MRI of notochordal anomalies

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Authors: R. Van Eetvelde¹, M. Lemmerling²; ¹Lokeren, Oost-Vlaanderen/BE, ²Beervelde/BE
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Learning objectives

- to review embryological development and function of the notochord
- to review notochordal (developmental) anomalies and their MRI features
- to review the most important differential diagnoses

Background

The notochord is a cellular rod extending from Rathke's pouch to the coccyx that defines the primitive axis of the body. Phylogenetically, it is the common factor defining all species of the highest phylum of the animal kingdom, the Chordata. During the second week of normal embryonal development, the embryo consists of dorsally lying ectoderm (skin, spinal cord) and ventrally lying endoderm (foregut) which is in contact with the yolk sac. During the third week of gestation, a midline primitive streak appears at the caudal end of ectoderm. Proliferating ectodermal cells from the primitive streak migrate laterally between ectoderm and endoderm to form the mesodermal layer. This conversion of the embryonic disk from a bilaminar disk to a trilaminar disk composed of ectoderm, mesoderm and endoderm is called gastrulation (Fig.1). The cephalic end of the primitive streak thickens and forms Hensen's node. Proliferating cells from Hensen's node migrate cranially in the midline to form the notochord. The notochord separates endoderm from ectoderm in the midline and is believed to act as an embryonic organizer. It induces chondrification and segmentation of the mesenchymal elements of the vertebral bodies and also influences the development of the neural plate (1,2).

Findings and procedure details

Two subtypes of notochordal anomalies exist.

1. Developmental anomalies causing 'split notochord syndrome'

Normally the notochord separates the ventral endoderm and dorsal ectoderm during embryogenesis. Split notochord syndrome is a spectrum of congenital spinal malformations that develop due to an adhesion between endoderm and ectoderm causing the 'splitting' of notochord. All forms of split notochord syndrome are frequently associated with vertebral anomalies such as anterior and posterior spina bifida, butterfly vertebrae and hemivertebrae.
a. **Most frequent manifestations are** NEURENTERIC CYSTS

*Basics*

Neurenteric cysts are rare (0.3-0.5% of all spinal tumors) benign congenital abnormalities that develop as a result of an abnormal connection between the primitive ectoderm and endoderm. If located in the spine they are associated with vertebral anomalies in 50 % of cases (3,4).

*Clinical presentation*

Patients with spinal neurenteric cysts typically present in the second and third decades of life with size-dependent myelopathic and/or radicular signs (4,5).

*Location*

Spinal and rarely intracranial, in the mediastinum, abdomen, pelvis or subcutaneous tissue (3-7).

*Spinal Neurenteric cysts*

- 50 % cervical spine, 25 % thoracic spine, 25 % thoracolumbar junction
- 90 % intradural/extramedullary and 10 % intradural/ intramedullary
- ventral or dorsal to the spinal cord

*Imaging features*

- multi- or unilobulated

MRI (Fig.2)

T1: variable signal intensity depending of protein content

T2: high signal intensity

T1 Gd+: no enhancement

*Differential diagnoses*

- Dermoid cyst (Fig.3)
• heterogeneous signal intensity: mixture of fat (<> neurenteric cyst), soft tissue and fluid
• most often in the lumbosacral region (60%) and cauda equina (20%) and only rarely in the cervical or thoracic spine (<> neurenteric cyst)

- Arachnoid cyst or extradural meningeal cysts (Fig.4)
  • follow the CSF signal intensity on all sequences

_Treatment_

Complete surgical resection is the treatment of choice if clinically needed.

**b. Less frequent (and more severe) manifestations include**

**b1. ENTERIC FISTULA/SINUS/DIVERTICULA**

**b2. CONGENITAL SPLIT CORD MALFORMATIONS**

_Basics_

This disorder may result from splitting of the notochord around an adhesion between the endoderm and the ectoderm. This split notochord might influence the formation of two neural tubes and subsequently two hemicords. The notochord also influences vertebral formation and thus it is common to have associated segmentation anomalies at the site of diastematomyelia. Females are affected more commonly (80%) than males (1,8).

_Clinical presentation_

Most patients with diastematomyelia present in childhood. The neurological disturbances produced by deformity of the cord vary greatly from individual to individual. Most frequently there is difficulty in walking, caused by a weakness or paralysis of the muscles in the legs. Sphincter disturbances of the bladder and rectum are present in about 50% of cases (8).

_Imaging findings_ (Fig.5)

- Two subtypes:

  1. Type 1 or diastematomyelia (60 %): two dural tubes
  2. Type 2 or diplomyelia (40%): one dural tube

**2. Mass formation from notochordal remnants:**
In humans there is normally complete degeneration of the notochord starting during fetal development and ended by the second decade of life. However, in some cases, notochordal remnants persist and cause lesions like Tornwaldt cysts or chordomas.

**TORNWALDT CYSTS**

*Basics*

Tornwaldt bursa are benign developmental lesions resulting from persistent notochord remnants. If the opening of the bursa is occluded, cysts develop. They are found in approximately 0.2%- 5 % of routine MRI studies and show no gender predilection (9).

*Clinical presentation*

Usually asymptomatic. When infected they can cause halitosis, nasal discharge and an unpleasant taste in the mouth (10).

*Location*

- Typical midline location in the posterior superior wall of the nasopharynx
- Immediately beneath the mucosa and anterior to the longus colli muscles

*Imaging features*

- Rounded and unilocular with a mean size of 6 mm
- No edema in the surrounding soft tissues or involvement of adjacent bone

MRI (Fig.6)

T1: variable signal intensity depending of protein content and/or associated hemorrhage

T2: typically high signal intensity

T1Gd +: no enhancement (possibly thin peripheral cyst wall enhancement)

*Differential diagnoses*

Usually none because of its typical location and imaging appearance.

Differential diagnosis may include:
normal or prominent adenoidal tissue with cystic degeneration
- mucous retention cyst (mucocele)

*Treatment*

If symptomatic by deroofing (marsupialization) or complete excision (9,10).

**CHORDOMA**

*Basics*

Chordomas are uncommon malignant tumors accounting for 4 % of all primary bone tumours and 1 % of intracranial tumours. They originate from embryonic remnants of the primitive notochord. They are most common in the 4th to 7th decade of life with a 2:1 male predilection. Generally they are slow growing but they have an extremely high recurrence rate. Distant metastasis is rare (11,14).

*Clinical presentation*

They present due to mass effect on adjacent structures (brainstem, cranial nerves, nasopharynx, spinal cord), or as a mass (sacroccygeal).

*Location*

Located along the course of the embryonic notochord:

- 30-50 % sacro-coccygeal
- 30-35% spheno-occipital
- 15-30% vertebral body, most commonly in the cervical spine (C2)

*Imaging features*

Midline, lytic, destructive lesion with soft tissue involvement with general size of 2-5 cm (12-14).

MRI (Fig.7, Fig.8)

T1: intermediate to low signal intensity with small foci of hyperintensity (hemorrhage, which can be confirmed with gradient echo imaging as dark areas, or mucus pool)
T2: high signal intensity (high fluid content) with intratumoral areas of heterogeneous hypointensity (calcification, hemorrhage or highly proteinaceous mucus pool)

T1Gd+: heterogeneous enhancement, sometimes with a honeycomb appearance (intratumoral areas of low signal intensity). Occasionally slight or even absent enhancement (necrosis or large amount of mucinous material)

**Differential diagnoses**

**Chondrosarcoma**

- most often confused with intracranial chordomas
- if cranial, majority arises along the petro-occipital fissure (<> chordomas typically have a midline skull base location)
- similar signal intensity on T1 and T2 as chordoma
- linear, globular or arclike calcifications possible (<> uncommon for chordoma)

**Meningeoma**

- dural attachment
- causes bone sceloris (<> chordoma are destructive bone lesions)
- homogenous enhancement

**Treatment**

Extensive surgical removal and postoperative radiation therapy are most effective (13).

**Images for this section:**
Fig. 1
Fig. 2: Midsagittal T1-weighted image (a) shows an intradural/extradural neurenteric cyst at T11 with isointense signal intensity. The corresponding T2-weighted image (b) illustrates that such cysts are homogeneously hyperintense.
**Fig. 3:** Midsagittal T2-weighted image (a) shows a dermoid cyst at L3 as a heterogeneous intraspinal mass. Associated tethered cord is present. The signal intensity of the lesion on the midsagittal T1-weighted image (b) is hypointense with a hyperintense signal in the peripheral portion indicating the presence of fat. No contrast enhancement was noted after administration of intravenous gadolinium (not shown).
Fig. 4: Midsagittal T1-weighted (a) and T2-weighted (b) images show a meningeal cyst as a well-defined extradural mass at L2 with signal intensity virtually identical to that of cerebrospinal fluid.
Fig. 5: These sagittal T2-weighted images show a split spinal cord at level T1 (a) with associated congenital fusion of multiple vertebral bodies (b). A single dural sac containing two hemicords is present and is best appreciated on the axial T2-weighted image (c). The axial image of another patient shows the second type of split cord malformation. The two hemicords have their own dural sac and are separated by a bony spur (d).
**Fig. 8:** This axial T2-weighted image of a 64-year-old man shows a heterogeneous hyperintense mass in the right side of the clivus (a). After intravenous injection of gadolinium the coronal T1-weighted image shows heterogeneous enhancement with a honeycomb appearance (b).

**Fig. 6:** Axial images show a cyst at the midline of the nasopharynx with a typically high signal intensity on T2-weighted images (a), isointense to cerebrospinal fluid on T1-weighted images (b) and with only a thin peripheral cyst wall enhancement on the axial post-gadolinium images (c).
**Fig. 7:** This axial fat-suppressed flair image of a patient with headache and left-sided trigeminal nerve neuropathy shows a large soft tissue mass centered on the clivus with obliteration of the prepontine cistern (a). The corresponding axial T2-weighted image shows the homogeneous high signal intensity in the tumor (b). The midsagittal T1-weighted image nicely demonstrates pontine compression (c). The axial post-gadolinium T1-weighted image illustrates that no enhancement is noted in this particular patient.
Conclusion

MRI is an excellent technique to investigate/differentiate notochordal remnants and anomalies.

Personal information

References


