Tumors and tumorlike lesions of the craniovertebral junction

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Learning objectives

1. Describe the common neoplasms involving of the CVJ.
2. Identify the possible differential diagnosis from the anatomic location.
3. Discuss potential pitfalls and mimics that can be mistaken for neogrowths

Background

The craniovertebral junction (CVJ) is a complex articulation comprised of the clivus, foramen magnum and upper two cervical vertebrae. It is the potential site of a variety of radiologic diagnoses and misdiagnoses.

Neoplasms that arise within the CVJ consist of osseous tumors, nervous system and those extending from surrounding soft tissue.

There is no single symptom or neurological finding pathognomonic for a lesion in this location. The role of imaging is to determine the nature and resectability of the tumors, such as tumor extent, direction of encroachment, whether the lesion has a vascular or intramedullary component and craniovertebral stability.

Imaging findings OR Procedure details

Neoplasms of the CVJ mainly arise from extradural osseous and soft tissues. Those arising from the intradural extramedullary structures include those from neural coverage, that is, meningiomas, nerve sheath tumors, and some congenital cystic
lesions. Intramedullary tumors include glial and nonglial tumors, such as ependymomas, astrocytoma and hemangioblastoma.

**NEOPLASMS FROM LOCAL BONE AND SOFT TISSUES**

The most common primary malignant bone tumors are chordoma, myeloma, lymphoma, chondrosarcoma, osteosarcoma, Ewing's sarcoma. The most common benign primary bone tumors of the CVJ and the cervical spine in the adults are, in decreasing occurrence, ABC, benign giant cell tumor, osteoblastoma, eosinophilic granuloma, solitary plasmacytoma, and hemangioma.

Chordomas are locally aggressive tumors from primitive notochordal remnants. They can occur the vertebral axis, especially for the sphenoccipital region and the sacrum. Sometimes, it extends from its classic midline position to involve the jugular fossa and foramen magnum and cause erosion of the C1 and other cervical vertebras.

CT can depict well-defined expansile soft tissue mass from clivus with lytic bone changes. Intratumoral calcifications are common. On the MRI, the typical findings are T2W bright lesion with some intratumoral hemorrhage/mucus pools and post-Gd avid enhancement. (Fig. 2-3)

Chondrosarcomas may have similar imaging appearance to choromas. Most of them are low-grade lesions. These chondroid lesions are typically located in the paramedian petroclival synchondrosis, but they rarely occur in the midline associated with the sphenooccipital synchondrosis.

Plasmacytoma and metastases are common malignant lesions found in the CVJ. Myeloma cell tumors in the CVJ can arise from the nasopharyngeal submucosal tissue (extramedullary plasmacytoma) or the marrow space of the clivus. Plasmacytomas reveal homogeneous signal intensity on T1- and T2-weighted images.

Osteosarcomas arise from the CVJ are very rare. Local pain is usually the presenting symptom. CT reveals ossified soft tissue mass with bone lytic change.

Nasopharyngeal carcinoma (NPC) is a common primary head and neck malignancy in southern China and Southeast Asia. Skull base involvement is a common findings in NPC. Rarely patients who have NPC may present with symptoms of CVJ tumors and multiple cranial neuropathies and few or no mucosa-associated symptoms. (Fig. 5)

**INTRADURAL EXTRAMEDULLARY NEOPLASMS**

Intradural extramedullary neoplasms of CVJ mostly originate from neural coverings, including meningioma, nerve sheath tumor and congenital cystic lesions. As meningiomas are the most common primary nonglial intracranial tumors, they are also the most common tumors at the foramen magnum (3% of all meningiomas). Ten
patients of foramen magnum meningiomas have extradural extensions. The lesions are typically attached at the anterior rim of foramen and may invade nearby vertebral artery and cervical nerve roots. Like those at other intracranial locations, the meningiomas usually appear intermediate T1- and T2- weighted images and homogeneous and strong enhancement. (Fig. 6)

The second most common nonosseous tumors of the CVJ are neurilemmomas. They usually originate from lower cranial nerves and upper cervical roots. Besides the typical intradural extramedullary location, they can be completely extradural (15%) or both intra- and extradural ("dumbbell" shape). Neurilemmomas show hyperintense signal intensity on T2-weighted imaging with occasional "target" sign, which represents denser areas of collagen and Schwann cells. (Fig. 7)

Congenital cystic lesions of the CVJ include arachnoid, epidermoid, and neurenteric cysts. Arachnoid cysts are rare lesions in the CVJ. On MRI, the cysts exhibit signals identical to CSF on all pulse sequences. They may exert a mass effect on adjacent brain or bone.

**INTRAMEDULLARY NEOPLASMS**

Common neoplasms of the CVJ include ependymomas, astrocytomas and hemangioblastomas. Ependymomas are the most common intramedullary neoplasms in the adults. A variety of histological subtypes of intramedullary ependymomas have been described. The cellular ependymoma is most common. Though these tumors lack a true capsule, they are generally well circumscribed and do not infiltrate surrounding spinal cord tissue. They are almost always benign. The typical imaging findings is a circumscribed, enhancing cord mass with hemorrhage. Polar (rostral or caudal) or intratumoral cysts are commonly observed (50-90%).

Cervicomedullary gliomas are a distinct subset of brainstem glioma and generally considered as cervical cord tumors with rostral extension into the caudal medulla. Most are low-grade (80-90%). MRI demonstrate mild/intermediate enhancing infiltrative mass with asymmetric or even exophytic growth and often associated cord expansion. (Fig. 8)

Hemangioblastomas (HBs) constitute 1.0%-7.2% of all spinal cord neoplasms and show no gender predilection. These lesions are typically seen as intramedullary mass with round, well-defined margins and serpentine "flow voids" at posterior aspect of the spinal cord (Fig. 9). Most cord HBs are solitary and occur in patients younger than 40 years, usually with impaired proprioception. Multiple lesions indicate the manifestations of von Hippel-Lindau syndrome.
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**Fig. 1:** Common neoplasms that arise from the craniovertebral junction.
Fig. 2: A 39-year-old woman presented with left-side multiple lower cranial neuropathy and tinnitus. MRI depicted a T1W hypointense, T2W and avid-enhancing chordoma at left clivo-occipital junction and jugular fossa.
**Fig. 3:** A 12-y/o-girl of clival chordoma presented with left-side ptosis, slurred speech and dysphagia.
Fig. 4: Osteosarcoma in a 23-year-old man with torticollis. Axial and coronal CT show increased osteoblastic change and periosteal reactions at C1 lateral mass and transverse process. MRI shows a necrotic mass of T1-weighted and T2-weighted intermediate signal intensity and prominent enhancement after gadolinium administration.
Fig. 5: Nasopharyngeal carcinoma in a 63-year-old man with initial presentation of lower cranial neuropathies. MRI depicts a infiltrative mass in the clivus with intracranial extension. Scanty mucosal lesion is found.
**Fig. 6:** MRI of a 28-year-old man demonstrates a well-demarcated meningioma attached to the anterior margin of foramen magnum with severe brainstem and cord compression. Dural tails are depicted at the rostral and caudal aspect of the lesion.
Fig. 7: Neurofibroma in a 35-year-old man of type I neurofibromatosis. One intra- and extradural dumbbell-shaped mass is depicted at left C1-2 intervertebral foramen, which shows hyperintense on T2-weighted imaging and homogeneous enhancement after gadolinium administration.
**Fig. 8:** Low-grade astrocytoma in a 30-year-old woman. MRI depicts an infiltrative T2-weighted hyperintense tumor involving brainstem and upper cervical cord as well as cord expansion.
**Fig. 9:** One lobulated avid-enhanced intramedullary hemangioblastoma at the posterior aspect of C1-4 spinal cord. Associated serpentine flow voids are seen in the tumor and regional perimedullary space/brainstem.
Conclusion

Knowledge of the pathologic features of these tumors and how these features are reflected in their imaging appearances may help radiologists differentiate them.

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References


