

Primary Osteosarcoma of the Head and Neck in Pediatric Patients

A Clinicopathologic Study of 22 Cases with a Review of the Literature

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BACKGROUND. Primary osteosarcomas of the head and neck in the pediatric age group, not associated with previous irradiation or a known syndrome, are rare. The literature contains several single cases and small study series; however, to the authors's knowledge, there has been no comprehensive large study to evaluate the clinicopathologic aspects of these tumors.

METHODS. Twenty-two cases of osteosarcomas of the head and neck in patients 18 years of age or younger, diagnosed between 1970 and 1997, were retrieved from the Otorhinolaryngic–Head & Neck Tumor Registry of the Armed Forces Institute of Pathology (AFIP). No secondary sarcomas (radiation-induced or those arising after chemotherapy) or those associated with known syndromes were included. Clinical, radiographic, and histologic features were reviewed, and patient follow-up was obtained.

RESULTS. The patients included 11 girls and 11 boys, 1–18 years of age (mean, 12.2 yrs). Patient symptoms related to tumor location were painless swelling, loss of teeth, headaches, or a mass lesion, present for an average of 5.9 months. No genetic abnormalities were documented. The tumors most frequently involved the mandible ($n = 19$), followed by the sphenoid sinus ($n = 2$) and the maxilla ($n = 1$). The tumors ranged in size from 1.1–10.0 cm (mean, 4.5 cm). All tumors were invasive and malignant by radiology and/or histology. The tumors were Grade 1 ($n = 11$), Grade 2 ($n = 8$), or Grade 3 ($n = 3$). All cases, except one chondroblastic osteosarcoma, were osteoblastic osteosarcomas. Thirteen patients underwent initial surgical resection with ($n = 5$) or without ($n = 9$) additional radiation and/or chemotherapy. The remaining 9 patients had an initial biopsy for diagnosis followed by surgery ($n = 4$) or surgery and radiation and/or chemotherapy ($n = 5$). Follow-up was available for 19 patients: 13 were alive at last follow-up with no evidence of disease (mean, 13.1 yrs); 1 was alive with disease (1.3 yrs); 3 had died without evidence of disease (mean, 23.2 yrs); and 2 had died of disease (mean, 7.8 yrs). The 3 patients with high-grade osteosarcoma were alive without disease (mean, 20.0 yrs).

CONCLUSIONS. Primary head and neck osteosarcomas in the pediatric population are typically low- to moderate-grade lesions in the mandible. Despite the invasive nature and high grade of a few of these tumors, there is an excellent overall long-term prognosis for patients in this age group with tumors in these locations. *Cancer* 2001;91:598–605. Published 2001 by the American Cancer Society.*

KEYWORDS: osteosarcoma, osteogenic sarcoma, bone, head and neck, sinuses, mandible, nasal cavity, pediatric, adolescent, child, treatment, prognosis, outcome.

Osteosarcomas are the most common primary malignant neoplasms of bone^{1–5} that can occur anywhere in the body but are found most commonly in the long bones, especially around the knee.

Osteosarcomas of the head and neck represent a small percentage of osteosarcomas with studies reporting occurrences between 0.5–8.1%^{1,5–12} and an average between 6–7%. Head and neck osteosarcomas are infrequent and usually present in the third to fourth decades of life, almost a decade after the most common presentation of long-bone tumors.^{2,13,14} Osteosarcomas of the head and neck usually occur as secondary tumors after radiation therapy or chemotherapy for another tumor,^{15–23} although syndromic association^{24–26} and development of osteosarcomas from a preexisting benign tumor²⁷ also have been described. Pediatric patients with primary osteosarcoma of the head and neck are rare, with only a handful of case reports and small study series (up to 7 patients) reported in the English-language literature (MEDLINE 1966–2000).^{5,8,11,26,28–38} Given the relative infrequency of osteosarcomas in the head and neck in pediatric or adolescent patients and the lack of a comprehensive series to date, we undertook a study of a large group of patients in these categories. We compared their clinical presentations, radiographic findings, pathologic features, treatment protocols, and clinical outcome with those patients reported in the literature.

MATERIALS AND METHODS

Twenty-two osteosarcomas of the head and neck in pediatric patients were identified in the files of the Otorhinolaryngic–Head and Neck Tumor Registry of the Armed Forces Institute of Pathology (AFIP) between 1970 and 1997. We defined pediatric patients as those persons 18 years of age or younger at the time of initial presentation. These 22 cases were identified in a review of 1947 (1.1%) pediatric benign or malignant tumors of the head and neck region diagnosed during the same time period. Twenty-one cases were obtained from civilian sources, including foreign countries, and 1 case was received from a military hospital.

As a point of comparison, the English-language literature was reviewed for all reported primary osteosarcomas (nonradiation associated and nonsyndromic) in the head and neck in children 18 years old or younger.

Inclusion in this study required the production of osteoid (unmineralized osseous matrix) by atypical, neoplastic osteoblasts with distinctive biologic activity and morphologic patterns, i.e. osteosarcoma. All lesions showed destruction of bone or invasion of soft tissue by radiographic and/or pathologic measures. A consensus was reached for tumor grade by all authors on the hematoxylin and eosin stained slides, available in all cases. We specifically excluded any patients who had received radiation treatment and/or chemotherapy for a previous benign or malignant process (i.e., thymic enlargement, acne, retinoblastoma, leukemia,

and lymphoma) or patients who had secondary osteosarcomas as part of a syndrome.

Materials within the files of the AFIP were supplemented by a review of the patients' demographics, symptoms at presentation, history of previous irradiation or chemotherapy, radiographic findings, surgical pathology reports, operative reports, cancer registry records, and written questionnaires or oral communications with treating physicians. Follow-up data included information regarding the exact location of the primary tumor site, the specific treatment modalities used, and the current status of the disease and patient. This clinical investigation was conducted in accordance and compliance with all statutes, directives, and guidelines of the Code of Federal Regulations, Title 45, Part 46, and the Department of Defense Directive 3216.2 relating to human subjects in research.

RESULTS

Clinical

The patients included 11 girls and 11 boys (Table 1). The patients age range was 1–18 years, with a mean age at presentation of 12.2 years. There were no differences between the genders in age at presentation (females: mean, 12.3 yrs; males: mean, 12.1 yrs). Seventeen patients were Caucasian, 2 were African-American, and 3 were Native American.

Nearly all of the patients presented with swelling or a mass lesion ($n = 21$), occasionally associated with tenderness or pain ($n = 5$; Table 1). In a few cases ($n = 4$) the mass lesion caused disruption of the dentition, including malocclusion and loosening of teeth. Two patients had noticed weight loss, whereas 3 patients reported a history of trauma 1–3 months before initial presentation. One patient presented with headaches and lethargy without a mass lesion on physical examination. The duration of symptoms ranged from 1–28 months, with a mean of 5.9 months. There were no differences among the various anatomic sites in average duration of symptoms.

Patients with prior exposure to radiation or chemotherapy or both were excluded from consideration by study design. Further, no patient had evidence for a genetic abnormality (syndrome) or other associated diseases.

Radiographic Studies

Radiographic studies were performed on all patients in this study, but the physical images were returned to the contributing hospital before this study began in a few cases, which allowed review of only the radiology reports in those cases. All patients had plain X-ray films of their tumors. Advanced imaging technique films were available in a number of cases, including

TABLE 1
Clinical and Radiographic Features of 22 Osteosarcomas of the Head and Neck in Pediatric Patients

Patient	Gender/age ^a	Clinical symptoms (duration of symptoms, in months)	Radiographic studies
1	M/9	Headaches and lethargy (3)	Erosion of the sella tursica and sphenoid sinus
2	F/7	Swelling, tenderness, paresthesias of the lip (1)	Microcalcifications in an elevated periosteum, expansile mass in mandible, with soft tissue extension
3	M/9	Painless swelling of face; history of trauma (12)	CT and X-rays demonstrated mandibular mass with soft tissue extension
4	M/16	Pain, mass behind lower teeth (5)	Mass over the mandible and lower teeth region
5	M/10	Mass in mandible; paresthesias of lower lip (0.8)	Cluster of radioopacities at the mandibular angle
6	F/13	Mass in jaw; malocclusion of teeth (1)	Well demarcated unilocular radiolucent mass with calcifications
7	M/13	Swelling of lower jaw (6)	Lytic lesion of the bone with soft tissue extension, minimal periosteal elevation
8	M/17	Mass in tongue; history of trauma (12)	Expansile radiolucent to radioopaque mass with microcalcifications
9	M/9	Mass in right cheek (6)	Expansile, radiolucent without peripheral sclerosis, displacing tooth
10	F/14	Mass in mandible; loosening of teeth (4)	Smooth mass, eroding through the buccal plate with periosteal elevation
11	M/14	Expanding mass; 5 lbs. weight loss; history of trauma (5)	Anterior mandible at symphysis, cystic with bone destruction; spiculations
12	F/4	Mass (4)	Mass in the mandible on CT
13	F/10	Mass in mandible (6)	Multilocular mass in the mandible
14	F/15	Mass in mandible, gingiva and floor of the mouth (8)	Mass on the internal ramus of the mandible with soft tissue invasion
15	F/15	Mass anterior jaw; teeth loosening (4)	Periosteal elevation, multilocular, radiolucent
16	F/17	Maxilla mass extending into orbit (8)	Radiolucent destructive lesion
17	F/11	Swelling of jaw; 14 lbs. weight loss (5)	Large mass occluding the mandible, maxillary sinus, and nasopharynx
18	M/17	Rapidly enlarging mandibular mass (2)	Large radiolucent mass with internal "bony" structures
19	M/18	Rapidly enlarging mandibular mass (2)	Radiolucent premolar and first molar region mass with irregular borders
20	F/16	Swelling in the mandible (1)	Mass in the mandible expanding the bone
21	F/13	Swelling of the left cheek (28)	Mandible mass with soft tissue extension
22	M/1	Nostril mass with ptosis of eyelid (8)	Mass extending from nasal cavity into sphenoid, ethmoid & base of skull

M: male; F: female.

^a Age is listed in years.

computed tomograms and/or magnetic resonance images. The imaging findings are recorded in Table 1, and most images demonstrated an expansile radiolucent mass, centered in and destroying the bone, frequently demonstrating soft-tissue extension. Periosteal elevation was noted in a few cases ($n = 3$).

Treatment and Follow-up

All patients (100%) were managed initially by surgical excision, either by an excisional biopsy or by a more radical procedure (Table 2). Three patients who were not U.S. citizens were lost to further follow-up. Of the remaining 19 patients, 2 received radiation therapy alone after the surgical resection, 5 received chemotherapy, and 2 had a combination of radiation and chemotherapy. Details of the chemotherapeutic regimens are unknown. The durations of treatment and lengths of time until patient response are unknown. There were not enough patients in each therapeutic modality to reach statistical significance as to whether treatment affected outcome.

Overall, no patients developed metastatic disease, although 7 patients (31.8%) developed recurrent local and/or residual local disease. When recurrent local

disease was diagnosed, additional surgery ($n = 7$) and adjuvant therapy ($n = 5$) were used. There was no specific anatomic site that was predisposed to local recurrence, as the tumors involved the maxilla, mandible, sphenoid sinus, and ethmoid sinus. Of the locally recurrent tumors, 3 were Grade 1, 3 were Grade 2, and 1 was Grade 3. Of the patients who died of local disease, all had developed local recurrences. Conversely, 5 patients with local recurrences still were living at last follow-up, 4 without any current residual/recurrent disease (including the Grade 3 tumor patient). The patients who developed recurrent disease usually developed the recurrent tumor within 1 year of their original diagnoses.

Follow-up ranged from 0.8–38.9 years. As a group, the patients had an excellent overall survival, with a mean follow-up of 13.5 years, irrespective of the outcome. Two patients died with or as a result of locally recurrent disease (mean, 7.8 yrs). Both of these patients had been treated with radical surgery and radiation, and one patient also received chemotherapy. One of the lesions (Grade 1) involved the sphenoid sinus with extension into the base of the skull, whereas the other patient had a mandibular lesion

TABLE 2
Tumor Location, Grade, Treatment, and Clinical Outcome of 22 Osteosarcomas of the Head and Neck in Pediatric Patients

Patient	Side/cm	Exact location	Tumor grade	Initial treatment	Additional therapy	Recurrent disease	Status (yrs)
1	M/1.1	Sphenoid	1	Biopsy	S, Rads: 4500	Y	D, LD (0.8)
2	L/1.2	Mandible	3	Biopsy	S: partial mandibulectomy, C	N	A, NED (11.8)
3	R/2.5	Mandible	1	RR	None	N	D, NED (1.8)
4	R/2.5	Mandible	1	Biopsy	S: H	N	A, NED (11.5)
5	L/2.5	Mandible	2	H	S, Rads, C	Y: multiple	D, NED (29.0)
6	L/3.0	Mandible	1	H	C	N	A, NED (4.0)
7	L/3.0	Mandible	3	RR	S: subtotal resection, C	Y: two	A, NED (28.0)
8	R/3.5	Mandible	1	RR	S: definitive resection	N	A, NED (7.6)
9	R/3.5	Mandible	1	H	None	N	A, NED (8.8)
10	R/4.0	Mandible	1	Biopsy	S: partial mandibulectomy	N	A, NED (2.7)
11	R/4.5	Mandible	2	Biopsy	S: H, C	Y	A, NED (4.3)
12	L/5.5	Mandible	2	H	S, Rads, C	Y: multiple	D, LD (14.8)
13	L/6.0	Mandible	2	H	None	N	A, NED (11.5)
14	R/7.0	Mandible	2	Biopsy	S: H, Rads: 3000	N	A, NED (29.2)
15	M/7.0	Mandible	1	H	NR	NR	LTF
16	R/7.0	Maxilla	1	H	S: wider excision	Y	A, LD (1.3)
17	L/8.0	Mandible	3	H	C	N	A, NED (20.4)
18	R/10.0	Mandible	2	H	NR	NR	LTF
19	R	Mandible	1	Biopsy	S: H	N	A, NED (4.6)
20	R	Mandible	1	Biopsy	S: H	Y: one	A, NED (26.4)
21	L	Mandible	2	H	None	N	D, NED (38.9)
22	L	Nasal cavity; ethmoid, sphenoid, base of skull	2	RR	NR	Y: residual tumor	LTF

M: midline; L: left; R: right; RR: radical resection; H: hemimandibulectomy; Y: yes; N: no; S: surgery; Rads: radiation therapy; C: chemotherapy; NR: not reported; D, LD: Dead, with local disease and extension into the base of the brain or the base of the skull; A, NED: Alive, no evidence of disease; LTF: Lost to follow-up; A, LD: Alive, local persistent/recurrent disease.

(Grade 2), which also recurred and developed an extension into the base of the brain.

At last follow-up (1.3 yrs), one patient was alive and had local disease in the maxilla (Grade 1), having received surgical excision only. The remaining patients were either still living and had no evidence of disease at last follow-up (*n* = 13; mean, 13.1 yrs) or had died of unrelated causes and had no evidence of disease (*n* = 3; mean, 23.2 yrs).

These findings yielded a 63.2% 5-year raw survival rate and a 52.6% 10-year raw survival rate. Therefore, it appears that patients may develop recurrences, but the recurrences do not adversely affect their overall survival rate when the recurrences are managed appropriately. The 3 patients who had high-grade osteosarcomas were disease free at last follow-up (mean, 20.0 yrs), even though 1 patient initially had developed 2 local recurrences during the first 3 years of follow-up.

Pathologic Features

Macroscopic findings

The mandible (*n* = 19) was affected most frequently, followed by 2 tumors in the sphenoid sinus and 1 in

the maxillary sinus (Table 2). There were no cases presenting in the larynx, orbit, or initially in the skull base. The tumors ranged in size from 1.1–10.0 cm, with a mean of 4.5 cm. Most of the lesions were received as multiple, irregular fragments of bone and soft tissue, especially in the biopsy specimens. The resection specimens (mainly hemimandibulectomies) demonstrated bony destruction by tumor. The margins of resection were free of tumor in most cases according to the referral macroscopic descriptions.

Microscopic findings

All lesions, in all locations, demonstrated the typical features of osteosarcoma, including osteoid (noncalcified, eosinophilic osseous matrix) production by atypical, neoplastic osteoblasts. There was an overall loss of normal intramedullary architecture, destruction of cancellous bone, and a number of areas of tumor necrosis. Histologic invasion into the surrounding bone or soft tissue was demonstrated in nine cases. There was no appreciable difference in the histologic grade of the invasive component compared with the intraosseous component. The tumor cells

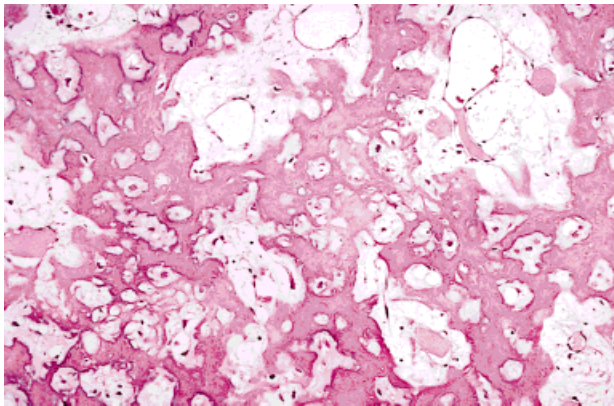


FIGURE 1. A low-grade osteosarcoma reveals atypical cells producing unmineralized osseous matrix.

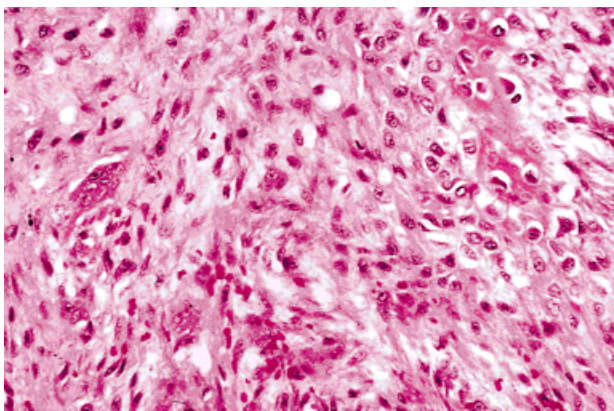


FIGURE 2. An intermediate-grade osteosarcoma with abundant atypical cells and admixed "lace-like" osteoid.

demonstrated cytologic atypia: increased nuclear to cytoplasmic ratio, nuclear chromatin irregularities, and an increase in mitotic figures, including atypical forms. All tumors, with one exception (a Grade 2 chondroblastic osteosarcoma), were osteoblastic osteosarcomas. The cases were graded as low-grade tumors (Grade 1), intermediate-grade (Grade 2), and high-grade (Grade 3), as previously described.^{2,7,8,38-40} Eleven cases were well differentiated (Grade 1, see Figure 1) tumors, 8 cases were moderately differentiated tumors (Grade 2, see Figure 2), and 3 cases were poorly differentiated (Grade 3, see Figure 3) tumors.

DISCUSSION

Osteosarcomas of the head and neck are rare and have a reported incidence of 1.7–5% of all head and neck primary tumors,^{6,28,41} whereas maxillofacial osteosarcomas account for 4–9% of all osteosarcomas.^{1,5,7,8,10-12,29,35,41,42} These often occur as secondary tumors after radiation or chemotherapy. Fur-

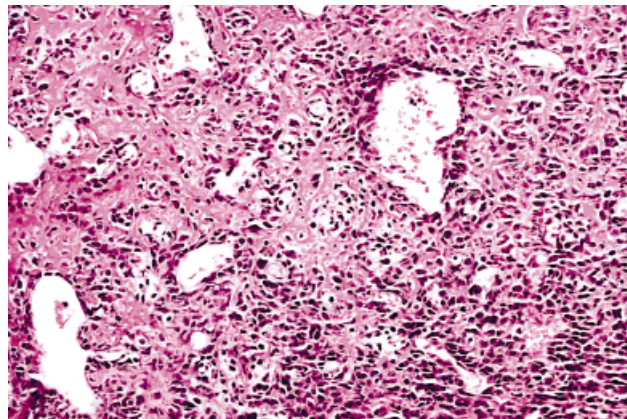


FIGURE 3. A high-grade osteosarcoma with malignant spindled areas and a small amount of recognizable osseous matrix.

ther, the incidence of head and neck osteosarcomas is even more uncommon if only the pediatric patient population is examined, accounting for less than 1% of all head and neck malignant tumor cases reviewed in pediatric patients (age < 18 yrs) at our institution. Head and neck osteosarcomas accounted for approximately 0.47% of all osteosarcomas in all sites in adults, while the 22 cases of head and neck osteosarcomas composed 2.7% of all pediatric osteosarcomas in all anatomic sites of the body. Therefore, osteosarcomas of the head and neck in pediatric patients are more common than those in the adult population, and only a small percentage of all osteosarcomas occur in the head and neck region.

An accurate comparison with the reported cases in the literature is difficult because of the overall lack of case data in pediatric patients. The results are, instead, incorporated into studies about all head and neck sarcomas or all head and neck tumors in general, specifically in adult patients. Still, where possible, our overall findings are similar to the aggregate of those in the English-language literature (Table 3). Remarkably, there is no gender predilection in this reported series or in those patients reported in the literature. There was only a slight, insignificant difference in the average age at presentation between our cases (12.2 yrs) and those in the literature (13.7 yrs), but the very young tended not to be affected as frequently as adolescents.

All of the patients in this study series and those in the literature presented clinically with symptoms related to anatomic location, especially by swelling ($n = 41$),^{1,5-7,12,38,39} with or without pain, followed by dental problems, ulceration, and neurasthenias. The patients experienced a fairly short overall duration of symptoms (mean, 5.9 mos), without any notable differences when comparing the anatomic site of in-

TABLE 3
Literature Summary of Pediatric Patients with Primary Head and Neck Osteosarcomas^a

All cases	Parameter ^b
Gender	
Females	23
Males	23
Age at presentation	
Range, all	0.02–18 yrs
Average, all	13.7 yrs
Females (average)	13.7 yrs
Males (average)	14.1 yrs
Clinical presentation	
Swelling	22
Ulceration	8
Pain	3
Proptosis	1
Exact location	
Mandible	27
Maxilla or maxillary sinus	15
Ethmoid sinus	1
Tumor size (cm)	
Range	2–17 cm
Average	6.9 cm
Pathology	
Osteogenic osteosarcoma	11
Osteoblastic	5
Fibroblastic osteosarcoma	3
Parosteal osteosarcoma	2
Round cell osteosarcoma	2
Chondroblastic osteosarcoma	2
Grade	
1	7
2	7
3	3
4	1
Treatment	
Surgery alone	19
Surgery and radiation	8
Surgery and chemotherapy	6
Surgery, radiation and chemotherapy	3
Radiation alone	1
Radiation and chemotherapy	1
Outcome	
Alive, no evidence of disease	22
Range of months survived	7–216 mos
Mean survival	92.0 mos
Dead, with disseminated disease	10
Range of months survived	5–36 mos
Mean survival	19.6 mos

^a Nonradiation induced and not part of a syndrome, in reports where clinical and histologic parameters were described.^{5,8,11,26,28–38}

^b Parameter was not always stated in the report, and therefore the numbers do not necessarily equal the total values in the columns.

involvement. To our knowledge, this parameter has been reported only in isolated case reports.^{5,17,43}

Although the tumors affected different anatomic sites, by far the majority involved the mandible (this study series, 86%; literature, 64%), followed by the

maxilla/maxillary sinus (36%), and other sinuses (sphenoid, *n* = 2; ethmoid, *n* = 2). The reason for this site predilection is unclear to us, although researchers have hypothesized that because the mandible retains growth centers for more than 30 years, osteosarcomas of the head and neck could occur more commonly in this location.^{7,8,13,14,33} However, it is possible that primary osteosarcomas of other head and neck locations are easier to recognize and, hence, do not require a second opinion from a referral center such as ours.

When the size of the tumor was reported in published studies,^{5,19,29,35,44} the mean size was 6.9 cm, slightly larger than the mean size of 4.5 cm for this study series. However, if the single largest tumor (17 cm) reported in the literature is removed from consideration,⁵ then the overall mean size for the cases in the literature is 5.0 cm, closer to the 4.5 cm reported herein.

All of the tumors in our study series were categorized as osteogenic sarcomas, predominantly osteoblastic subtype, with one chondroblastic subtype. The cases in the literature were divided into osteoblastic (*n* = 5), fibroblastic (*n* = 3), parosteal (*n* = 2), chondroblastic (*n* = 2), and round-cell type (*n* = 2; Table 3). Despite these different subtypes, most of the cases reported in the literature were Grade 1 or Grade 2 tumors (Grade 1, 41%; Grade 2, 41%), almost identical to the tumor grading of the cases in this study series (Grade 1, 50%; Grade 2, 36%). Therefore, it appears that there is a penchant for head and neck osteosarcomas to occur as lower grade tumors.^{2,34,43}

Surgery was the initial treatment of choice for osteosarcomas and was used for all of the patients in this series and for nearly all of the cases in the literature (Table 3). The use of adjuvant therapy, either radiation (*n* = 2), chemotherapy (*n* = 5), or a combination of radiation and chemotherapy (*n* = 2) did not seem to yield a prolonged survival. There were an insufficient number of cases in each category to determine a statistically significant result. This finding is similar to that reported in the literature (Table 3). Overall, however, complete surgical eradication of the tumor appears to be the minimum therapeutic intervention required.

The patients in this study series had an overall good long-term prognosis, regardless of age, gender, anatomic location of the tumor, tumor grade, or whether or not they received adjuvant therapy. There were no metastases in this series of patients. The exact adjuvant therapeutic regimens and lengths of time to relapse in these patients are unavailable. Our follow-up and treatment findings are difficult to correlate with the literature because outcome data for pediatric patients has been included with adult data in the published studies.^{2,6,9,10,12,14,18,37,42,45–49} When data are

separated, the published studies do not indicate whether the figures are for disease-free or raw survival rates. Nonetheless, those studies that did report survival rates indicated ranges of 10–35% 5-year survival rates.^{5,8–10,37,40,50} These figures are quite a bit lower than the patients in this study, where 84.2% were alive or had died of unrelated causes at last follow-up, an average of 15.0 years after initial presentation. None of our patients developed metastatic disease, even though metastases to the lung and bone have been reported.^{7,35} The results for our series of patients support the assertion in the literature that maxillofacial osteosarcomas have, in general, a better prognosis than long-bone osteosarcomas, which have an overall 5-year survival rate of 30–40%.^{51,52} There are some data that suggest that this prognosis is increasing with better adjuvant therapies.⁷ In contrast to findings in the literature, we did not find that high-grade osteosarcomas have a poorer outcome when they arise in the head and neck than if they arise in the peripheral skeletal system.^{39,41,52} In fact, the 3 patients with Grade 3 tumors were all alive at last follow-up without evidence of disease over a mean period of 20.0 years, even though 1 patient had developed intercurrent disease.

Osteosarcoma is a malignant tumor of bone formation. The presence of neoplastic lace-like osteoid and osteoblastic pleomorphism and cytologic atypia, with or without ischemic changes, distinguishes these tumors from osteoblastomas and other fibro-osseous benign or malignant lesions. Even the Grade 1 osteoblastic osteosarcomas had parallel bony trabeculae, mitotic activity, stromal cellularity, and radiographic and/or pathologic evidence for bony destruction and/or soft tissue invasion. Osteoblastoma has only mature osteoblasts lining the woven and lamellar trabecular bone without a destructive pattern or soft tissue invasion. Additionally, osteoblastomas have an increased vascularity and evidence of osteoclastic activity. Occasionally, well differentiated osteosarcomas can be difficult to distinguish from osteoblastomas on initial review. However, the presence of atypical cells and destructive radiologic appearance of such osteosarcomas will distinguish them from osteoblastomas. None of our well differentiated osteosarcomas were histologically similar to osteoblastomas. Fibrous dysplasia has Y- or C-shaped trabeculae with blander stroma and absence of bony destruction or soft tissue invasion. Osteoma, purely an osseous lesion, lacks the atypical osteoblasts, mitotic activity, and cellular stroma of osteosarcoma.

In summary, children and adolescents of either gender can develop primary nonradiation-associated osteosarcomas in the head and neck region, most commonly arising in the mandible. These patients

usually will present with symptoms of swelling and a mass of short duration, although dental problems and neurasthesias also have been observed. After radiographic studies, complete surgical extirpation followed by appropriate adjuvant therapy for osteosarcoma will allow pediatric patients to demonstrate a good overall prognosis, irrespective of gender, anatomic location, size of tumor, or tumor histology. In our experience, even with multiple local recurrences and locally invasive qualities, these tumors tend to behave as low-grade neoplasms with a good long-term patient survival.

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