagnosis of associated multisystem abnormalities will be provided where applicable.

1. Introduction

Abnormalities from maldevelopment of the caudal cell mass may be local, systemic or with syndromic associations. Some of these abnormalities may need surgical intervention; others require medical, orthopedic or urologic intervention. Antenatal and postnatal imaging aid diagnosis and prognosis, and guide treatment for some of these entities. Radiologists must familiarize themselves with the spectrum and clinical significance of these entities and search for coexistent multisystem abnormalities since antenatal and postnatal imaging may aid diagnosis and prognosis and guide treatment. This article reviews the embryology, clinical manifestations, imaging diagnosis and treatment of developmental aberrations of the caudal cell mass.

2. Embryology

The sacrum, coccyx and caudal portion of the conus medullaris, filum terminale and lower lumbar and sacral nerve roots develop from a large aggregate of undifferentiated cells representing remnants of the primitive streak. The embryological processes underlying development of the caudal cell mass include canalization (secondary neurulation) and retrogressive differentiation. These processes are distinct from the preceding events of gastrulation and primary neurulation, which result in formation of the mesoblast/primitive streak and neural tube respectively [1].

In the 30-day embryo, the caudal cell mass forms cell clumps and cysts; the cysts coalesce into an ependyma-lined tubular structure, which unites with the more proximal neural tube. The adjacent developing lower gastrointestinal and/or genitourinary tracts may be simultaneously affected by caudal cell mass malformations [2]. Sacral ossification proceeds craniocaudally with ossification centers of S1 to S3 appearing between 10 and 12 weeks of gestation and with the ossification center for S5 appearing between 5 and 8 months of gestation [3,4]. The coccyx does not ossify during gestation.

Over the following paragraphs, we discuss specific abnormalities resulting from aberrant development of the caudal cell mass. For each abnormality, we review the embryology, clinical manifestations, associated abnormalities, antenatal and postnatal imaging diagnosis, and treatment (also see included Table 1).

3. Normal conus medullaris and filum terminale

In early embryogenesis, the location of each neural segment exactly matches that of the corresponding vertebral segment so that the spinal

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Table 1
Developmental aberrations of the caudal cell mass.

Entity	Embryology	Clinical manifestations	Imaging findings	Treatment strategies/goals
Tight filum terminale	Anomalous production of fat cells by pluripotential caudal cell mass or failure to connect areas of cavitation to central canal Failure of involution of the terminal cord	Frequently asymptomatic Progressive motor and sensory changes in back and legs Foot deformities Scoliosis Urinary dysfunction	Fibrolipomas or cysts Tethered cord	Management differs based on age Prophylactic spinal cord release to relieve pain and sensorimotor deficits
Caudal dysgenesis	Developmental insult to the caudal mesoderm, which results in impaired migration of neurons, paraxial mesoderm and lateral mesoderm	Currarino triad Caudal regression syndrome Sirenomelia OEIS complex VACTERL association Motor abnormalities corresponding to the level of bone anomaly	On fetal ultrasound, inappropriate crown-rump length; if severe, “Buddha's attitude” Possible associated absence of lumbar and/or sacral spine Anorectal malformations Urogenital anomalies – renal dysplasia and/or vesicoureteral reflux	Manage fecal and urinary continence Prevention of urinary infections Surgical management for associated orthopedic abnormalities and physical therapy
Terminal myelocystocele	Elongated spinal cord traverses an extraspinal cerebrospinal fluid filled cyst, adherent to the subcutaneous fat, and inserts within its lateral or posterior wall	Lack of bladder or bowel control Impaired lower extremity function Scoliosis, lordosis	Caudal meningocele and hydromyelia Widened posterior elements of the vertebral bodies Dilated terminal canal	Early repair with resection of the nonfunctioning caudal cyst wall Reconstruction of the proximal neural placode and duraplasty Manage fecal and urinary continence
Anterior sacral meningocele	Focal hypogenesis of sacral or coccygeal segments with intrapelvic herniation of cerebrospinal-fluid dura matter through an anterior osseous defect or via sacral foramen	Small lesions are asymptomatic May occur in the context of Currarino triad	“Pear-shaped” pelvic cyst on fetal ultrasound Narrow stalk connects the cyst with the spinal canal Possible “Scimitar sacrum” Accompanying osseous defects	Obliterate communication between the meningocele and the spinal subarachnoid space Excision of meningocele and untethering of the spinal cord if necessary Manage fecal and urinary continence
Sacrococcygeal teratoma	Tumors derived from Hensen's node; arise due to abnormal persistence of pluripotential primitive streak	Impaired bowel and/or urinary control Impaired lower extremity function Prenatal hydrops	Pelvic heterogeneous mass – cystic and solid that may contain fat and calcifications	Prenatal therapy includes laser, radiofrequency ablation, bladder drainage for obstructive uropathy, and aspiration of cystic components Tumor excision including coccyx Manage fecal and urinary continence

cord extends to the caudal end of the spinal canal [5]. During the second trimester, the vertebral column elongates more rapidly than the spinal cord [4] with discrepancy of levels and relative ascent of the spinal cord. By full-term, at nearly 40 weeks, the conus medullaris (the caudal termination of the spinal cord) lies between L1 and L3 [5]; its termination below the inferior endplate of L2 in a term infant should be considered abnormal and search should be made for a tethering mass (usually lipoma), bony spur or thick filum. Although the conus may be slightly lower in preterm infants; recent evidence suggests that the conus should be no lower than the bottom of L3 by 24 weeks of gestation [6].

The normal filum terminale is a thin fibrous filament about 20 cm in length, which contains glial and ependymal cells and has intra-and extra dural components. The upper, intradural portion (filum terminale internum) is approximately 15 cm in length, is surrounded by subarachnoid space (which is the site of access to the CSF via lumbar puncture), and descends from the tip of the conus medullaris through the bottom of the subarachnoid space and dura mater at about S2. The extradural component of the filum terminale (filum terminale externum, also called the coccygeal ligament) [7,8] is about 5 cm long; it extends through the S2 inferior spinal dura to insert on and blend with the first coccygeal segment dorsal connective tissue. At L5-S1, the normal filum terminale should measure < 1 mm in diameter on axial MR. The conus is best visualized on axial MR, since the cauda equina nerve roots may obscure the precise conus level on sagittal images.

4.1. Anomalies of the filum terminale/tight filum terminale (simple spinal dysraphism)

Anomalous development of the filum terminale can manifest as ***fibrolipomas/cysts*** of the filum terminale and as ***tight filum terminale***. The embryologic events leading to ***fibrolipomas*** and ***cysts*** of the filum include anomalous production of fat cells by the pluripotential caudal cell mass or failure to connect areas of cavitation to the central canal. These may be intradural, extradural or involve both portions. Filar fibrolipomas (Fig. 1) are hyperintense on T1-weighted images and hypointense on T2 weighted images, occurring within an enlarged filum. Although frequently asymptomatic, fibrolipomas may become symptomatic at any age requiring careful neurologic examination and urodynamic testing with surgery performed if necessary [9,10].

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