



Discover Generics

Cost-Effective CT & MRI Contrast Agents



WATCH VIDEO

AJNR

Craniofacial chondrosarcomas: imaging findings in 15 untreated cases.

Y Y Lee and P Van Tassel

AJNR Am J Neuroradiol 1989, 10 (1) 165-170

<http://www.ajnr.org/content/10/1/165>

This information is current as of June 19, 2025.

Craniofacial Chondrosarcomas: Imaging Findings in 15 Untreated Cases

Ya-Yen Lee¹
Pamela Van Tassel

Radiographic findings of 15 untreated chondrosarcomas of the cranial and facial bones were reviewed. These tumors have a propensity to occur in the wall of a maxillary sinus, at the junction of sphenoid and ethmoid sinuses and vomer, and at the undersurface of the sphenoid bone. Because of its slow-growing nature, chondrosarcomas tend to be large, multilobulated, and sharply demarcated when detected. Frequent bone changes are a combination of erosion and destruction, with sharp transitional zones and absent periosteal reaction. Tumor matrix calcifications, not necessarily chondroid, are almost always present. Both CT and MR may be necessary for thorough evaluation of tumor extent.

Chondrosarcoma, a malignant but usually slow-growing cartilaginous tumor, constitutes approximately 11% of malignant bone tumors [1] but rarely occurs in the craniofacial region. Because of its propensity to occur in the deep facial structures or base of the skull, the true extent and origin of the tumor may be overlooked if not properly evaluated radiographically. We review a relatively large series of craniofacial chondrosarcomas and discuss the differential diagnosis and choice of imaging technique.

Materials and Methods

This retrospective radiologic review was based on the pretreatment radiographic studies of 15 patients with craniofacial chondrosarcomas seen at our institution over a period of 40 years, excluding three intracranial dural chondrosarcomas, which are to be reported separately. An attempt was also made to correlate the radiographic findings with the histologic grades of the tumors.

The ages of the patients ranged from 10 to 73 years, with a mean of 40 years. There were 12 females and 3 males. Two of the chondrosarcomas were secondary tumors, one associated with extensive fibrous dysplasia and the other developing 25 years after irradiation. There were no other associated systemic bone disorders, such as osteochondromatosis or enchondromatosis. No case was found to have regional nodal or systemic metastases at presentation. Histologically, there were 11 classic (six grade-I and five grade-II), three mesenchymal, and one dedifferentiated chondrosarcomas.

Radiologic studies available for review included plain films, conventional tomograms, arteriograms, and MR images. Plain radiographs of the paranasal sinuses or skull, routine studies in earlier years, were obtained in 14 patients. Conventional tomograms, routinely obtained before the advent of CT, were available in 12 patients. Nine patients had CT in the axial plane, some with optional coronal images, with or without IV administration of contrast medium. Arteriograms were acquired selectively in five patients for presurgical diagnosis. More recently, one patient has had an MR study, which was obtained on a 1.5-T superconducting scanner. Radiologic analysis was made in terms of the following parameters: location; configuration and size; associated bone changes (erosion, destruction, type of transitional zone, and periosteal reaction); tumor matrix mineralization; and vascularity.

Received December 31, 1987; accepted after revision April 25, 1988.

¹ Both authors: Division of Diagnostic Imaging, Department of Diagnostic Radiology, M. D. Anderson Hospital and Tumor Institute at Houston, The University of Texas System Cancer Center, 1515 Holcombe Blvd., Houston, TX 77030. Address reprint requests to Y-Y Lee.

AJNR 10:165-170, January/February 1989
0195-6108/89/1001-0165

© American Society of Neuroradiology

Results

Table 1 presents a summary of the following findings.

1. *Location.* Twelve patients had tumors in the paranasal sinuses and nasal cavity, and three patients had tumors at the undersurface of the skull base. Further analysis of the 12 paranasal sinus and nasal cavity tumors showed that six originated in the maxillary sinuses (Fig. 1) and six had a geometric center at the junction of the sphenoid sinuses and vomer (Fig. 2). Among the six maxillary tumors, four were primary lesions and originated from the antral walls without involvement of the alveolar ridge of the maxilla (Fig. 3); the other two were secondary tumors. One of these occurred in a case of extensive polyostotic fibrous dysplasia, with tumor involving the entire maxillary sinus (Fig. 4); the other one, in the alveolar ridge, developed 25 years after radiation therapy (5500 rad) to an unknown type of maxillary tumor (Fig. 5). All three tumors arising from the undersurface of the skull base occurred at the greater sphenoid wing without intracranial extension (Fig. 6).

2. *Configuration and size.* All the lesions were sharply demarcated and multilobulated (Fig. 2). They tended to be large, ranging from $3 \times 2 \times 3$ cm to $8 \times 6 \times 7$ cm.

3. *Bone changes.* All tumors were associated with combined erosive and destructive bone changes (Figs. 1–3) except for one dedifferentiated chondrosarcoma in which destruction was the predominating feature (Fig. 4). The transitional zone in all cases was sharp except for the above-mentioned dedifferentiated tumor, which arose from preexisting extensive fibrous dysplasia, and a mesenchymal chondrosarcoma of the maxillary sinus. No periosteal reaction was identified in any case.

4. *Tumor matrix mineralization.* All tumors had matrix calcification, which varied greatly from the classical ring, arc, or snowflake chondroid calcifications to nonspecific amorphous

calcifications. There was no close correlation between the amount or character of tumor classification and histologic grade, although the extensive calcification and classic chondroid calcification tended to occur in the lower-grade classic chondrosarcoma. Peripheral eggshell calcifications were also observed (Figs. 3 and 6).

5. *Tumor vascularity.* Arteriograms were obtained in five patients and showed avascular tumors except for one mesenchymal maxillary tumor, which was hypovascular.

Discussion

Chondrosarcomas rarely occur in the head and neck region; they constitute just 6.7% of all reported chondrosarcomas [2]. Although the laryngeal cartilage is well known as a site of occurrence of chondrosarcoma, the rate at which this tumor occurs in the craniofacial region is actually higher. Although several theories have been put forth to explain the origin of chondrosarcomas, their exact etiology remains unknown [3]. Chondrosarcomas can arise from the cartilage, cartilaginous bone, or tissue not normally harboring cartilage. Multidirectional differentiation of primitive mesenchymal cells is the most accepted explanation for the occurrence of chondrosarcomas in such structures as the dura, membranous bones, and soft tissues. In addition, chondrosarcomas can originate from the cartilaginous tissue of an existing exostosis, enchondroma, or osteochondroma. Malignant transformation may occur after irradiation or in severely disordered bone, as seen in fibrous dysplasia.

Histologically, chondrosarcomas have been classified into three subtypes: classical, mesenchymal, and dedifferentiated. The classical chondrosarcomas can be further subdivided into grades I, II, and III on the basis of their mitotic rate, cellularity, and nuclear size [4]. The rarer mesenchymal and dedifferentiated chondrosarcomas are more malignant [5, 6]. Eleven (73.3%) of the 15 chondrosarcomas were of the classical subtype (six grade I, five grade II, and no grade III); three (20%) were mesenchymal; and one (6.6%) was dedifferentiated.

The geometric center of the maxillary chondrosarcomas in our series appears to be in the antral wall, unlike the origin of primary osteosarcoma, which tends to arise from the alveolar ridge [7]. However, the number of our cases is too small to make this finding conclusive. Another group of chondrosarcomas appears to originate at the junction of the sphenoid and ethmoid sinuses and the vomer of the nasal cavity, along their articulations. This observation is not mentioned in the literature. The extent of chondrosarcoma in the paranasal sinus and nasal cavity is frequently misdiagnosed because of poor accessibility to the tumor on clinical examination and inadequate radiologic examination. Such a tumor can easily be misdiagnosed as a nasopharyngeal tumor because of its posteroinferior extension when the anterior extent is not appreciated on the nasal examination. For the same reason, a maxillary tumor might be misdiagnosed as a nasal cavity tumor by its medial extension.

The chondrosarcomas of the undersurface of the sphenoid bone, in the region of the pterygoid plate, had growth into the



Fig. 1.—Axial CT of maxillary sinus chondrosarcoma. Note bone erosion (arrowhead) and sharply marginated destruction (small arrows). Extension into right infratemporal fossa is appreciated but poorly defined, with a large amorphous calcification (large arrow).

TABLE 1: Craniofacial Chondrosarcoma

Case No.	Age Gender	Original Site	Configuration and Size (cm)	Bone Changes			Histologic Grading	Remarks
				Bone Erosion/ Destruction	Transitional Zone	Periosteal Reaction		
A. Paranasal sinuses and nasal cavity:								
1.	24 M	Medial maxillary wall	Multilobulated 5 x 4 x 5	++/+	Sharp and long	-	Mesenchymal	
2.	48 F	Medial maxillary wall	Multilobulated 3 x 4 x 3	+/+	Sharp	-	Classic, grade I	
3.	70 F	Anterior maxillary wall	Multilobulated 3 x 2 x 3	+/+	Sharp	-	Classic, grade II	
4.	24 F	Posterior maxillary wall	Multilobulated 5 x 4 x 5	+/+	Sharp	-	Mesenchymal	
5.	73 F	Alveolar ridge	Multilobulated 5 x 5 x 4	++/++	Long	-	Classic, grade I	25 years after X-ray therapy
6.	58 M	Entire maxillary sinus	Multilobulated 8 x 6 x 7	+/+	Sharp	-	Dedifferentiated	Associated extensive fibrous dysplasia
7.	40 F	Junction of sphenoidal and vomer	Multilobulated 4 x 3 x 5	+/+	Sharp	-	Classic, grade II	
8.	54 F	Junction of sphenoidal sinuses and vomer	Multilobulated 3 x 3 x 3.5	+/+	Sharp	-	Classic, grade II	
9.	15 F	Junction of sphenopalatine and vomer	Multilobulated 7 x 6 x 6	++/++	Sharp	-	Classic grade II	
10.	54 F	Junction of sphenoidal sinuses and vomer	Multilobulated 4.5 x 3 x 4	+/+	Sharp	-	Classic, grade II	
11.	12 F	Junction of sphenoidal sinuses and vomer	Multilobulated 6 x 5 x 5	++/+	Sharp	-	Classic, grade I	
12.	18 F	Junction of sphenoidal sinuses and vomer	Multilobulated 6 x 4 x 5	+/+	Sharp	-	Classic, grade I	
B. Undersurface of skull base:								
13.	60 F	Pterygoid palate	Multilobulated 3 x 3 x 4	+/+	Sharp	-	Mesenchymal	
14.	10 M	Pterygoid palate	Multilobulated 3 x 3 x 6	+/+	Sharp	-	Classic, grade I	
15.	71 F	Undersurface of sphenoid wing, lateral to pterygoid palate	Multilobulated 4 x 5 x 5	+/+	Sharp	-	Classic, grade I	

Note.— + = mild; ++ = moderate; +++ = marked.

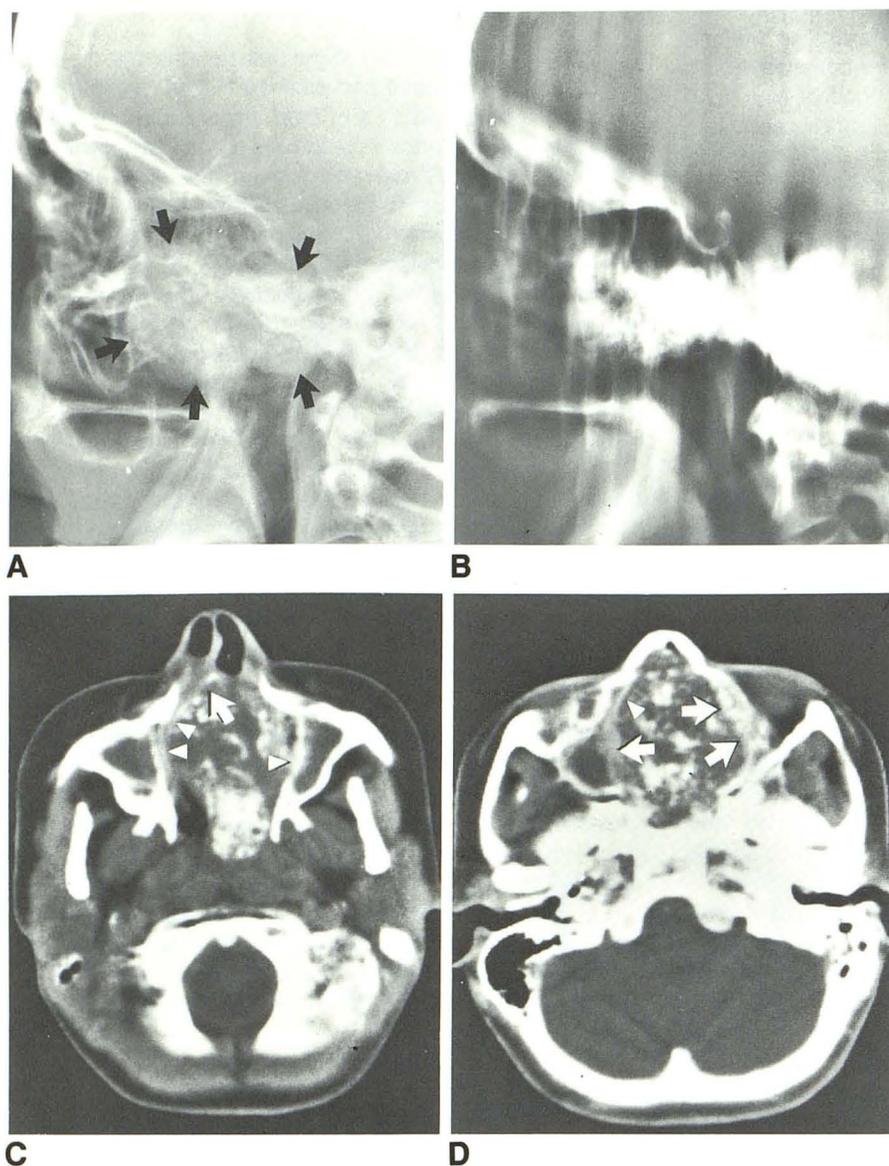


Fig. 2.—Two chondrosarcomas of paranasal sinuses and nasal cavity.

A and B, Plain film (A) and tomogram (B) show that tumor (arrows) is sharply demarcated and multilobulated, with extensive chondroid calcification.

C and D, Axial CT scans of a different patient show bone erosion (arrowheads) and destruction (arrows) in addition to matrix calcification. Geometric center in both cases appears to be at junction of sphenoid and ethmoid sinuses and vomer, although this is less well appreciated on axial CT.

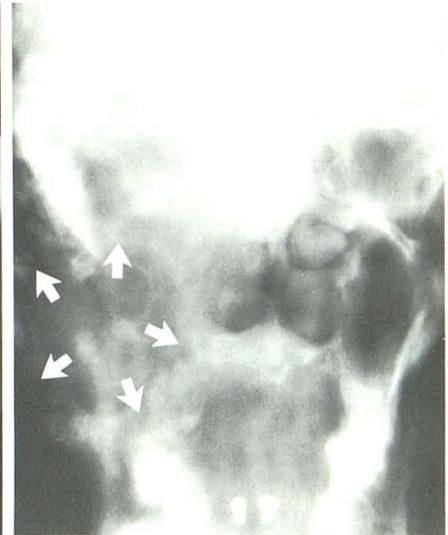
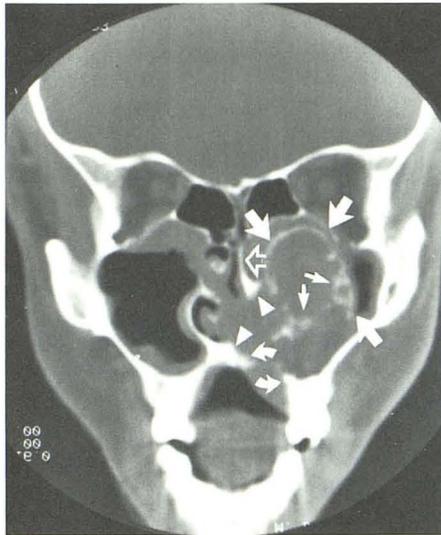
infratemporal fossa or parapharyngeal space. None of the cases studied had tumor extending intracranially, although this may occur [8, 9]. There may be confusion between chondrosarcoma and chondroblastic osteosarcoma of the mandible, as reported by Dahlin and Unni [10]; in their series, some chondroblastic osteosarcomas of the jaw were called chondrosarcomas by other observers. There was no mandibular chondrosarcoma in our series; and, almost exclusively, the malignant primary mandibular bone tumors seen at our institution have been osteosarcomas (Ayala AG, personal communication). However, this disagreement is only of academic interest, since mandibular osteosarcomas and chondrosarcomas affect patients in the same age range and have practically identical 5-year survival rates.

Owing to its slow-growing nature and symptom-insensitive location, craniofacial chondrosarcoma tends to be very large and multilobulated when detected clinically. The dominant

bone change is a combination of erosion and destruction. Often, the transitional zone is sharp and there is no periosteal reaction. Matrix calcification is almost always observed, but it is poorly correlated with the degree of tumor malignancy, although extensive chondroid calcification tends to occur in low-grade classical chondrosarcomas. Occasionally, the presence of sequestered bone fragments in the more frequently occurring squamous cell carcinoma may be confusing. As expected, most chondrosarcomas are avascular as compared with osteosarcomas, which are hypervascular [11].

The differential diagnosis of craniofacial chondrosarcoma should include osteosarcoma, chondroma, osteochondroma, or aggressive osteoblastoma. Osteosarcoma, which is more frequent and malignant, has a propensity to arise from the maxillary alveolar ridge or the mandible, demonstrates more destructive bone changes, and appears hypervascular [7]. However, on some occasions, chondroblastic osteosarcoma

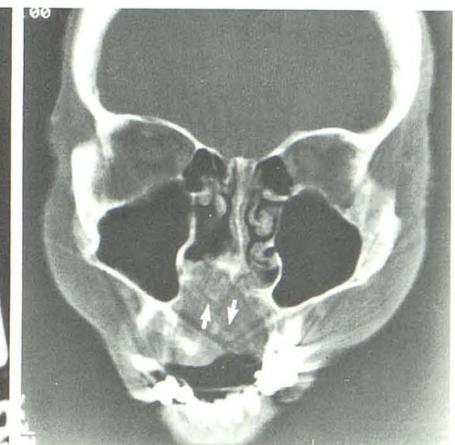
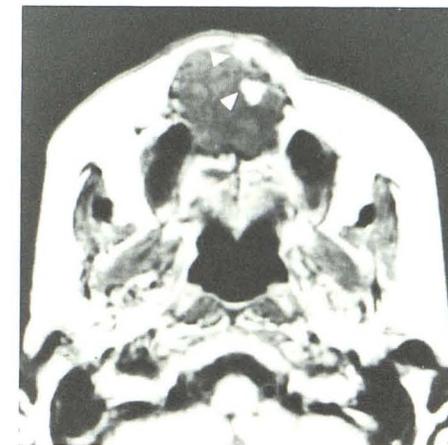
Fig. 3.—Coronal CT of maxillary sinus mesenchymal chondrosarcoma. Note amorphous (*small arrows*) and eggshell (*large arrows*) calcifications. Tumor seems to originate from medial wall of antrum. Alveolar ridge is free of involvement. Bone destructions have sharp (*arrowheads*) and long (*curved arrows*) transitional zones. Bone erosion (*open arrow*) is also noted. Tumor margins could not be demarcated from adjacent thickened mucosa.



3

4

Fig. 4.—Tomogram of maxillary sinus differentiated chondrosarcoma. Note extensive fibrous dysplasia of craniofacial bones. Destruction with long transitional zones (*arrows*) is the predominating feature.



A

B

Fig. 5.—Radiation-induced alveolar ridge chondrosarcoma.

A and B, Axial T1-weighted MR, 600/25 (A) and coronal CT (B). There is sharp demarcation of tumor margins on MR, which also shows post-biopsy hemorrhages (*arrowheads* in A); however, the matrix calcifications (*arrows* in B) are better appreciated on CT.



A

B

Fig. 6.—Pterygoid plate chondrosarcomas.
A, Tomogram shows fairly well defined peripheral eggshell calcification with central amorphous calcification in a classic chondrosarcoma.
B, Coronal CT of poorly defined irregular eggshell calcification in a mesenchymal chondrosarcoma.

might be very difficult to differentiate radiographically as well as histologically. The incomplete eggshell calcification and associated bone erosion may make chondrosarcoma indistinguishable from osteochondroma, chondroma, or aggressive osteoblastoma. One clear definition of osteochondroma is demonstration of continuation of the cortex and medullary cavity of the parent bone into the tumor [12]. Chondrosarcoma might be very difficult to distinguish from aggressive osteoblastoma [13] and chondroma [14] because of their matrix mineralizations, which are usually more organized; however, a soft-tissue component of tumor would tend to rule out the latter two. Skull-base chondrosarcoma with intracranial extension should be differentiated from chondroma and meningioma. Chondroma tends to originate from the midline and extend laterally, usually causing extensive bone destruction and not erosion [15]. Meningioma is frequently associated with reactive hyperostosis and intense homogeneous contrast enhancement on CT.

Since radical surgery remains the treatment of choice for chondrosarcoma, demonstration of tumor extent is essential. In recent years CT has replaced conventional tomography for radiologic evaluation of tumor extent. However, the inability to image in the sagittal plane and poor demarcation of tumor from inflammatory changes in the sinuses make CT less than optimal. These shortcomings of CT can be overcome by MR [16]; however, MR will not provide an adequate study of bone structures and matrix calcifications. Therefore, at the present time, both CT and MR are probably required for the complete study of craniofacial chondrosarcomas. Arteriography remains valuable for differentiation of chondrosarcoma from osteosarcoma when histologic diagnosis is uncertain.

REFERENCES

1. Dahlin DC, Unni KK. *Bone tumors*, 4th ed. Springfield, IL: Thomas, 1986:227-244
2. Pritchard DJ, Lunke RJ, Taylor WF, Dahlin DC, Medley BE. Chondrosarcoma: a clinicopathologic and statistical analysis. *Cancer* 1980;45:149-157
3. Jones HM. Cartilaginous tumors of the head and neck. *J Laryngol Otol* 1973;87:135-151
4. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone. A clinicopathologic analysis with emphasis on histologic grading. *Cancer* 1977;40:813-831
5. Dahlin DC, Unni KK. *Bone tumors*, 4th ed. Springfield, IL: Thomas, 1986:261-268
6. Dahlin DC, Unni KK. *Bone tumors*, 4th ed. Springfield, IL: Thomas, 1986:245-253
7. Lee YY, Van Tassel P, Nauret C, Raymond K, Edeiken J. Craniofacial osteosarcomas. Plain film, CT, and MR findings in 46 cases. *AJNR* 1988;9:379-385
8. Grossman RI, Davis KR. Cranial computed tomographic appearance of chondrosarcoma of the base of the skull. *Radiology* 1981;141:403-408
9. Bahr AL, Gayler BW. Cranial chondrosarcoma. *Radiology* 1977;124:151-156
10. Dahlin DC, Unni KK. *Bone tumors*, 4th ed. Springfield, IL: Thomas, 1986:286
11. Yaghma I. Angiographic features of chondroma and chondrosarcomas. *Skeletal Radiol* 1987;3:91-98
12. Kenney PJ, Gilula LA, Murray WA. The use of computed tomography to distinguish osteosarcoma and chondrosarcoma. *Radiology* 1981;139:129-137
13. Som PM, Bellot P, Blitzer A, Som ML, Geller SA. Osteoblastoma of the ethmoid sinus. *Arch Otolaryngol* 1979;105:623-625
14. Krayenbuhl H, Yasargil MG. Chondromas. *Progr Neurol Surg* 1975;6:435-463
15. Meyer JE, Oot RF, Lindfors KK. CT appearance of clival chordomas. *J Comput Assist Tomogr* 1986;10:34-38
16. Dillon WP. Magnetic resonance imaging of head and neck tumors. *Cardiovasc Intervent Radiol* 1986;8:275-282