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Chordomas: From Stem to Stern

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After participating in this educational activity, the diagnostic radiologist should be able to identify the usual sites of origin and imaging features of chordomas and describe their clinical presentations, treatment options, and differential diagnoses.

Category: Neuroradiology
Modality: Multiple

Key Words: Chordomas, Clinical Presentation of Chordomas, Imaging of Chordomas, Treatment of Chordomas

Although rare, chordomas can affect the entire axial skeleton, and they are the most common primary malignant bone tumor in both the sacrum and the spine. This article details tumor origin, epidemiology, pathology, and imaging features, in an effort to improve the ability of the radiologist to diagnose, provide a differential diagnosis, and further understand treatment of patients with chordomas.

Origins

Chordomas are rare tumors thought to arise from vestigial or ectopic remnant notochord tissue, and they span intracranially (clivus) to the caudal end of the axial skeleton (sacrum).[1] The nucleus pulposus is the only persistent adult derivative of the notochord. Occasionally, notochordal cells remain in the nucleus pulposus within mature intervertebral discs and are known as notochordal rests or vestiges,[2,3] from which tumors may arise.[1] Alternatively, rests in notochord-like tissue in the intravertebral region, which develop into benign notochordal cell tumors, may transform into malignant variant chordomas.[1-3] These notochordal remnants are in the midline, accounting for the midline or near-midline location of spinal chordomas.

Skull base chordomas form slightly differently. Three cartilage precursors (parachordal, hypophyseal, and prechordal) fuse with occipital sclerotomes to create a cartilaginous chondrocranium intermediate, which then undergoes endochondral ossification into ethmoid, sphenoid, occipital, and temporal components of the skull base.[1] Growth and cartilage ossification seem to be regulated by signaling pathways in nearby notochordal rests, which again are thought to be the source of skull base chordomas.[1] The spheno-occipital synchondrosis is a common site of chordoma origin.[4]

Incidence

Overall incidence rates have been estimated at 0.08 per 100,000 or approximately 1 per 1,000,000 persons, with males slightly more affected than females.[4,5] Although chordomas have been diagnosed at almost every age, the median age is estimated at 58.5 years, with the largest proportion of tumors in patients older than 50 years.[5] Of note, the median age at diagnosis of intracranial chordomas is younger than that of all other sites, and the oldest median age is associated with sacral chordomas,[6] with median ages of 49 and 69 years, respectively.[5]

Prognosis

In a review of 400 cases from 1973 to 1995, the overall 5- and 10-year survival rates for all races and sexes were 65% to 67.7% and 35% to 39.9%, respectively.[5] Additional research has shown 5-year survival rates of 61.6% for patients with skull base chordomas compared with 90.4% for those with nonskull base chordomas.[6] Survival of patients with nuclear pleomorphism (i.e., tumors usually of the dedifferentiated subtype) is significantly poorer than in patients without nuclear pleomorphism.[6]

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Features

Historically, chordomas were thought to be most frequent in the sacrum, followed by the skull base and vertebrae, but data now suggest a more equal distribution in the skull base (32.0%), mobile spine vertebrae (32.8%), and sacrum (29.2%).[4,7] Chordomas usually are considered low-grade neoplasms; however, frequently they invade locally and often recur.[7] Within the skull, origination from the spheno-occipital synchondrosis[4] leads to involvement of the clivus and sinuses. Small chordomas may be detected incidentally before they are large enough to cause symptoms from mass effect (Figure 1). Intradural extension is common, and chordomas may invade the petrous bone, epidural space, basal cisterns, cerebellopontine angle, and retropharynx, thereby causing mass effect on the pituitary, hypothalamus, optic chiasm, cranial nerves, and brainstem. Intracranial carotid artery encasement may occur, but occlusion is uncommon (Figure 2). Distant metastases are rare, but they can involve the lungs, liver, lymph nodes, and other bones.[4]

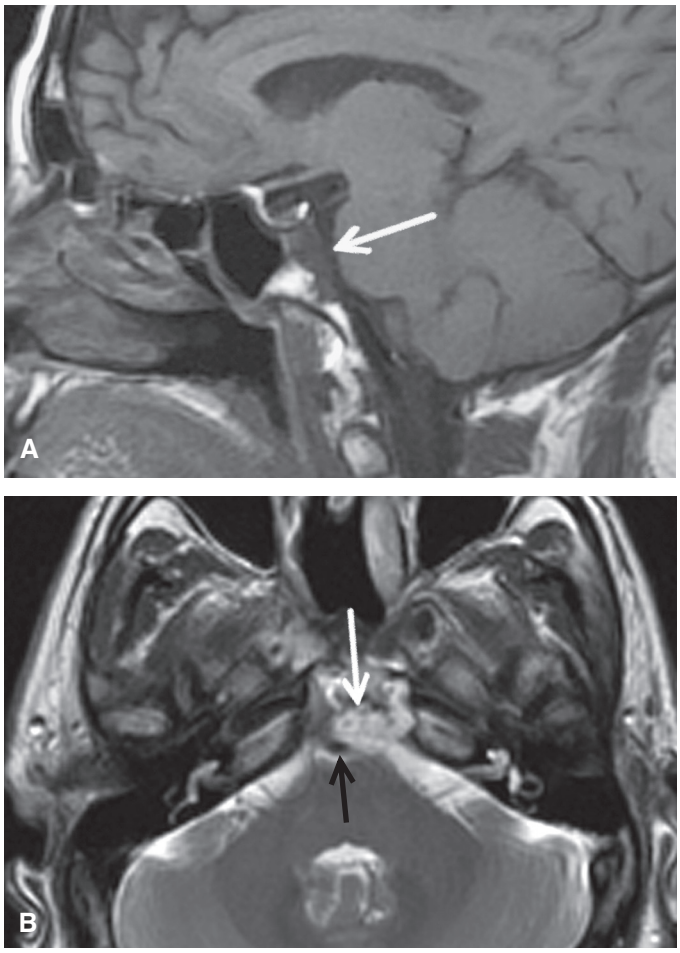

Figure 1. A 60-year-old woman with small early-stage clival chordoma. *A*: Sagittal, T1-weighted MR image shows irregular low-signal mass (*arrow*) in posterior superior clivus. *B*: Axial, T2-weighted MR image shows a high T2-signal mass in the posterior superior clivus (*white arrow*) with minimal extension into prepontine cistern but no mass effect on the basilar artery (*black arrow*) or other vessels.

Intracranial chordomas may encase an intracranial carotid artery, but occlusion of the artery is uncommon.

Within the spine, intervertebral discs are preserved initially, with subsequent invasion of the disc spaces, as tumor extends to adjacent vertebral bodies. There is often mass effect on foraminal nerve roots and/or the central spinal canal and spinal cord. However,

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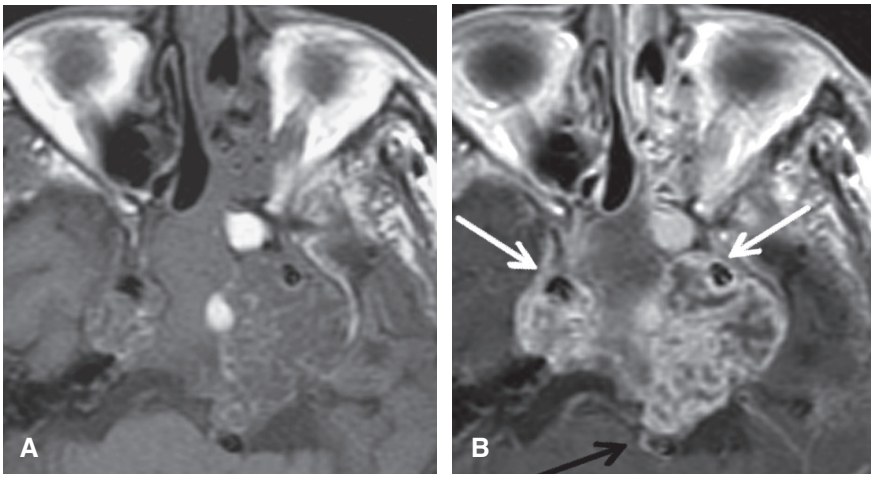
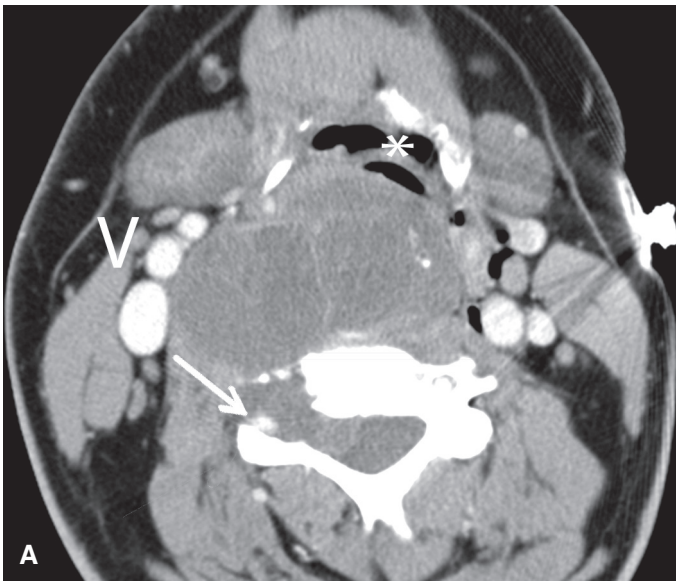


Figure 2. A 73-year-old man with large, invasive skull base chordoma. *A*: Axial, T1-weighted MR image shows a mass with T1 hypointense signal. *B*: Axial, postcontrast T1-weighted MR image shows invasion of cavernous sinuses with displacement and partial encasement of internal carotid arteries (*white arrows*). Signal flow void indicates patency. There is mass effect on the left cerebellopontine angle abutting the basilar artery (*black arrow*).

encasement of vertebral arteries in the cervical spine is infrequent.



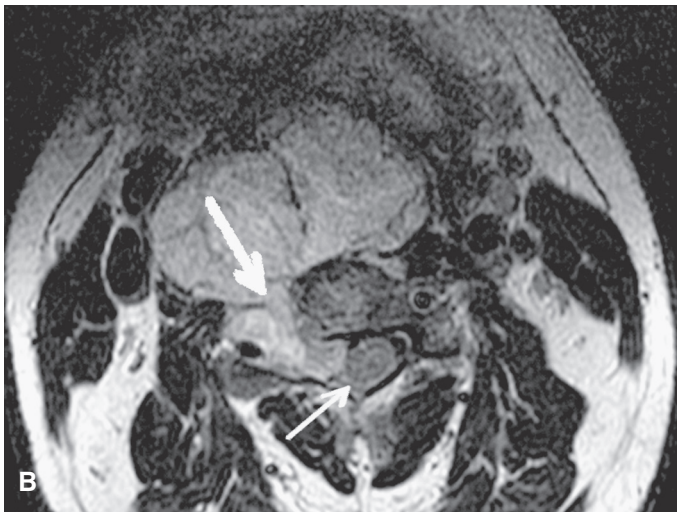


Figure 3. A 29-year-old man with large, invasive cervical spine chordoma. *A*: Axial, enhanced CT scan shows displacement and partial encasement of the right vertebral artery (*arrow*), displaced airway and esophagus (*asterisk*), and displaced right carotid artery and jugular vein (letter V). *B*: Axial, T2-weighted MR image shows the prevertebral mass extending through C3/C4 right neural foramen (*thick arrow*), destroying articular pillars, and extending into spinal canal with deformity and displacement of cord (*thin arrow*) to left.

Clinical Presentation

Clinical manifestations depend on tumor location, size, and mass effect. Chordomas often are clinically silent until later in the clinical course due to indolent, slow growth. Skull base tumors can present with cranial nerve palsies, epistaxis, and/or endocrinopathy. Spinal chordomas vary in presentation. The level of involvement within the cervical (Figure 3), thoracic (Figure 4), or sacral (Figure 5) spine dictates the distribution of symptoms from cord/nerve compression. Anterior invasion in the cervical spine can lead to dysphagia and a variety of respiratory symptoms from mass effect on the trachea and/or larynx (Figure 3A). Posterior invasion can create localized pain, radiculopathies, and sensory/motor symptoms. Sacrococcygeal tumors often present as dull pain worse with sitting. Cauda equina involvement can result in saddle anesthesia, leg paresthesias, motor weakness, and bladder and bowel dysfunction.

Pathology

Chordomas are histologically subdivided into classical (conventional), chondroid, and dedifferentiated types.[8] Gross specimens are tan, soft, and myxoid in consistency, with areas of necrosis, recent and old hemorrhage, and entrapped bone.[8] Conventional chordomas are composed of cords of tumor cells in lacunae embedded in hyaline cartilage-like mucopolysaccharide stroma. Cytoplasm is granular and eosinophilic in most tumor cells, which characteristically contain numerous intracytoplasmic vacuoles (physaliferous cells) arranged in nests, cords, or sheets within stroma.[8] Immunohistochemistry stains are positive for S-100 protein, cytokeratin, and epithelial membrane antigen (EMA) but negative for carcinoembryonic antigen (CEA) reactivity.[8]

Chondroid subtypes are characterized by islands of tissue histologically identical to hyaline cartilage matrix, coupled with foci of conventional chordoma cells.[8] Dedifferentiated subtypes demonstrate areas of conventional chordoma with chondroid or high-grade sarcomatous transformation[8] and spindle-shaped, pleomorphic hyperchromatic nuclei.

Imaging

Initial radiographs of the skull and spine may demonstrate osteolytic lesions with or without secondary sclerosis, depending on the age and progression of the tumor. Initial chest radiographs may demonstrate a posterior mediastinal mass of a thoracic chordoma. CT and MRI are needed for further localization, delineation of tumor

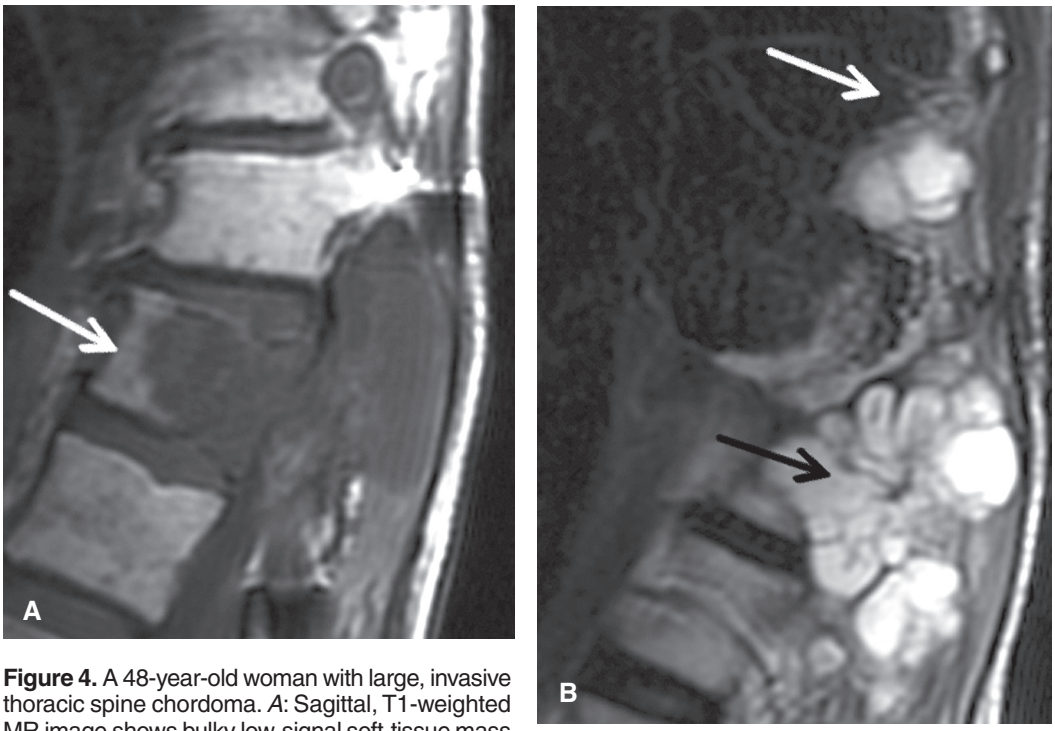


Figure 4. A 48-year-old woman with large, invasive thoracic spine chordoma. *A*: Sagittal, T1-weighted MR image shows bulky low-signal soft-tissue mass involving T12 vertebral body (*arrow*) extending into posterior elements and replacing normal bone marrow. *B*: Sagittal, T2-weighted MR image shows high-signal chordoma of T12 vertebral body (*black arrow*) with mass effect on right neural foramen. Paraspinal component extends cranially beyond image margins (*white arrow*).

margins, and evaluation for invasion of surrounding anatomic structures.

CT evaluation of chordomas demonstrates tumors of homogeneous or heterogeneous attenuation, and they can be quite subtle (Figure 6A). Bone involvement and calcifications are best shown by CT, whereas soft tissue extension is better shown by MRI. On CT, skull base tumors are more often isodense to brain parenchyma than hypodense or hyperdense.[9] Tumors almost always enhance, although the degree of enhancement varies.[9] Most masses demonstrate extensive osteolysis with or without sequestra or dystrophic calcification (Figure 5).[9] Calcifications can be nodular or flake-like and are seen in up to 50% of cases.[10] Irregular intratumoral calcifications are felt to represent sequestra from bone destruction rather than dystrophic calcification.[4] Chondroid variants are more likely to demonstrate true intratumoral calcifications.[4]

Intracranial chordomas typically are located centrally and are expansile, with a soft tissue mass arising from or near the clival region (Figures 1 and 2). Spine lesions may have associated vertebral body and/or posterior element collapse and/or subluxation. CT is particularly useful when the inferior clivus is involved and can show the degree of occipital condyle erosion, which is critical in determining the need for surgical fusion.[9] CT myelography or MRI can be used to more accurately evaluate epidural extension, degree of compression of neural elements, and presence of subarachnoid metastases.

MRI is the modality of choice to evaluate intradural extension.[9] Sagittal images best define posterior tumor margins. Coronal sequences demonstrate relationships of tumors with the optic chiasm and/or extension into the cavernous sinus. Tumors may extend into the sphenoid sinus, ethmoid air cells, sella turcica, suprasellar cistern, nasopharynx, parapharyngeal space, hypoglossal canal, jugular foramen, and prevertebral space. Skull base chordomas and chondrosarcomas often exhibit similar MR features such that the two cannot be differentiated accurately.[9]

Chordomas, regardless of location, are predominantly T1 iso- or hypointense (Figures 1A, 2A, and 4A) and T2 hyperintense (Figures 1B, 3B, 4B, and 6B–6C) with enhancement ranging from minimal to intense (Figures 2B and 6D).[4,9,10] Osseous destruction

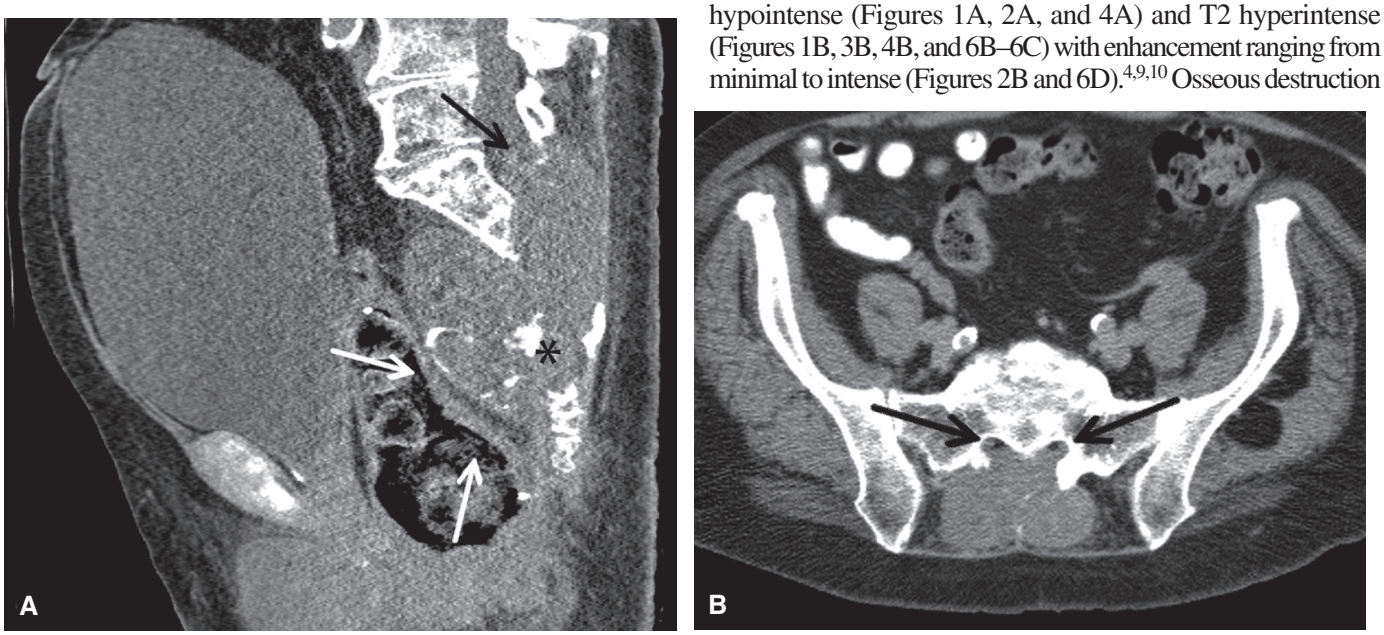


Figure 5. A 65-year-old man with large osteolytic sacral chordoma. *A*: Sagittal CT scan shows mass effect displacing rectum anteriorly (*white arrows*) and mass effect on central spinal canal (*black arrow*). Multiple calcifications (*asterisk*) could be dystrophic or intratumoral sequestra. *B*: Axial CT scan shows a large osteolytic sacral mass with mass effect on bilateral sacral neural foramina (*arrows*).

appears as cortical bone signal void replaced by soft-tissue signal intensity.[4] Heterogeneity dominates over homogeneity, which is thought to be due to variable signals from mixed-age hemorrhage, trabeculae, and compactness of tumoral cellular arrangement.[10] Calcifications obvious on CT may be seen only as intrinsic, inhomogeneous, low MR signal changes.[10] Focal areas of high signal intensity on T1 images usually are due to highly proteinaceous material such as intratumoral hemorrhage or mucous pools.[4,10] Gradient-echo imaging similarly can confirm blood products.[4] Low-signal septations with tumor lobulations are most appreciable on the background of predominantly high signal on T2 images (Figures 3B and 4B).[4,9] Fat suppression aids in differentiating enhanced tumor margins from bright fatty bone marrow.[4]

MR angiography (MRA) augments evaluation of vascular encasement, although arterial narrowing is uncommon due to the generally soft nature of chordomas.[4] Conventional angiographic evaluation is nonspecific but better demonstrates the degree of luminal narrowing, and it usually is reserved for patients where there is significant encasement or narrowing of arteries on MRA.[4] Tc-99m bone scintigraphy usually demonstrates reduced or normal radiotracer uptake.[11]

Treatment

Close tumor proximity to vital organs in the skull and spine makes treatment challenging, coupled with the fact that many tumors are resistant to conventional chemotherapies. Surgical resection of as much tumor as possible is preferred treatment, as longer survival rates have been associated with more extensive tumor removal.[4] Unfortunately, many chordomas recur as complete surgical resection is difficult (Figure 6). Typical surgical techniques include cranio-orbitozygomatic, transcondylar, and transmaxillary approaches for tumors extending from the upper clivus, inferior clivus, and into the nasopharynx or craniocervical junction.[4] Sacral chordoma en-bloc resections involve posterior-transperineal approaches or a combination of anterior and/or posterior approaches. Complete en bloc resection is obtained in only approximately 50% of sacral chordomas, with much lower rates intracranially and within the spine.[7]

Residual tumor frequently is treated successfully with radiation therapy.[4] However, tolerance doses of structures adjacent to chordomas, frequently the cranial nerves, brainstem, spinal cord, and rectum, are much lower than effective doses to treat chordomas.[7] By using hadrons, which are high-dose protons or charged particles such as carbon or helium ions, higher doses can be used with less of a negative effect on adjacent tissues and organs.[7] Unfortunately, higher costs and limited availability often restrict their use.

Differential Diagnosis

The differential diagnosis of chordoma includes chondrosarcoma, chondroma, meningioma, plasmacytoma, lymphoma, and metastatic adenocarcinoma.[10] Chondrosarcomas most closely resemble chordomas, both appearing as lobulated soft tissue masses with similar, variable MR signal intensity and enhancement.[12] However, chondrosarcomas arise from primitive mesenchymal cells or embryonic rests of cartilaginous matrix, containing pleomorphic chondrocytes rather than physaliferous cells. Location also usually differs as chordomas are more commonly centered midline at the clivus, whereas chondrosarcomas are off midline either in the petrous bone or at the sphenopetroclival or petroclival regions.[12]

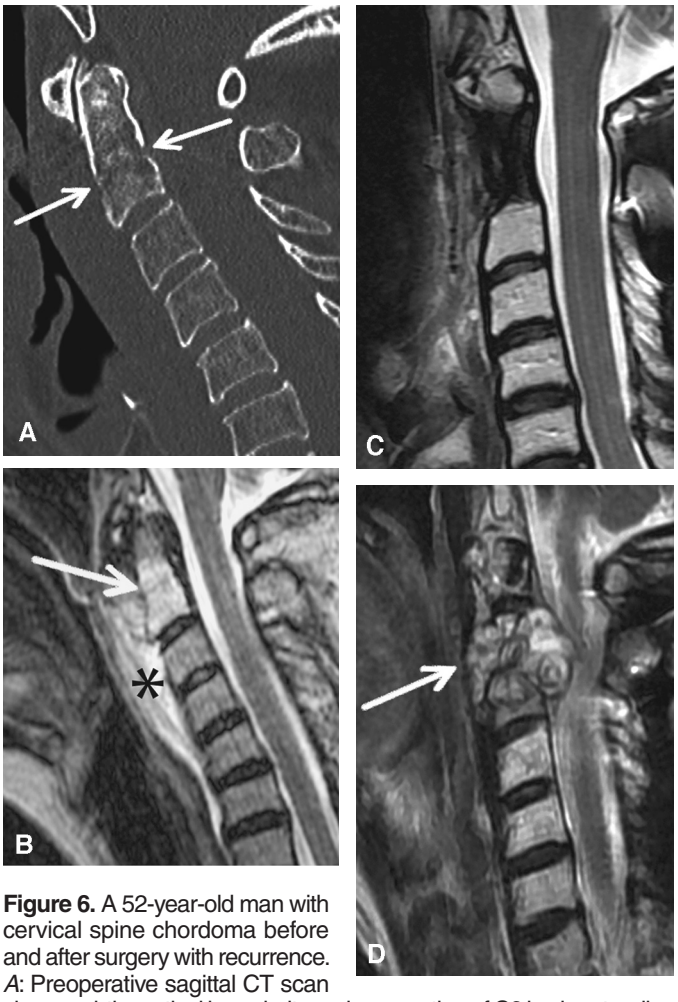


Figure 6. A 52-year-old man with cervical spine chordoma before and after surgery with recurrence. *A*: Preoperative sagittal CT scan shows subtle cortical irregularity and permeation of C2 body extending into odontoid process (*arrows*). *B*: Preoperative sagittal, T2-weighted MR image shows high signal of C2 (*arrow*) with C2-C6 prevertebral soft tissue thickening (*asterisk*). *C*: Sagittal, T2-weighted MR image shows no recurrence 2 years after surgical resection and fusion from occiput-C4. *D*: Sagittal, T1-weighted, postcontrast MR image 1 year later shows recurrence with new lobulated, heterogeneous signal at the surgical site (*arrow*) expanding into the ventral central spinal canal.

Meningiomas may mimic intra-axial chordomas in location, but close inspection should reveal a dural attachment indicative of extra-axial origin with osseous sclerosis instead of destructive invasion. Contrast enhancement is more frequently diffusely homogeneous than would be routinely expected in chordomas. Calcifications can occur. Plasmacytoma and lymphoma, if located intracranially and centrally, can invade and cause lytic bony destruction. Skull base metastases should be considered in the presence of a primary malignancy.

Immunohistochemical stains frequently are used to provide a diagnosis when imaging and clinical features overlap.[10] Chordomas are usually S-100, cytokeratin CAM5.2, and EMA positive but CEA negative.[10] Chondromas and chondrosarcomas are both S-100 positive but usually CAM5.2, EMA, and CEA negative.[10] Meningiomas are more likely to be EMA and CEA positive but CAM5.2 negative.[10] Metastatic adenocarcinoma is usually CAM5.2 and EMA positive but S100 negative.[10]

Conclusion

Chordomas are rare axial skeleton tumors of notochord tissue origin, which arise in the skull, spine, and sacrum. This CME activity emphasizes that CT and MRI are central to diagnosis

and treatment planning. Treatment depends on size, location, and degree of involvement of critical adjacent structures.

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1. All of the following are CT features of skull base chordomas, *except*
- **A.** nonenhancement
- **B.** isodense to brain parenchyma
- **C.** extensive osteolysis
- **D.** sequestra
- **E.** dystrophic calcifications

2. Which one of the following is the *best* imaging modality for evaluation of intradural extension of a chordoma?
- **A.** CT
- **B.** MRI
- **C.** PET/CT
- **D.** Ultrasound
- **E.** Plain radiography

3. All of the following are MR features of chordomas, *except*
- **A.** T2 hyperintense
- **B.** T1 hypointense
- **C.** minimal enhancement
- **D.** intense enhancement
- **E.** homogeneity more common than heterogeneity

4. Which one of the following imaging modalities is *most* helpful for evaluation of a chordoma involving the inferior clivus?
- **A.** MRI
- **B.** Ultrasound
- **C.** CT
- **D.** Plain radiography
- **E.** Cerebral angiography

5. Which one of the following is the initial preferred treatment for chordomas?
- **A.** Radiation therapy
- **B.** Conventional chemotherapy
- **C.** Steroid therapy
- **D.** Surgical resection of as much tumor as possible
- **E.** Watchful waiting until symptoms become incapacitating

6. All of the following immunohistochemistry stains are positive in classical (conventional) chordomas, *except*
- **A.** CEA reactivity
- **B.** S-100 protein
- **C.** EMA
- **D.** cytokeratin

7. All of the following malignant tumors should be considered in the differential diagnosis of a skull base chordoma, *except*
- **A.** chondrosarcoma
- **B.** meningioma
- **C.** lymphoma
- **D.** metastases
- **E.** oligodendroglioma

8. Extension of intracranial chordomas may involve
- **A.** intradural space
- **B.** petrous bone
- **C.** basal cisterns
- **D.** retropharynx
- **E.** all of the above

9. All of the following are potential clinical manifestations of chordomas, *except*
- **A.** cranial nerve palsies
- **B.** early onset of symptoms when the tumor is small
- **C.** bladder dysfunction
- **D.** endocrinopathy
- **E.** sensory/motor symptoms

10. Which one of the following statements regarding chordomas is *false*?
- **A.** They can arise from the nucleus pulposus.
- **B.** They are rare tumors.
- **C.** They usually affect the sacrum at an earlier age than they affect the clivus.
- **D.** They are midline axial skeletal tumors.
- **E.** The spheno-occipital synchondrosis is a common site of intracranial origin.