Notochord Derived "Pitfalls" of Central Skull Base

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Learning objectives

The purpose of this study is to describe CT and MRI findings of notochord remnants and benign notochord derived lesions of skull base and to differentiate these lesions from chordoma.
The notochord is a cellular rod that forms a central axis, around which the segments of the vertebral column develops. It lies on the ventral aspect of the neural tube and reaches from the sacrococcygeal region to the anterior end of the mid-brain, where it ends in the region of the future dorsum sella of the sphenoid bone. On either side, cartilaginous centers which form the body of vertebra, make their appearance on the fourth week; then extend around the notochord. The notochord, surrounded by the vertebral bodies atrophy and ultimately disappear, except the portion which lies in the center of the intervertebral fibrocartilages that persist throughout life as the central nucleus pulposus.

The posterior part of the base of the skull also develops around the notochord. The notochord runs within the cephalic mesoderm between the ectoderm of the developing neural tube and the endoderm of the pharynx. Later in development, in the presence of advancing chondrification and ossification of the skull base, the notochord undergoes degenerative changes and finally disappears.

Two pairs of cartilages develop around notochord, a pair of parachordal cartilages, one on either side of the notochord and a pair of prechordal cartilages in front of the notochord. From the parachordal cartilages the basiocciput and the occipital condyles develop. The prechordal cartilages form the nasal septum, ethmoidal labyrinth and the lateral and alar cartilages of the nose, the small and the great wing as well as the lateral pterygoid plate of the sphenoid. The cartilages meet and fuse below the pituitary gland, forming the floor of sella.

The notochord emerges from the vertebral axis through the odontoid process; it bends slightly and then enters and crosses the posterior region of the basiocciput (Figure 1). At the level of the basiocciput, the cartilages lie on the dorsal aspect of the notochord, hence in this region the notochord is placed between the cartilages and the wall of the pharynx and has contacts with pharyngeal epithelium. It then continues cephalad under the basiocciput and turns dorsally, penetrating and terminating within the basisphenoid near the region of the pituitary gland.

Several developmental and neoplastic processes may arise from this embryonic pathway. The developmental anomalies, benign and malignant lesions will be discussed separately.
Fig. 1: The pathway of notochord in the skull base. Notochord reaches the surface of the bone at three sites, posterior clivus, pharyngeal surface and dorsum sella; this may explain the ectopic locations of these lesions.

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Findings and procedure details

1. Developmental Anomalies

Canalis Basalis Medianus (CBM)

CBM is an uncommon variant or mild anomaly of basiocciput which occurs in approximately 2% of skulls. It is a well-defined channel originating from the basiocciput in the midline very close to anterior rim of foramen magnum. Two theories are proposed for the formation of CBM, one is vascular and proposes the persistence of emissary veins, the other, which is more probable, proposes it to be the vestige of the notochord.

There are six types of canalis basilaris medianus that have been described (figure 2), including the complete forms (superior, inferior, and bifurcating) and incomplete forms (long channel in basiocciput, superior basiocciput recess, and inferior basiocciput recess [foveola/fossa pharyngica]). These appear as well-defined luencies in various configurations on CT. Fossa navicularis magna, which is a notch-like defect in the anterior aspect of the basiocciput, may actually be a giant form of inferior basiocciput type of incomplete canalis basilaris medianus.

CBM is usually asymptomatic, although there are reports in the literature suggesting fossa navicularis magna can be a potential source of intracranial transmission of infection from the lymphatic tissue of pharynx through skull base.

On CT images, both the complete and incomplete forms have intact cortical rims (figure 3 and 4). The cortical bone is seen as hypointense rim on T2 weighted images (figure 3) and after intrathecal contrast injection, CSF leakage through the canal can be seen on MRI.

CBM is different from the craniopharyngeal canal (persistent hypophyseal canal) which is a vertical midline defect in the skull base that connects the pituitary fossa with the nasopharynx and thought to be a remnant of the Rathke's pouch (figure 5). Large craniopharyngeal canals or transsphenoidal canals may actually represent meningoencephaloceles and can be associated with other craniofacial anomalies.

Tornwaldt's cysts

During development, when the notochord is in contact with the endoderm of the primitive pharynx, a focal adhesion may occur, causing the notochord to carry a segment of the pharyngeal mucosa with it. This creates a potential space known as the "pharyngeal
bursa”, which is lined by pharyngeal mucosa. This bursa develops into a cyst only in when the orifice is obstructed as result of infection or after adenoidectomy. So embryologically, a Tornwaldt's cyst is a persistent communication between the roof of the nasopharynx and the notochord. These cysts may be found in ~ 3% of the adult population.

Tornwaldt's cyst occurs in the midline of the nasopharynx (may be slightly off-centered), above the upper border of the superior constrictor muscle. The cyst has an epithelial lining. It is seen as a soft tissue mass with sharply defined margins high on the posterior pharyngeal wall. Absence of surrounding soft tissue reaction and a lack of bony involvement are other features of the Tornwaldt's cysts.

MRI remains the examination of choice. On MRI scans, a Tornwaldt's cyst shows high signal intensity on both T1- and T2- weighted images and on FLAIR sequence, because of the presence of protein and/or associated hemorrhage within the cyst (Figure 6). Post-contrast studies usually demonstrate peripheral enhancement of the nasopharyngeal mucosa.

2. **Neoplastic Lesions**

A) **Benign Lesions**

**Ecchordosis physaliphora (EP)**

Ecchordosis physaliphora (EP) is a rare, congenital, benign hamartomatous lesion of gelatinous tissue derived from notochord. It is typically located at the midline of the craniospinal axis. This ectopic notochordal remnant is most commonly found in the intradural space of the prepontine cistern with an attachment to the dorsal surface of the clivus. EP is usually asymptomatic and when it is detected it has to be differentiated from other retroclival lesions including chordoma.

EP shows high signal on T2-weighted images, low signal on T1-weighted images, and shows no contrast enhancement. Lesions were defined as classical EP type A which has hyperintense excrescence (cyst-like component) on the dorsal surface of the clivus, classical EP type B which has hyperintense excrescence intradurally and a hyperintense lesion within the clivus (figure 7, 8, 9). Both types can have a tiny T2 hypointense protrusion posteriorly.

EP is attached to the posterior surface of the clivus with a tiny osseous stalk best seen on computed tomography (CT). When seen, the osseous stalk is defined as a morphological hallmark of ecchordosis and does not exist in other retroclival lesions. T2-hypointense protrusion seen on some EPs is thought as the corresponding MR appearance of this osseous stalk.
Chihara et al. also defined incomplete EP types, as EP bud which is a T2-hypointense protrusion of the clivus, and EP variant which is a hyperintense lesion within the clivus alone (figure 7). But it is difficult to distinguish EP variant from benign notochordal cell tumor based on MR findings because both lesions show hyperintensity on T2-weighted images and no contrast enhancement. CT may be helpful because benign notochordal cell tumors usually manifest mild osteosclerosis on CT. No bony destruction is seen on both EP variants and BNCTs.

The major clinically significant point in cases with EP is to distinguish the lesion from chordoma. Both are notochord-related lesions with similar MR signal characteristics, hypointense on T1-weighted images and hyperintense on T2-weighted images; however, chordoma is a malignant tumor with aggressive behavior in contrast to the benign nature of EP. Therefore, chordomas are usually symptomatic lesions showing contrast enhancement and extensive bone destruction, whereas EPs are nonenhancing incidental lesions. Although chordomas mostly occur in extradural location, rarely, they may be seen as an intradural retroclival lesion and confused with EP. Intradural chordomas usually do not present bony involvement. In such cases, the presence of contrast enhancement, suggesting chordoma, presence of a stalk connecting the retroclival lesion with the clivus, suggesting EP, may be the only hints in distinguishing intradural chordomas from EP.

**Benign Notochordal Cell Tumor (BNCT)**

The giant notochordal rest and notochordal hamartoma are terms that have been used to describe a benign notochordal cell tumor (BNCT). They are usually small and found most frequently in both ends of the axial skeleton followed by the mobile spine. Their histological features are different from those of classic chordoma or notochordal vestiges in fetal intervertebral disks.

Imaging is critical in separating the benign lesion from the chordoma. Chordomas are osteolytic tumors. In contrast, in BNCTs, bony trabecula are preserved but may be sclerotic because of appositional or reactive new bone formation. CT scans revealed osteosclerosis in the center of the affected areas, with preserved cortical margins. The sclerotic area displays nodular or diffuse high attenuation with a smooth margin, which displays hyperintensity compared with normal bone marrow on T2-weighted images. The high attenuated areas on CT, histopathologically corresponds to thickened bone trabeculae encompassing tumor, reflecting a benign process on imaging. The presence of extraosseous extension essentially excludes BNCT (figure 10).

The lesions show low signal intensity on T1-weighted MR images and intermediate to high signal intensity on T2-weighted images, hyperintense on short T1 inversion recovery sequences. Gadolinium-DTPA enhanced T1 weighted MR images do not show enhancement. No abnormal uptake was detected on bone scintigrams.
Chordomas are distinguished from BNCTs by trabecular destruction, with a lytic and expansile appearance and extraosseous involvement on imaging. The most significant discriminators between BNCTs and chordomas are extraosseous components, bone destruction, and contrast enhancement on T1-weighted MRI that are present in chordomas.

B) Malignant Lesions

Chordoma

Chordomas are uncommon malignant tumors that account for 1% of intracranial tumors and 4% of all primary bone tumors. They originate from embryonic remnants of the notochord (extending from the dorsum sella to the coccyx). Since chordomas arise in bone, they are usually extradural and result in local bone destruction. They are locally aggressive, but uncommonly metastasize.

On CT, chordoma is seen as a centrally located, well-circumscribed, expansile soft-tissue mass that arises from the clivus with associated extensive lytic bone destruction. Intratumoral calcifications appear irregularly and are usually thought to represent sequestra from bone destruction rather than dystrophic calcifications in the tumor itself.

On MRI, T1-weighted images show intermediate to low signal intensity whereas it has high signal intensity on T2-weighted images. The majority of intracranial chordomas demonstrate moderate to marked enhancement following contrast injection (figure 11).

Chordoma usually can be easily differentiated from other notochord derived lesions by its aggressive behavior. Although differentiating intradural chordoma, which is a subtype of chordomas and doesn't have a bony component, from other benign lesions especially from EP, can be hard. In this case, contrast enhancement of chordomas and T2 hypointense stalk of EP are the only hints that help us. The specific features and hints that help us to differentiate notochord related neoplastic processes are summarized in table 1.
Fig. 1: The pathway of notochord in the skull base. Notochord reaches the surface of the bone at three sites, posterior clivus, pharyngeal surface and dorsum sella; this may explain the ectopic locations of these lesions.

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Fig. 2: Upper line presents the complete forms of CBM, superior (A), inferior (B), bifurcating (C). Lower line presents the incomplete forms of CBM, long channel in basiocciput (D), superior basiocciput recess (E), inferior basiocciput recess (foveola/fossa pharyngica)(F).

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**Fig. 3:** Complete form of CBM, inferior type. Transverse (A), sagittal reformatted (B) CT images. Note that the cortical bone is present at the borders of the canal. Sagittal T2WI (C) and sagittal fat saturated T1 WI following intrathecal gadolinium (D) demonstrate CSF leakage trough the CBM.

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**Fig. 4:** Incomplete form of CBM, inferior basiocciput recess, axial and reformatted sagittal CT images.

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Fig. 5: This figure illustrates the pathway of notochord, locations of CBM (inferior complete type) and craniopharyngeal canal.

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Fig. 6: Tornwaldt's cyst. Schematic drawing of Tornwaldt's cyst (A), as it develops by the communication of the nasopharyngeal mucosa with the notochord. On sagittal T1 weighted image (B), there is a T1 hyperintense lesion at the roof of nasopharynx. On axial T2 weighted image (C), the lesion is hyperintense. On postcontrast images (D), only peripheral enhancement is seen.

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Fig. 7: Types of EP. Classical EP, type A (A), hyperintense excrescence (cyst-like component) on the dorsal surface of the clivus, classical EP type B (B) which has hyperintense excrescence intradurally and a hyperintense lesion within the clivus. Both types can have T2 hypointensity which may present the stalk, seen surgically or on CT images (black triangle). Incomplete EP, with only EP bud (C), seen as hypointensity on T2 weighted images. EP variant (D), hyperintense lesion in clivus.

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Fig. 8: Ecchordosis physaliphora. Axial CISS image (A), axial CISS image (B) from another level, showing T2 hypointensity (arrow), sagittal reformatted CISS image (C), axial T2 weighted image (D), showing intraclival compartment of EP.

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**Fig. 9:** Ecchordosis physaliphora. Transverse CISS image (A), sagittal reformatted CISS image; white arrows show T2 hypointensity (EP bud). Coronal CISS image (C). Axial T2 image (D), axial CISS image from another level (E), shoe intradural and intraosseous components of EP (arrows). Axial post contrast image (F), lesion shows no enhancement.

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**Fig. 10:** Benign notochord cell tumor, axial T2 weighted image. Note that the lesion has no extraosseous component.

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**Fig. 11:** Chordoma. Axial CT image (A), with bone destruction. Axial T2 weighted image (B) and sagittal T1 weighted image (C). Post contrast axial image (D) shows the enhancement of the tumor.

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**Table 1:** Features of benign and malignant notochord derived lesions.

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Conclusion

Benign notochord derived lesions and notochord remnants are rarely seen but it is important to differentiate them from chordoma, as chordoma is aggressively treated.
References


