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Craniofacial Osteosarcomas: Plain Film, CT, and MR Findings in 46 Cases

Ya-Yen Lee[1]
Pamela Van Tassel[1]
Craig Nauert[1]
A. Kevin Raymond[2]
Jack Edeiken[1]

Forty-six osteosarcomas of the cranial and facial bones were reviewed radiographically by using the conventional parameters for long bone tumors. There were 32 de novo osteosarcomas (11 maxillary, 13 mandibular, and eight cranial) and 14 postradiation osteosarcomas. All the maxillary tumors originated from the alveolar ridge, and the majority of mandibular lesions began in the body of the mandible. The postradiation osteosarcomas occurred in portions of bones at the borders of the radiation field; the latent period ranged from 4 years, 2 months to 50 years (mean, 14 years). The majority of de novo or postradiation craniofacial osteosarcomas were osteolytic with a long transition zone and no periosteal reaction; the exception was in the mandible, where nearly half the cases were osteoblastic and periosteal reaction was occasionally present. Tumor matrix mineralization occurred in more than 75% of the cases, and osteoid matrix calcification was most frequent, even though most tumors were chondroblastic. Soft-tissue extension of tumor was present in all cases and contained calcifications in more than half.

Conventional radiographs are of limited value in evaluating head and neck osteosarcomas because of the superimposed bony structures. CT provides excellent detection of tumor calcification, cortical involvement, and, in most instances, soft-tissue and intramedullary extension. MR is even more effective in demonstrating the intramedullary and extraosseous tumor components on both T1- and T2-weighted images. However, CT and plain films are superior to MR in detecting the matrix calcifications and bone destruction or reaction.

With the exception of multiple myelomas, osteosarcoma is the most frequently occurring primary malignant bone tumor. It generally affects older children or young adults, and the most common sites are the distal femur and proximal tibia. Radiographically, the tumors of the long bones have been evaluated by using the parameters of bone destruction, transition zone, tumor matrix mineralization, and periosteal reaction [1–6]. The cranial and facial tumors, being infrequent, have not been extensively evaluated. Our large patient referral population in a cancer hospital setting has allowed us to collect pretreatment radiographic studies of cranial and facial tumors in 46 patients; we have analyzed these tumors by using the above parameters and compared them with tumors found in the long bones.

Materials and Methods

Of 79 patients with craniofacial osteosarcomas registered at our institution from March 1945 to April 1987, 46 had pretreatment radiographic studies available for review. Of the 32 osteosarcomas de novo, 11 were in the maxilla, 13 in the mandible, and eight in the cranial bones. The remaining 14 patients had postradiation osteosarcomas. No case was found in preexisting bone disorders such as Paget disease or fibrous dysplasia.

The pretreatment radiologic studies available for review consisted of 46 plain films, 28 conventional tomograms, and 29 CT scans. In addition, four patients had MR imaging. The lesions were analyzed by using the conventional parameters for osteosarcoma in the long bones: (1) bone destruction, (2) transition zone, (3) tumor matrix mineralization, (4) periosteal

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[1] 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.

[2] Division of Pathology, Department of Anatomical Pathology, M. D. Anderson Hospital and Tumor Institute at Houston, The University of Texas System Cancer Center, Houston, TX 77030.

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reaction, and (5) soft-tissue extension and calcification. The patient's gender, age at initial presentation, presenting symptom(s), and latent period in postradiation cases were recorded.

Surgical specimens were reviewed and subclassified histologically into (1) osteoblastic, (2) chondroblastic, or (3) fibroblastic subtypes [7].

Results

The following data are summarized in Table 1.

Osteosarcomas de novo (n = 32)

Facial Bones (n = 24)

(1) Maxilla (*n* = 11).—Seven males and four females, ages 13 to 52 years (mean, 34.7 years) were in this category. Painless local swelling was the presenting symptom in eight patients (73%). Cervical lymphadenopathy did not occur. The epicenter of the tumor was in the alveolar ridge in all 11. Six (55%) of these tumors were osteolytic, with two osteoblastic

TABLE 1: Craniofacial Osteosarcomas (*n* = 46)

	Average Age (yrs)	Gender Ratio (M/F)	Painful Swelling (no.)	Location (no.)	Bone Destruction (no.)	Transitional Zone (no.)	Mineralized Tumor Matrix (no.)	Periosteal Reaction (no.)	Soft-Tissue Extension (no.)	Soft-Tissue Calcification (no.)	Histologic Subtype (no.)
Osteosarcoma de novo (*n* = 32)											
Facial bones (*n* = 24)											
1. Maxilla (*n* = 11)	34.7	7:4	(3)	Alveolar ridge (11)	Lytic (6) Blastic (2) Mixed (2) None (1)[a]	Long (10) None (1)[a]	Osteoid (9)[b] None (2)	(None)	(11)	(6)	Osteoblastic (1) Chondroblastic (9) Fibroblastic (1)
2. Mandible (*n* = 13)	35.5	5:8	(11)	Body (10) Symphysis (2) Angle (1)	Lytic (4) Blastic (6) Mixed (3)	Long (9) Short (4)	Osteoid (8) Chondroid (5)	Spiculated (1) Laminated (2)	(13)	(8)	Osteoblastic (2) Chondroblastic (10) Fibroblastic (0) Unknown (1)
Cranial bones (*n* = 8)	34.5	4:4	(1)	Parietal (2) Temporal (2) Occipital (1) Ethmoid (1) Sphenoid (2)	Lytic (7) None (1)[c]	Long (7) None (1)[c]	Osteoid (4)[d] None (4)	(None)	(8)	(4)	Osteoblastic (4) Chondroblastic (0) Fibroblastic (3) Unknown (1)
Postradiation osteosarcoma;[e] (*n* = 14)	38.2	8:6	(10)	Periphery of radiation field (14)	Lytic (9) Blastic (3) Mixed (2)	Long (14)	Osteoid (7) Chondroid (2) None (5)	(None)	(14)	(7)	Osteoblastic (1) Chondroblastic (5) Fibroblastic (2) Unknown (6)

[a] Parosteal osteosarcoma.
[b] One with additional chondroid calcification.
[c] Parosteal osteosarcoma.
[d] One with additional chondroid calcification.
[e] Latent period: 4–50 yr; mean, 14 yr.

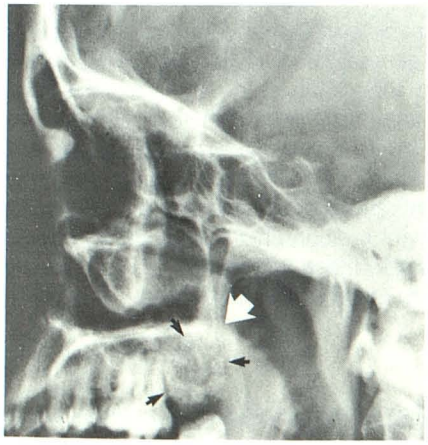


Fig. 1.—Lateral plain film of maxillary chondroblastic osteosarcoma in 39-year-old man. Subtle osteoblastic changes of posterior alveolar ridge (*white arrow*) of maxilla and clustered chondroid tumor calcification (*black arrows*).

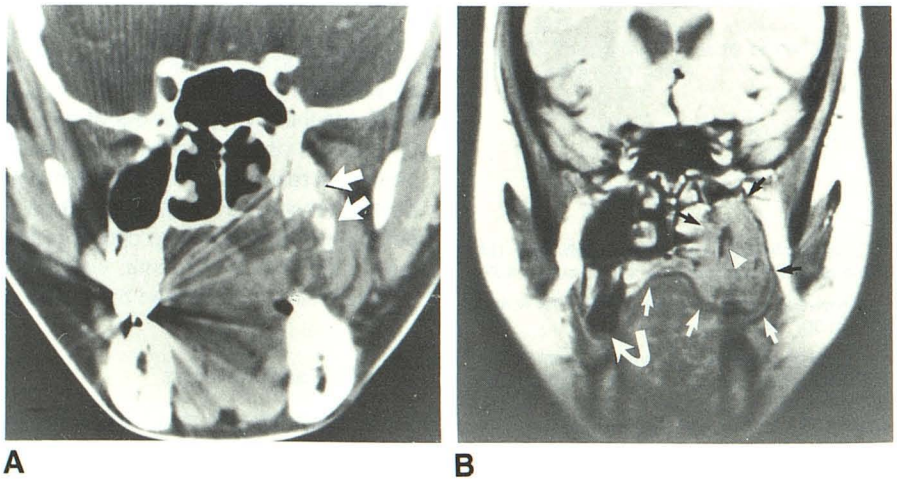


Fig. 2.—Left maxillary chondroblastic osteosarcoma in 42-year-old woman.

A, CT scan shows a large osteolytic destructive mass of left posterior alveolar ridge extending into hard palate, maxillary antrum, nasal cavity, and infratemporal fossa, with osteoid calcification (*arrows*).

B, MR image in T1-weighted pulse sequence shows sharp demarcation of tumor extent (*straight arrows*). Noted is artifact caused by tooth fillings (*curved arrow*). Signal void within mass denotes matrix calcification (*arrowhead*).

(18%) and two mixed lesions (18%) (Figs. 1 and 2). All showed an aggressive-looking long transition zone. The remaining case was a parosteal osteoblastic osteosarcoma with no underlying bony changes (Fig. 3). Osteoid tumor matrix mineralization was identified in nine patients (81%) with one of these showing additional chondroid calcification. Soft-tissue extension was present in all 11, with calcifications identified in six (55%). Periosteal reaction did not occur. Histologically, nine of the lesions were chondroblastic, one osteoblastic, and one fibroblastic.

(2) Mandible (n = 13).—Five males and eight females, ages 7 to 62 years (mean, 35.5 years) were in this category. All presented with local swelling, which was painful in 11 cases. There was no cervical lymphadenopathy. Tumors arose in the body of the mandible in 10 (77%) of the 13 patients. Four (31%) were osteolytic, six (46%) were osteoblastic, and three (23%) were mixed (Figs. 4 and 5). Osteoid calcification was the most frequent form of tumor matrix mineralization, occurring in eight (62%) of the 13 patients, with the remaining five showing chondroid calcification. The transition zone was long in nine cases (69%), while in four cases (31%) this zone was short and the borders were sharply marginated. Periosteal reaction occurred in three patients (23%); it was laminated in two cases (Fig. 6) and spiculated in the third (Fig. 7). Soft-tissue extension occurred in all 13 cases and contained calcification in eight (62%). Histologically, there were 10 chon-

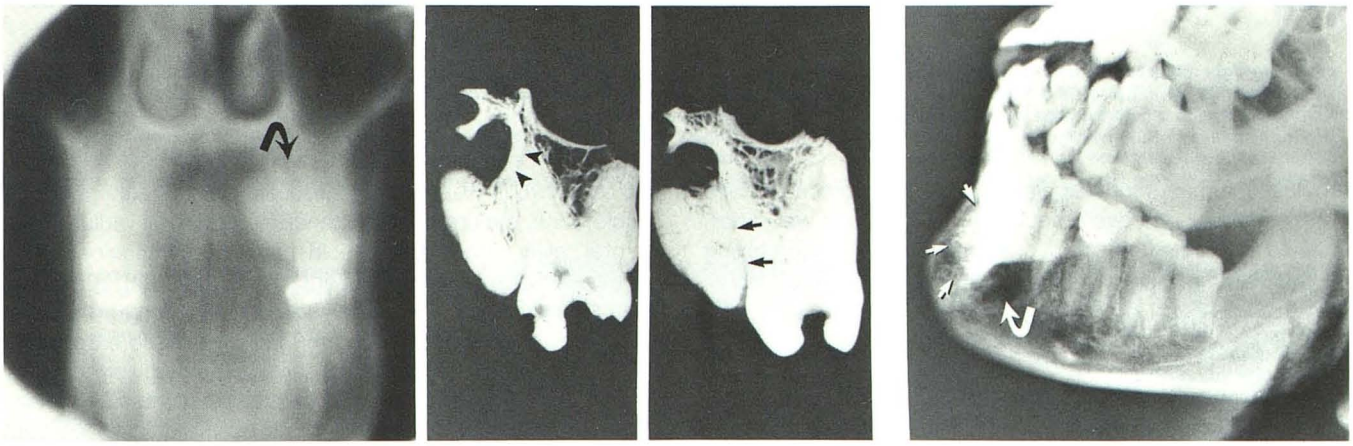


Fig. 3.—Maxillary parosteal osteosarcoma in 25-year-old man.
A, Anteroposterior conventional tomogram shows dense homogeneous osteoblastic mass arising from medial aspect of left alveolar ridge, with a faint radiolucent cleft in between (*curved arrow*).
B, Anteroposterior radiograph of surgical specimen. Noted are the base (*arrowheads*) of this mushroomlike parosteal tumor and the cleft between tumor and tooth (*arrows*).

Fig. 4.—Oblique plain film shows chondroblastic osteosarcoma arising from symphysis of mandible in 19-year-old boy. Characteristic chondroid mineralization in medullary cavity (*arrows*). Radiolucent defect denotes biopsy site (*curved arrow*).

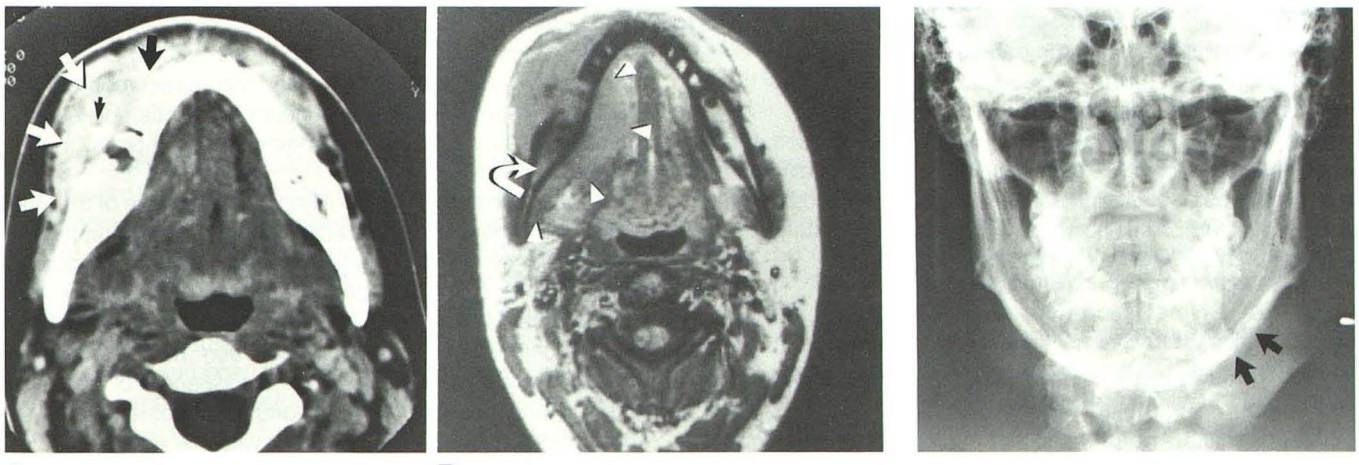


Fig. 5.—Chondroblastic osteosarcoma in 28-year-old woman.
A, Contrast CT scan with soft-tissue setting. Osteolytic lesion of body of right mandible with osteoid tumor mineralization (*small arrow*) and lateral soft-tissue extension (*large arrows*). Medial extraosseous extension is obliterating the fat plane lateral to the genioglossus muscle but is not well-defined.
B, MR image in T1-weighted pulse sequence. Better demonstration of medial border of soft-tissue component (*arrowheads*) and medullary infiltrative extension, decreasing normal high signal intensity of medullary bone (*curved arrow*).

Fig. 6.—Anteroposterior plain film shows left mandibular chondroblastic (high-grade surface) osteosarcoma in 30-year-old man. Subtle laminated periosteal reaction (*arrows*).

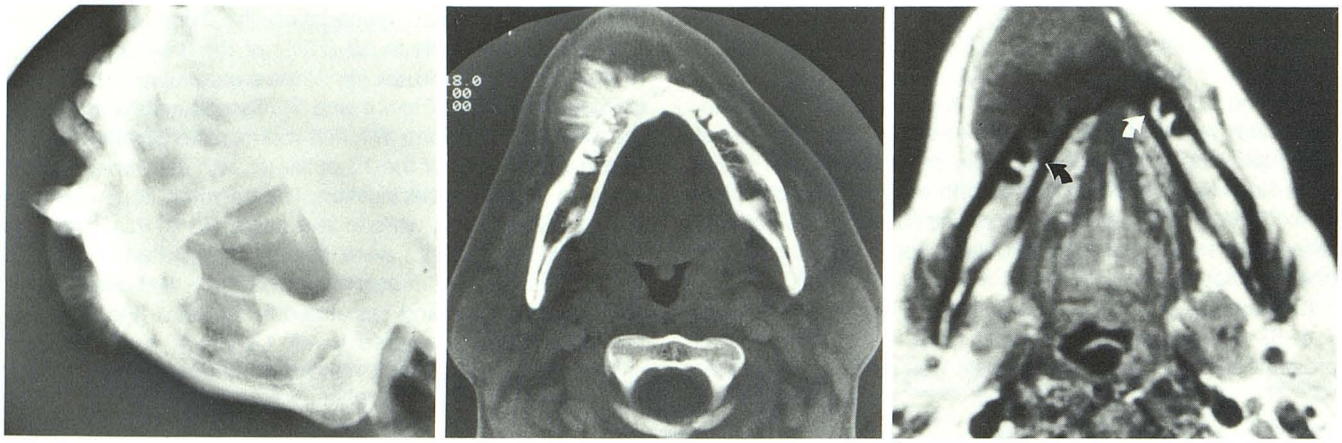


Fig. 7.—Right mandibular chondroblastic osteosarcoma in 40-year-old woman.
***A–C*, Plain film (*A*), CT (*B*), MR (*C*) of extensive spiculate pattern of parosteal reaction, best demonstrated on CT. However, MR, in T1-weighted pulse sequence, shows to best advantage the extent of medullary involvement (*arrows* in *C*).**

droblastic tumor subtypes (including a high-grade surface osteosarcoma), and two osteoblastic lesions (one well-differentiated osteosarcoma and one osteosarcoma resembling osteoblastoma); the 13th case was not available for review.

Cranial Bones (n = 8)

Four males and four females, ages 16 to 56 years (mean, 34.5 years) were in this category. There was no uniform presenting symptomatology. Painful swelling occurred in only one case. There was no cervical lymphadenopathy. The bony site of tumor origin included two parietal, two temporal, one occipital, one ethmoid, and two sphenoid. Seven were lytic (Figs. 8 and 9) and one was a parosteal osteoblastic osteosarcoma of the mastoid tip. Four (50%) had osteoid tumor mineralization; among these one had additional chondroid calcification. The transition zone was long in all cases except for the parosteal osteoblastic osteosarcoma. None had periosteal reaction. All had soft-tissue extension, and four of them contained calcification (50%).

Specimens were available for review in seven cases; there were four osteoblastic tumors (including one osteosarcoma resembling osteoblastoma and one parosteal osteosarcoma) and three fibroblastic lesions (including one telangiectatic subtype). There was no chondroid tumor.

Postradiation Osteosarcomas (n = 14)

Eight males and six females, ages 7 to 50 years (mean, 38.2 years) were in this category. A painless mass was the usual presenting symptom, occurring in 10 patients (71%). None presented with cervical lymphadenopathy. The latent period between radiation and detection of osteosarcoma ranged from 4 years, 2 months to 50 years, with a mean of 14 years. The information on radiation dosages was scanty, obtained in only six patients, and ranged from 3500–6400 rad (35–64 Gy).

Radiographically, these tumors were osteolytic in nine (64%) of the 14 patients, osteoblastic in three (21%), and mixed in two (14%) (Fig. 10). Disease was not confined to a single bone, except for one mandible previously radiated for an oral cavity carcinoma. Osteoid matrix calcification occurred in seven (50%) of the 14 patients; five cases (36%) had no matrix calcification. The transition zone was long in all, and none had periosteal reaction. All had soft-tissue extension, with calcification seen in half of these. Pathology specimens were available for review in only eight patients; five were chondroblastic, one osteoblastic, and two fibroblastic lesions.

Discussion

Osteosarcoma is a malignant tumor arising from undifferentiated connective tissue of bone. To qualify as an osteosarcoma, the neoplasm should have proliferating malignant cells that produce either osteoid substance or material histologically indistinguishable from it. Malignant fibroblastic or chondroblastic tumors with no neoplastic osteoid production are classified as fibrosarcomas and chondrosarcomas, respectively. Depending on the dominant differentiated element, osteosarcomas are divided into osteoblastic, chondroblastic, and fibroblastic types.

Osteosarcoma is primarily a malignant neoplasm of the long bones, with the greatest predilection for the metaphyses, most frequently the distal femur and proximal tibia. Excluding myeloma, it is the most common primary bone tumor and primarily affects older children and young adults [8, 9]. Seventy-five percent of patients are between 10 and 25 years old; very few osteosarcomas occur before age 5 or over age 30 [9]. Radiographically, most long bone osteosarcomas (46%) demonstrate a mixed pattern, with the osteoblastic or osteolytic type accounting for 32% and 22%, respectively. Periosteal reaction is associated with 80% of long bone lesions [9]. The majority of osteosarcomas have matrix mineralization, calcifications of the osteoid or osteoid-like substance within the tumor; the osteoid pattern creates a solid cloud or ivorylike increased density, the chondroid pattern creates a stippled, flocculent, or ring–arc pattern [6].

Osteosarcomas affecting the craniofacial bones occur infre-

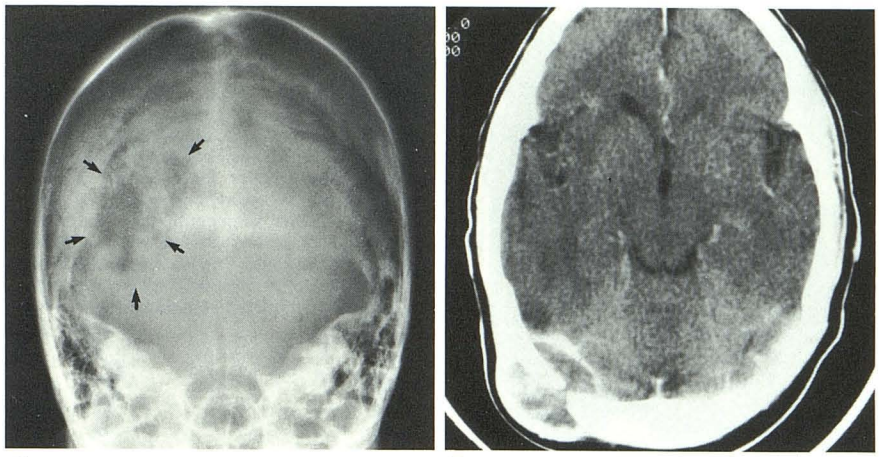


Fig. 8.—Fibroblastic osteosarcoma in 53-year-old woman with stage IV breast cancer.

A, Towne's view of skull shows poorly defined nonsclerotic osteolytic lesion in right parietooccipital bone (*arrows*).

B, Contrast-enhanced CT scan provides good demonstration of scalp and epidural tumor extension. No matrix calcification is noted. The lesion was treated as metastatic tumor with radiation (30 Gy). Surgical resection after no response to radiation revealed fibroblastic osteosarcoma.

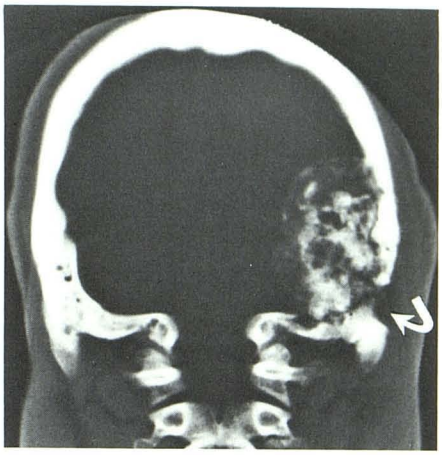
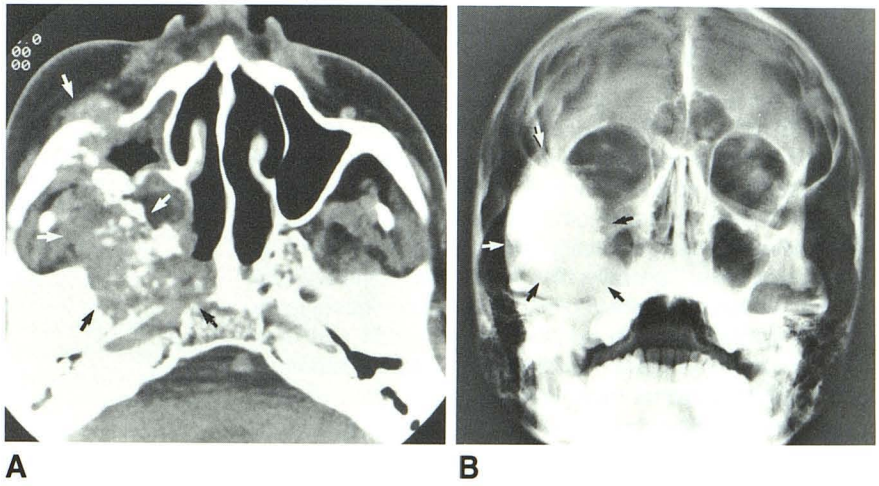


Fig. 9.—CT scan with bone setting shows osteoblastic osteosarcoma resembling osteoblastoma of left petrous ridge in 24-year-old man. Large, densely calcified mass arising from left petrous ridge with thin calcific rim periphery. Noted is a biopsy defect (*curved arrow*). No brain edema underneath is seen.

Fig. 10.—*A*, Postradiation chondroblastic osteosarcoma in 72-year-old man who had radiation therapy (64 Gy) for right nasopharyngeal carcinoma 12 years previously. Contrast-enhanced CT scan shows large destructive tumor (*arrows*) involving right maxilla, zygoma, sphenoid and temporal bones, and clivus, with soft-tissue extension and matrix calcification.

B, Postradiation chondroblastic osteosarcoma in 15-year-old girl who had radiation therapy (dose unknown) for right retinoblastoma 14 years previously. Water's view of skull shows large osteoblastic mass (*arrows*) involving right frontal bone, zygoma, and maxilla. Note involvement of multiple bones in both postradiation cases.



quently, constituting only 8.6% of Dahlin and Unni's series of 1274 cases [10]. The average age of our patients was 34 years, more than a decade older than those with osteosarcoma of the long bones. The presenting symptoms varied with the location of the tumors. Mandibular tumors frequently presented with focal painful swelling, while, conversely, the maxillary lesions usually produced no pain, as was the case with the cranial and postradiation osteosarcomas. There was no cervical lymphadenopathy at initial presentation in either de novo or postradiation cases.

All maxillary de novo osteosarcomas in our series arose from the alveolar ridge, and most of the mandibular lesions were located in the body of the mandible. The postradiation osteosarcomas occurred at the periphery of the radiation field, and usually affected more than one bone. Excluding the mandibular osteosarcomas, the radiographic presentation in two-thirds of the craniofacial osteosarcomas was osteolytic, occurring in 22 (67%) of the 33 cases. Except for two cases of parosteal osteosarcoma with no radiologic abnormalities of the underlying bone, all had a long transition zone, an observation significantly different from that in long bone tumors. The dominant pattern in the mandibular tumors was osteoblastic (46%), and occasionally the transition zone was short. Periosteal reaction was rarely observed in craniofacial osteosarcoma, and only in the mandibular lesions. Tumor matrix mineralization was frequent, particularly in mandibular and maxillary lesions. Osteoid calcification was the most common form, although chondroid differentiation was the dominant histologic subtype.

Secondary osteosarcoma associated with preceding bone disorders such as Paget disease is well recognized [11]. In our craniofacial series, prior radiation therapy for neoplastic or infectious disease was the only predisposing factor for secondary osteosarcomas. The clinicopathologic correlation of radiation-induced osteosarcoma has been well described [12–14]. Our series differed from previous reports in that the

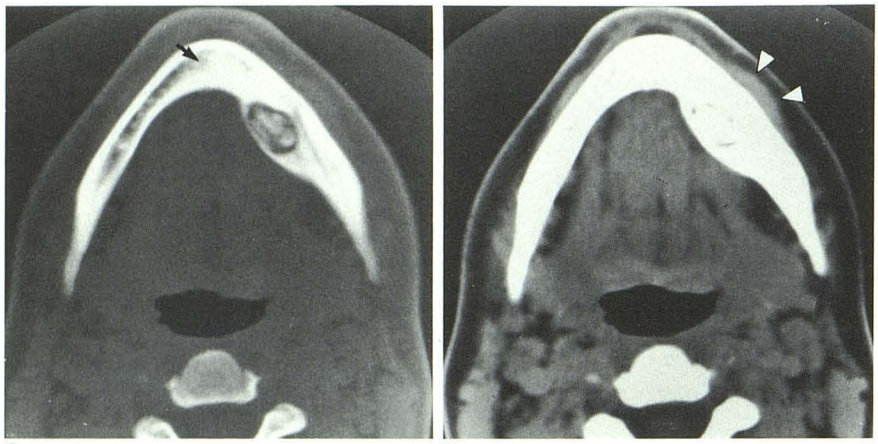


Fig. 11.—Osteosarcoma in 25-year-old woman presenting with painful left jaw swelling of 9 months' duration.

CT scan with bone setting (*A*) shows expansile osteoblastic left mandible with central radiolucency containing islands of osteoid matrix calcifications. Aggressive osteoblastoma appears a likely diagnosis. However, the presence of intramedullary extension (*arrow*) and soft-tissue component (*arrowheads*) demonstrated on the same scan in soft-tissue setting (*B*) indicates the malignant nature of this tumor. The histologic diagnosis was low-grade, well-differentiated osteoblastic osteosarcoma, resembling osteoblastoma.

mean latent period was slightly longer and there were more chondroblastic than fibroblastic subtypes [14]. The unique locations of postradiation osteosarcoma at the periphery of the radiation fields result from the administered radiation being unable to destroy all viable cells but able to induce malignant transformation [12]. The previously noted relative frequency of craniofacial bones as sites of postradiation osteogenic sarcoma [13] is supported by our collection of 14 of these, of a total of 46.

The presence of tumor matrix mineralization, either osteoid or chondroid, with aggressive bone destruction and soft-tissue extension, leads directly toward a radiologic diagnosis of osteosarcoma without much difficulty. The differential diagnosis between osteosarcoma and chondrosarcoma may be troublesome radiographically, and sometimes impossible even histologically. Generally, the chondrosarcomas are even rarer in the head and neck region. They appear less aggressive radiographically, with less bone destruction and more bone erosion. Osteosarcoma resembling osteoblastoma or parosteal osteosarcoma might mimic "benign" bone growth, aggressive osteoblastoma or osteochondroma, but the presence of a soft-tissue component and intramedullary extension will provide a clue (Fig. 11). It may be difficult to differentiate between postradiation osteosarcoma and the simple radiation osteitis often associated with it [14]. Nevertheless, the associated soft-tissue extension strongly favors tumor and justifies a prompt tissue biopsy. Occasionally, osteosarcoma may be indistinguishable from the more common metastatic carcinoma, when it presents without matrix mineralization.

Conventional radiographs are of limited worth in evaluating head and neck osteosarcomas because of the superimposed bony structures. However, they can be very useful adjuncts to CT for maxillary and mandibular tumors when there are extensive metallic tooth fillings or permanent dentures. CT provides excellent detection of tumor calcification, cortical involvement, and, in most instances, soft-tissue as well as intramedullary extension. MR is even more effective in demonstrating the intramedullary and extraosseous tumor components unequivocally on both T1- and T2-weighted images [15, 16]. The T1-weighted images provide a better anatomic definition, and T2-weighted images demonstrate additionally the peritumoral edematous reaction. Furthermore, in the paranasal sinuses the T2-weighted images will distinguish relatively hypointense tumor extent from the hyperintensity of retained mucous debris in the obstructed sinuses [17]. However, CT and plain films are superior to MR in detecting the matrix calcifications and bone destruction or reaction.

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