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Imaging features of cartilaginous tumors of the head and neck

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ABSTRACT

There is a wide spectrum of head and neck cartilaginous lesions which include both neoplastic and nonneoplastic processes. Cartilaginous tumors of the head and neck are uncommon, posing a diagnostic challenge. Benign cartilaginous tumors that may occur in the head and neck include chondroma, chondroblastoma, chondromyxoid fibroma, osteochondroma, and synovial chondromatosis. Chondromesenchymal hamartoma is a rare non-neoplastic cartilaginous lesion that is included for the 1first time in the new WHO classification and radiologically can mimic a tumor. Malignant cartilaginous tumors include chondrosarcoma and chondroid variant of chordoma. Characteristic tumor locations, internal chondroid matrix calcification, and typical T2 hyperintense signal secondary to high-water content within the extracellular matrix of the hyaline cartilage are useful imaging features that narrow the differential diagnosis and help in diagnosing these diseases. This article presents a narrative review of the anatomy of the head and neck cartilaginous structures, discusses the current knowledge and imaging spectrum of benign and malignant cartilaginous tumors and tumor-like lesions of the head and neck.

Keywords: Cartilage, Cartilaginous lesions, Cartilaginous tumors, Chondroid, Head and neck

INTRODUCTION

Radiologic diagnosis of cartilaginous tumors of the head and neck can be challenging due to their rarity.^[1] Their imaging appearance may overlap with other pathologies in the same location, further adding to the diagnostic dilemma. Thus, an understanding of the anatomy of head and neck cartilaginous structures is critical to correctly identifying the spectrum of cartilaginous lesions occurring in the head and neck. This review article describes the anatomy of the head and neck cartilaginous structures and radiologic features of benign and malignant cartilaginous tumors of the head and neck.

ANATOMY OF THE CARTILAGINOUS STRUCTURES OF THE HEAD AND NECK

Cartilage is an avascular connective tissue that plays a pivotal role in the growth and development of the human skeleton and histologically is composed of specialized cells, called chondrocytes. Chondrocytes can synthesize and secrete different components of the extracellular matrix (ECM), such as collagens, proteoglycans, and glycoprotein, which provide tensile strength and

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flexibility to tissue.^[2,3] The perichondrium is a thin fibrous membrane covering the external surface of the cartilage and containing superficial blood vessels. These vessels provide nutrition to the cartilage by means of diffusion.^[3]

In humans, there are three main types of cartilage: Hyaline cartilage, elastic cartilage, and fibrocartilage, each of which has different types and proportion of collagens and proteoglycans.^[2] Hyaline cartilage is the most abundant cartilage in the human body, found primarily at the articular surfaces of the synovial joints and at the epiphyseal plates of long bones. It facilitates joint movement and, in the head and neck region, is present at the skull base, within the nasal septum, the laryngeal cartilages, and in the tracheal rings.^[2] Ossification of the laryngeal cartilages increases with advancing age [Figure 1]. Male laryngeal cartilages tend to ossify to a greater extent compared to females.^[4,5] Nonuniform ossification of the laryngeal cartilages is one of the pitfalls in evaluating cartilaginous invasion in patients with laryngeal cancer. It is very important to be familiar with this normal variation to avoid over staging of tumors.^[6]

Elastic cartilage, as the name implies, has high elasticity due to the elastin fibers. In the head and neck, elastic cartilage is predominantly found in the epiglottis, pinna of the external ear, external auditory canal, and the eustachian tube.^[2] Unlike the hyaline cartilage which forms the thyroid, cricoid and arytenoid cartilages, the elastic cartilage of the epiglottis does not undergo ossification with age.^[7] Fibrocartilage is a poorly vascularized tissue composed of fibroblasts and chondrocytes. It has a high density of collagen Type I which provides its high tensile strength.^[8,9] It is predominantly found at the sites of ligamentous and tendinous attachments, intervertebral discs, symphyses, synchondroses, and menisci.^[8,9] In the head and neck region, fibrocartilage is found in the articular disc and articular surfaces of the temporomandibular joint, as well as spheno-petrosal and petro-occipital synchondroses at the skull base.^[8] Embryonic remnants of cartilaginous matrix within these synchondroses can be origins of cartilaginous tumors in the skull base.^[10] [Table 1] summarizes the types of cartilage and related cartilaginous structures in the head and neck.

IMAGING FEATURES OF CARTILAGINOUS TUMORS IN THE HEAD AND NECK

The 2017 4th edition of the WHO classification of head and neck tumors is the most updated pathologic classification of head and neck tumors and tumor-like conditions.^[11] Benign cartilaginous tumors that may occur in the head and neck include chondroma (aka enchondroma), chondroblastoma, chondromyxoid fibroma (CMF), osteochondroma, and synovial chondromatosis. Chondromesenchymal hamartoma is a rare non-neoplastic cartilaginous lesion that is included for the 1st time in the new WHO classification^[12] and is



Figure 1: Physiologic ossification of the thyroid cartilage. Axial contrast-enhanced neck CT in soft tissue window of a 15-year-old boy (a) and a 41-year-old man (b) show physiologic ossification of the thyroid cartilage which increases with age. Note absence of ossification of the thyroid cartilage (arrow) on (a) and non-uniform ossification of the thyroid cartilage (arrow) on (b).

Table 1: Head and neck structures derived from each type of cartilage.

Types of cartilage	Head and Neck Structures
Hyaline cartilage Elastic cartilage	Skull base, nasal septum, larynx, trachea Epiglottis, external ear (that is, ear pinna and outer cartilaginous portion), the eustachian tube of the middle ear
Fibrocartilage	Intervertebral disks of the spine, articular disc and articular surface of the temporomandibular joint, spheno-petrosal and petro-occipital synchondroses of the skull base

included in this discussion, because it can mimic a tumor. Malignant cartilaginous tumors include chondrosarcoma and chondroid variant of chordoma. [Table 2] lists the cartilaginous tumors and tumor-like lesions affecting the head and neck and their characteristic imaging features.

In general, cartilaginous tumors with well-differentiated chondroid matrix frequently contain internal calcification which can be visualized on plain radiography and CT. Typical calcification patterns of chondroid matrix are ring-andarc-like or popcorn-like which correlate with histological enchondral ossification.^[13] On MRI, cartilaginous tumors typically have intermediate signal on T1WI and high signal on T2WI reflecting the high-water content within the ECM of the hyaline cartilage. The areas of dense calcification are hypointense on both T1- and T2WI.^[13-15] The lesions may enhance avidly, and the intensity of enhancement varies with the extent of the calcific component [Figure 2]. Studies on the diagnostic utility of FDG PET/CT have produced conflicting results with some studies indicating limited utility of FDG PET in differentiating benign tumors from low-grade chondroid malignancies because of significant overlap of SUV max, while

Table 2: Characteristic imaging features of cartilaginous tumors of the head and neck.			
Cartilaginous lesions	Common locations	Characteristic imaging features	
Benign cartilaginous tumors and tumor-like lesions			
Chondroma (aka enchondroma)	Cricoid and thyroid cartilages	Well-defined expansile hypodense lesions with variable amounts of coarse, chondroid, mottled calcification Difficult to distinguish from chondrosarcoma on imaging	
Chondroblastoma	Skull base (the squamous portion of the temporal bone)	Nonspecific well-defined expansile bone lesions containing a variable amount of internal chondroid matrix calcification Intratumoral cystic degeneration or secondary aneurysmal bone cysts in one third of tumors	
Chondromyxoid fibroma	Skull base, sinonasal cavity	Well-defined, expansile bone lesions with a sclerotic rim, intralesional calcification approximately 10% of tumors	
Osteochondroma	Mandible, particularly mandibular condyle and coronoid process	Exophytic bone lesions with continuity of bone marrow from the parent bone into the tumor and cartilaginous cap at the periphery Abnormal thickening of the cartilaginous cap (exceeding 1.5–2.5 cm) indicating malignant degeneration	
Synovial chondromatosis	Temporomandibular joint	Joint effusion, abnormal synovial thickening and enhancement suggestive of synovitis, adjacent bone erosion due to mechanical pressure, and calcified or noncalcified intra-articular loose bodies	
Chondromesenchymal hamartoma	Sinonasal cavity, orbit	Rare benign, slow-growing, locally destructive, tumor-like lesion Associated with pathologic <i>DICER1</i> genetic variants Expansile heterogeneous mixed solid and cystic masses, internal calcifications in 50% of lesions	
Malignant cartilaginous tumors			
Chondrosarcoma	Skull base (particularly petroclival fissure) and cricoid cartilage	Expansile soft tissue masses with bone erosion and invasion of adjacent structures, intratumoral calcification up to 50% of skull base chondrosarcomas MRI - heterogeneous T2 hyperintense signal with a variable degree of contrast enhancement Tumor location - more frequently located off- midline near the petro-occipital fissure	
Chondroid chordoma	Clivus	Similar to conventional chordoma including expansile soft tissue mass with adjacent bone erosion and occasional internal calcification	

other studies report a strong correlation between histological grading of cartilaginous tumors and SUV max.^[16,17]

BENIGN CARTILAGINOUS TUMORS AND TUMOR-LIKE LESIONS

Chondroma

Chondroma is a benign, slowly growing cartilaginous tumor. In the head and neck, the most common location is the cricoid cartilage, followed by the thyroid cartilage. Approximately, 70–75% of laryngeal chondromas arise from the endolaryngeal surface of the posterior lamina of the cricoid cartilage.^[18] Less common locations are arytenoid cartilage, epiglottis, trachea, nose, ear pinna, and other soft tissues such as muscles of mastication.^[18-20] On CT, chondromas are seen as expansile well-circumscribed hypodense lesions with variable amounts of coarse, chondroid, mottled calcification, arising from a cartilaginous structure [Figure 3]. When they occur within submucosal regions, the overlying mucosa is altered in contour but is



Figure 2: Characteristic CT and MRI features of well-differentiated cartilaginous tumors. A 19-year-old man with sinonasal chondrosarcoma presenting with sinonasal obstructive symptoms. Axial maxillofacial CT image in bone window (a) shows a large right sinonasal mass containing internal ring-and-arc calcifications, a classic feature of calcified chondroid matrix (arrow). The tumor is heterogeneously hyperintense on T2W MRI image (b) and moderately enhancing on post-contrast T1W MRI image with fat suppression (c). Note the T2 hypointense area in the posterior aspect of the tumor (arrowhead in b) corresponding to the area of dense calcification on CT.



Figure 3: Laryngeal chondroma. A 51-year-old woman with laryngeal chondroma presenting with hoarseness for 6 months. Axial contrast-enhanced (a) and coronal-reformatted (b) CT images show a small expansile hypodense lesion at the left lamina of the thyroid cartilage (arrows). She underwent open resection of the lesion. Histopathology revealed a hypocellular tumor containing hyaline cartilage. The overlying perichondrium was intact with no evidence of destructive growth.

otherwise maintained. MRI appearance of chondromas is less specific because of its relative insensitivity in detecting chondroid calcifications. It can, however, provide additional information with regards to the extent of the lesion due to its high soft tissue contrast resolution.^[21] Chondromas and chondrosarcomas are difficult to distinguish on the basis of imaging, and as such histopathology is always required for confirming the diagnosis.^[21,22]

Chondroblastoma

Chondroblastomas are rare benign but locally aggressive bone tumors which commonly involve the epiphysis of the long bones such as distal femur, proximal tibia, and proximal humerus. Craniofacial chondroblastomas are uncommon, accounting for 2-7% of all chondroblastomas.[1,23,24] The most common location in the head and neck region is the skull base, with a predilection for the squamous portion of the temporal bone adjacent to the temporomandibular joint. The other less common sites include mandibular condyle [Figure 4], paranasal sinuses, and maxilla.^[25-28] Histopathological features of chondroblastomas include polygonal mononuclear cells (chondroblasts), multinucleated giant cells, and chondroid matrix. A fine network of pericellular calcification (so called "chicken-wire" pattern) can be seen in varying amounts. Clinical manifestations depend on tumor location, for example, with skull base chondroblastomas presenting as cranial neuropathy and chondroblastoma of the TMJ presenting with jaw pain, etc. Radiologically, chondroblastomas are seen as nonspecific well-defined expansile osseous lesions containing a variable amount of internal chondroid calcification. Invasion of the surrounding structures indicates the locally aggressive nature of these lesions.^[29,30] Intratumoral cystic degeneration or secondary aneurysmal bone cysts occur frequently in up to one third of these tumors.^[1,23] Although the tumors are locally aggressive, distant metastasis is rare. Surgical resection with complete tumor removal is the standard treatment.^[31]

Chondromyxoid fibroma

CMF account for less than 1% of all primary osseous tumors and predominantly affect the metaphysis of the long bones, particularly in the lower extremities. CMF of the skull and facial bones are exceedingly rare, with less than 100 cases reported in English literature.^[32] The previously reported locations of craniofacial CMF are skull base, sinonasal cavity, and, less frequently, the calvarium.^[31,32] These tumors demonstrate slow growth and tend to invade surrounding bones and soft tissue structures. Radiologic findings of CMF are nonspecific. On CT, CMFs are wellcircumscribed, expansile osseous lesions with a sclerotic



Figure 4: Chondroblastoma. A 36-year-old woman with chondroblastoma arising from the right mandibular condyle presenting with worsening right jaw pain. Axial T2WI MRI image (a), axial (b) and coronal (c) contrast-enhanced T1W MRI images with fat suppression show an exophytic enhancing T2 hyperintense mass arising from the right mandibular condyle projecting medially into the right masticator space (arrows). Note edema and enhancement of the right lateral pterygoid muscle (arrow heads).

rim. Intralesional calcification is an uncommon imaging feature and can be identified in approximately 10% of all CMF.^[33] MRI characteristics of CMF are similar to other cartilaginous tumors which show low signal on T1WI and heterogenous high signal on T2WI due to its cartilaginous and myxoid components, and heterogeneous enhancement on postcontrast sequence [Figure 5]. The nonspecific imaging appearance spurs a wide differential diagnosis which includes chondrosarcoma, chordoma, chondroblastoma, and chondroblastic osteosarcoma. Total tumor resection is the treatment of choice, with curative intention. In general, curettage is not recommended due to its higher recurrent rate (up to 25%). The role and efficacy of radiation therapy for local tumor control remains controversial.^[32]

Osteochondroma

Osteochondroma is a common benign bone lesion which usually occurs at the end of long bones such as distal femur.^[34] Osteochondroma in the head and neck region is relatively rare. The most common location is the mandible, particularly the mandibular condyle and coronoid process, but it can arise from any part of the mandible or other osseous structures in the head and neck.^[34-38] On imaging, head and neck osteochondromas are similar to osteochondromas seen



Figure 5: Chondromyxoid fibroma. A 56-year-old woman with chondromyxoid fibroma of the right central skull base presenting with multiple right cranial neuropathy. Axial T1WI (a), axial T2WI (b) and coronal contrast-enhanced T1W MRI image with fat suppression (c) show a large expansile mass arising from the right greater wing of sphenoid (arrows). The tumor demonstrates mixed signal intensity on both T1WI and T2WI with small lobular foci of T2 hyperintensity, and diffuse contrast enhancement.

elsewhere in the body and are seen as exophytic osseous lesions with continuity of bone marrow from the parent bone into the tumor [Figure 6]. A layer of cartilage known as the cartilaginous cap is seen at the growing end of the lesion. The cartilaginous cap is better seen on MR and typically has low signal on T1WI and high signal on T2WI.^[36] On postcontrast images, there is enhancement at the periphery of the lesion which correlates on histology with fibrovascular tissue covering the nonenhancing cartilaginous cap.^[39] The cartilaginous portion of osteochondroma may undergo malignant transformation. Abnormal thickening of the cartilaginous cap (exceeding 1.5–2.5 cm) on imaging is an ominous sign and is viewed with suspicion for malignant degeneration [Figure 7].^[40]

Synovial chondromatosis

Synovial chondromatosis is a benign but locally aggressive disease characterized by chondroid metaplasia of the synovium, with subsequent formation of intra-articular loose bodies, causing destruction of the joint.^[41,42] In the head and neck, it frequently involves the temporomandibular joint. Synovial chondromatosis can be divided into two subtypes – primary and secondary.^[41,42] Pathogenesis of primary form remains unknown, but according to recent cytogenetic



Figure 6: Osteochondroma. A 28-year-old woman with osteochondroma of the right mandibular condyle presenting with chronic right jaw pain. Axial (a) and coronal-reformatted (b) CT images in bone window show an exophytic osseous lesion originating from the right mandibular condyle, continuous with the medullary cavity of the mandible (arrow). Note remodeling of the skull base from chronic long-standing pressure effect (arrowhead).



Figure 7: Osteochondroma with secondary chondrosarcoma. A 70-year-old man with secondary chondrosarcoma originating from the osteochondroma of the cervical spine presenting with palpable lump in the neck. Axial T2W MRI image (a) shows an exophytic T2 hyperintense mass arising from the right ventral aspect of C4 vertebral body (arrow). Axial CT image in bone window (b) shows a few small calcific foci within the tumor (arrowhead). Note a small underlying osteochondroma deep to the tumor (black arrow).

studies, it is believed to be a benign neoplastic process rather than metaplastic disease.^[43] In contrast, secondary form is associated with underlying joint diseases such as mechanical joint abnormalities or arthritic conditions. Malignant transformation of primary synovial chondromatosis to chondrosarcoma may occur although it is uncommon.^[44]

Radiologic features of synovial chondromatosis include joint effusion, abnormal synovial thickening and enhancement suggestive of synovitis, adjacent bone erosion due to mechanical pressure, and calcified on noncalcified intra-articular loose bodies [Figure 8].^[41,42] In chronic disease, there may be imaging features of coexistent secondary osteoarthritis and asymmetric joint space narrowing. There is potential for intracranial tumor extension due to its close proximity to skull base. CT is the best imaging modality for detecting calcified intraarticular loose bodies and for assessment of bone erosion.



Figure 8: Synovial chondromatosis. A 45-year-old woman with synovial chondromatosis initially presenting with chronic nasal congestion. The left temporomandibular joint abnormality was incidentally found on maxillofacial CT. Coronal-reformatted CT image in bone window (a) and coronal contrast enhanced T1W MRI image with fat suppression (b) show abnormal synovial thickening and enhancement with extensive bone erosion of the left mandibular condyle and adjacent skull base (arrows). Note abnormal widening of the left temporomandibular joint.

MRI can provide additional information on soft tissue abnormalities, including synovitis, and bone marrow involvement. On MRI, the signal characteristics of intraarticular loose bodies are variable depending on the degree of mineralization. Noncalcified loose bodies typically have low-to-intermediate signal on T1WI and high signal on T2WI whereas calcified loose bodies show fat signal with a peripheral hypointense rim on T1WI and low signal on T2WI [Figure 9].^[41] During the early course of the disease, it can be difficult to diagnose synovial chondromatosis on MRI because loose bodies are not well formed, or they may be obscured by joint effusion.^[45]

Chondromesenchymal hamartoma

Chondromesenchymal hamartoma is a rare benign, slowgrowing, locally destructive, tumor-like lesion that contains mixed proliferating mesenchymal and cartilaginous elements such as mature and immature hyaline cartilage. This rare tumor-like lesion occurs predominantly in infants and young children. The most common sites are sinonasal cavity and orbit.^[46,47] The mass can invade skull base and extend intracranially.^[48] There is an association of pathologic DICER1 genetic variants, a hereditary cancer predisposition syndrome, with nasal chondromesenchymal hamartoma.^[49,50] As such, otolaryngologic evaluation should be performed in individuals with pathogenic DICER1 genetic variants who have persistent nasal obstruction.[49] Surgical resection is the treatment of choice.^[48] On imaging, chondromesenchymal hamartomas are seen as expansile heterogeneous mixed solid and cystic masses [Figure 10]. Approximately, 50% of nasal chondromesenchymal hamartomas contain internal calcifications. On MRI, tumors typically have low-to-intermediate signal on T1, heterogeneous hyperintense signal on T2, and marked heterogeneous contrast enhancement.[51,52]



Figure 9: Synovial chondromatosis with calcified intra-articular loose bodies. A 50-year-old woman with synovial chondromatosis presenting with left jaw pain. Axial CT image in bone window (a) shows several small calcified intra-articular loose bodies in the left temporomandibular joint (arrow) which are hypointense on axial T2W MRI image (b) (arrowhead). Note diffuse synovial thickening and distension of the joint capsule (black arrow).



Figure 10: Chondromesenchymal hamartoma. A 16-year-old man with *DICER1* mutation and sinonasal chondromesenchymal hamartoma presenting with chronic sinonasal obstructive symptoms. Axial (a) and coronal reformatted (b) CT images in bone window show polypoid opacification with scattered calcifications in bilateral ethmoid sinuses (arrows).

MALIGNANT CARTILAGINOUS TUMORS

Chondrosarcoma

Chondrosarcoma is the third most common primary malignant bone tumor after multiple myeloma and osteosarcoma.^[53] Head and neck chondrosarcomas accounts for 1–12% of all chondrosarcomas. They are usually low-grade (WHO Grade 1 and 2) with slow growth and indolent clinical course.^[54] Distant metastasis is uncommon in low-grade tumors but is more frequent in higher grade tumors (that is, WHO Grade 3). The most common location in the head and neck is the skull base, with approximately 60–70% of tumors emanating from the petroclival fissure.^[54] The other sites such as larynx and sinonasal cavity are less frequent.^[55,56] Chondrosarcomas usually occur in isolation but are occasionally associated with Ollier disease or Mafucci syndrome, rare diseases characterized by

multiple enchondromas.^[54,57] The standard treatment for chondrosarcoma is surgical resection. Adjuvant radiation is usually given when there is residual tumor following resection or close resection margins to improve local tumor control. Chondrosarcoma is generally considered to be unresponsive to chemotherapy.^[55]

Head and neck chondrosarcomas appear to be a distinct pathologic entity compared to chondrosarcomas arising elsewhere in the body.^[55] A meta-analysis study has shown that head and neck chondrosarcomas tend to be lower grade and have significantly higher 10-year disease-specific survival and overall survival compared to chondrosarcomas at other sites.^[55] A recent study also found that the incidence of IDH mutation is different among the head and neck chondrosarcomas of different subsites.[58] Approximately, 86% of skull base chondrosarcomas have an IDH mutation while the incidence of IDH mutation in laryngotracheal chondrosarcomas is only 12%. More interestingly, no IDH mutations are found in maxillofacial chondrosarcomas. The authors hypothesized that this could be related to different types of ossification (that is, enchondral ossification in the skull base compared to intramembranous ossification in the maxillofacial bones) and different pathway of tumorigenesis in each location.[58]

Radiologically, chondrosarcomas are expansile soft tissue masses with bone erosion and invasion of adjacent structures. Up to 50% of skull base chondrosarcomas show intratumoral calcification on CT.^[59] Intratumoral chondroid calcification (for example, ring-and-arc or popcorn appearance) is one of the most helpful imaging features suggesting cartilaginous origin of the tumor and best depicted on CT. MRI can provide more information regarding bone marrow and soft tissue involvement. MR signal characteristics of chondrosarcoma are low-to-intermediate signal on T1 and hyperintense signal on T2, with a variable degree of contrast enhancement [Figure 11].^[60] Low-grade chondrosarcomas may contain internal T2 hypointense septations and septal enhancement which corresponds to fibrovascular tissue on histopathology while the nonenhancing areas within tumors represent the areas of hyaline cartilage, cystic mucoid tissue, and tumor necrosis.^[39]

The main differential diagnosis of skull base chondrosarcoma is chordoma; it can be difficult to differentiate the two due to overlapping imaging appearance. The tumor location is a helpful imaging clue that may help in differentiation between these two entities. Skull base chondrosarcomas are more frequently located off-midline near the petrooccipital fissure, whereas chordomas are more often located in the midline of the clivus.^[61] DWI MRI with quantitative assessment of apparent diffusion coefficient (ADC) values may be helpful in distinction, with some studies demonstrating skull base chondrosarcomas have



Figure 11: Skull base chondrosarcoma. A 31-year-old man with skull base chondrosarcoma presenting with progressive left cranial neuropathy. Axial T2W MRI image (a) and coronal contrast-enhanced T1W MRI image with fat suppression (b) show a large expansile heterogeneously enhancing T2 hyperintense extraaxial mass centered at the left petroclival fissure (arrows) extending into the preportine cistern and causing mild mass effect on the adjacent brainstem and left temporal lobe. The tumor contains subtle small calcifications (arrowhead), a clue to the diagnosis, best seen on CT (c).

higher ADC values compared to chordomas, though further research is required.^[61-63]

In the larynx, chondrosarcoma is the most common nonepithelial tumor, and it is much more common than chondroma.^[64] The most frequent sites of tumor are cricoid cartilage followed by thyroid cartilage. CT characteristics of laryngeal chondrosarcoma is an endolaryngeal soft tissue mass with or without extralaryngeal extension.[65] Approximately, 70-80% of laryngeal chondrosarcomas show matrix calcification visible on CT [Figure 12].^[56] The main differential diagnosis of calcified laryngeal mass is chondrosarcoma and chondroma; however, it is difficult to distinguish them based on their radiologic features. There is growing body of evidence that chondrosarcoma and chondroma are closely related in both histopathology and genetic molecular profiles. Some lesions may contain both components which pose challenges for tissue diagnosis after biopsy caused by under sampling.^[64] Differential diagnosis is much broader and include both epithelial and nonepithelial tumors if the mass is not calcified.^[65]

Chondroid chordoma

Chondroid chordoma is a distinct variant of chordoma which contains hyaline cartilage. It accounts for 14% of all skull base chordomas.^[33,66] The most common location of



Figure 12: Laryngeal chondrosarcoma. A 59-year-old man with laryngeal chondrosarcoma presenting with progressive hoarseness of voice. Axial (a) and coronal-reformatted (b) contrast-enhanced CT images show an expansile partially calcified mass arising from the left lateral aspect of the cricoid cartilage (arrows), severely narrowing the airway. Note characteristic chondroid matrix calcification within the tumor.



Figure 13: Chondroid chordoma. A 36-year-old male with chondroid chordoma. The tumor was incidentally found on head CT which was performed as part of work up for patient's headache (not shown). Axial T2W MRI image (a) and sagittal contrast-enhanced T1W MRI image with fat suppression (b) show a lobulated extraaxial T2 hyperintense, mildly enhancing mass centered at the superior aspect of the clivus (arrows) causing mild mass effect on the adjacent brainstem.

chondroid chordomas is the skull base, but it occasionally occurs in other locations such as the nasal septum.^[66,67] On histopathology, chondroid chordomas may mimic other cartilaginous tumors such as chondrosarcoma especially in a limited biopsy sample, and immunohistochemical tests are usually required for diagnosis.^[68] It is crucial to differentiate between chondroid chordoma and chondrosarcoma because they have different clinical outcomes and prognosis.^[69,70] It has been shown in a previous study^[69] that chondrosarcomas have significantly longer overall and disease-free survivals when compared to chondroid chordomas and conventional chordomas. Radiologic features of chondroid chordoma are similar to those of conventional chordoma and include an expansile soft tissue mass with adjacent bone erosion and occasional internal calcification [Figure 13].^[66,71]

CONCLUSION

The important imaging clues in diagnosis of the head and neck cartilaginous tumors include characteristic location, internal calcified chondroid matrix, and typical T2 hyperintense signal due to high water content in hyaline cartilage.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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