The Journal of Laryngology and Otology
November 1994, Vol. **108**, pp. 947–953

Sarcomas of nasal cavity and paranasal sinuses: chondrosarcoma, osteosarcoma and fibrosarcoma

V. KOKA, M.D., R. VERICEL, M.D., E. LARTIGAU, M.D., A. LUSINCHI, M.D., G. SCHWAAB, M.D.

Abstract

Forty-two patients were treated for sarcoma of the nasal cavity and paranasal sinuses at the Institut Gustave Roussy, Paris, between 1960 and 1993. Twelve patients had chondrosarcoma (CS), 14 had osteosarcoma (OS) and 16 had fibrosarcoma (FS). Ten patients had grade I, six grade II and 26 grade III tumours.

All but 10 patients had surgery for the primary tumour. A significantly increased risk of local failure was associated with the male sex ($p < 0.01$), grade III tumours ($p < 0.02$) and patients excluded from surgery ($p < 0.04$). The overall incidence of local and distant failure was 76 and 12 per cent respectively. Overall survival was 28 per cent at three years and 23 per cent at five years. Eight patients (20 per cent) were alive more than 10 years later. The factors significantly influencing survival were sex ($p < 0.01$), grade ($p < 0.05$) and local failure ($p < 0.01$).

Key words: Paranasal sinus neoplasms; Nasal neoplasms; Chondrosarcoma; Osteosarcoma; Fibrosarcoma; Surgery; Radiotherapy; Chemotherapy, adjuvant; Combined modality therapy

Introduction

Sarcomas of the nasal cavity and paranasal sinuses are infrequent and account for seven to 28 per cent of all the sarcomas of the head and neck region (Friedman *et al.*, 1989; Wanebo *et al.*, 1992). These tumours are biologically diverse depending on the type, histological grade and site of occurrence (Mandard *et al.*, 1989). There is a high rate of local recurrence with a much reduced risk of distant metastasis for sarcomas of head and neck (Malawer *et al.*, 1989) and death usually results from intracranial extension of uncontrolled local disease (Fu and Pezin, 1974). Because of the biological diversity, we tried to identify clinical and pathological characteristics that may predict prognosis and to evaluate the treatment modalities for chondrosarcoma (CS), osteosarcoma (OS) and fibrosarcoma (FS) of the nasal cavity and paranasal sinuses.

Methods

Forty-two patients were treated for CS, OS and FS of the nasal cavity and paranasal sinuses at the Institut Gustave Roussy, Paris, between 1960 and 1993.

Three patients had been referred after the primary treatment for CS had failed elsewhere. All the clinical, radiological and histological data of the primary tumour of these three previously treated patients had been obtained and was studied along with other previously untreated patients. The pathological features of the primary tumours were reviewed and tumours were graded depending on their histological appearance: grade I – well differentiated tumours; grade II – moderately differentiated tumours; and grade III – poorly differentiated tumours. Since it is difficult to employ the staging systems used for extremity lesions for classification of head and neck sarcomas, we adopted the TNM classification for cancer of nasal cavity and paranasal sinuses (UICC, 1987).

The outcome and survival were analysed along with the clinical and histological characteristics and treatment modalities. The disease-free period and overall survival were calculated by the method of Kaplan and Meijer (1958). The qualitative variables were compared by χ^2 and continuous variables by Student's *t*-test. Comparison of survival between the subgroups of patients was performed with the log–rank statistical test (Peto and Pike, 1973). The $p \leqslant 0.05$ was considered statistically significant.

Materials

The ages of the patients ranged from seven to 67 years (mean 36 years). Two-thirds of the patients in the series were aged between 20 and 50 years. The mean ages for OS, CS and FS cases were 33, 35 and 42 years respectively. There were six children (14 per cent) aged between seven and 15 years. One of them had CS, three had OS and two had FS (Table I).

There were 18 male (43 per cent) and 24 female (57 per cent) patients (Table I). The sex incidence was equal for OS but a female predominance was noted for CS (2:1) and FS (4:3). Three patients (seven per cent) had a previous history of irradiation to the head and neck area. One

From the Department of Head and Neck Surgery, Institut Gustave Roussy, Paris, France.
Accepted for publication: 22 July 1994.

TABLE I
PATIENT AND DISEASE CHARACTERISTICS

Characteristics	CS (n = 12) n	(%)	OS (n = 16) n	(%)	FS (n = 14) n	(%)	Total (n = 42) n	(%)
Age								
≤ 15 years (paediatric)	1	8%	3	19%	2	14%	6	14%
(Age range)	(15)		(7–11)		(10–15)		(7–15)	
> 15 years (adult)	11	92%	13	81%	12	86%	36	86%
(Mean age ± SD)	(36.7 ± 15.4)		(38.4 ± 12.8)		(46.6 ± 12.4)		(40.6 ± 12.4)	
(Age range)	(18–63)		(22–65)		(27–67)		(18–67)	
Sex								
Male	4	63%	8	50%	6	43%	18	43%
Female	8	67%	8	50%	8	57%	24	57%
T stage								
T^1	1	8%	1	6%	0	0%	2	5%
T^2	4	34%	3	19%	2	14%	9	21%
T^3	1	8%	0	0%	0	0%	1	2%
T^4	6	50%	12	75%	12	86%	30	72%
Grade								
I	5	42%	2	12%	3	21%	10	24%
II	3	25%	1	6%	2	14%	6	14%
III	4	33%	13	82%	9	65%	26	62%

CS = chondrosarcoma; OS = osteosarcoma; FS = fibrosarcoma; total = CS + OS + FS.

patient had received radiation therapy for an ameloblastoma of the upper alveolus, six years prior to the development of CS. A second patient received radiation treatment for a lymphoepithelioma of the nasopharynx five years prior to development of OS. The third patient developed OS, 20 years after irradiation of a recurrent fibro-osseous dysplasia of the maxilla. There were 10 (24 per cent) grade I tumours, six (14 per cent) grade II tumours and 26 (62 per cent) grade III tumours. The majority of OS (81 per cent) and FS (64 per cent) were of grade III whereas nearly half of CS were of low grade (Table I).

Of 42 patients, two (five per cent) had T_1 lesions, nine (21 per cent) had T_2 lesions, one (two per cent) had a T_3 lesion and 30 (72 per cent) had T_4 lesions (Table I). The site of origin was the maxillary sinus in the patients who had T_1 or T_2 lesions. Multiple sinuses were involved in those who had T_3 or T_4 lesions. No patient had regional or distant metastasis at the time of diagnosis.

Treatment

In principle, a radical surgery has been the treatment of choice if there was no contra-indication. Post-operative radiotherapy in a dose between 55 to 65 Gy was given only if the excision was incomplete macroscopically or in the case of positive surgical margins (microscopic). Radiotherapy or chemotherapy was employed alone, or in combination, for unresectable lesions. During recent years chemotherapy has been employed as a neoadjuvant treatment pre-operatively in patients with OS or FS. The chemotherapeutic agents used were methotrexate in high doses, adriamycin, cyclophosphomide, cisplatin, doxorubicin and vincristine.

The modality of treatment employed for the primary tumour is shown in Table II. Surgery was performed in 32 patients. The resection was considered to be 'complete' when there were no macroscopic remnants and microscopic surgical margins were negative. Excision was considered 'incomplete' if macroscopic tumour remnants were left *in situ* and/or when margins of excision were positive. Of 32 patients, 12 had complete and 20 incomplete excisions. Chemotherapy was employed pre-operatively as a neoadjuvant in seven patients with no response and four of them had positive surgical margins at excision. Post-operative radiotherapy was given to all patients in whom the excisions were incomplete except two who had had irradiation previously.

Radiotherapy was employed without surgery in eight patients; two of them received neoadjuvant chemotherapy with no tumour regression. Chemotherapy was employed alone with a palliative intent for unresectable OS in two patients.

Results

The overall three and five-year survival was 28 and 23 per cent respectively with a median survival of 24 months (mean 53 months). The median disease-free interval was nine months with a three and five-year disease-free survival of 27 and 20 per cent respectively. The survival at five years was 36 per cent in patients with CS, 24 per cent in patients with OS and 21 per cent in patients with FS ($p>0.1$; Table III).

The paediatric patients had a survival rate of 50 per cent at three years compared to 30 per cent survival rate in

TABLE II
TREATMENT

Type	CS (n = 12)	OS (n = 16)	FS (n = 14)	Total (n = 42)
Surgery	5 (1)	4 (1)	3 (1)	12 (3)
Radiotherapy	3	2 (2)	3	8 (2)
Surgery + radiotherapy	4	8 (2)	8 (2)	20 (4)
Chemotherapy alone	0	2	0	2

CS = chondrosarcoma; OS = osteosarcoma; FS = fibrosarcoma; total = CS + OS + FS; brackets () = number of patients in the corresponding subgroup who had neoadjuvant chemotherapy.

https://doi.org/10.1017/S0022215100128609 Published online by Cambridge University Press

TABLE III
FACTORS AFFECTING SURVIVAL

Factors	Overall survival rates				Difference in survival at 3 and 5 years (log–rank test)
	1 year	2 years	3 years	5 years	
Whole series	67%	42%	28%	23%	
Type					NS
Chondrosarcoma	83%	48%	48%	36%	
Osteosarcoma	60%	42%	24%	24%	
Fibrosarcoma	78%	42%	28%	21%	
Age					NS
Paediatric (≤ 15 years)	67%	50%	50%	17%	
Adult (≥ 15 years)	72%	42%	30%	27%	
Sex					p <0.01
Female	71%	54%	46%	37%	
Male	72%	33%	13%	13%	
Grade					p <0.05
I	90%	70%	70%	56%	
II	66%	33%	33%	33%	
III	68%	40%	21%	14%	
T stage					NS
$T_1 + T_2$	82%	64%	52%	42%	
$T_3 + T_4$	68%	39%	29%	23%	
Quality of resection					NS
Complete (no RT)	92%	65%	36%	24%	
Incomplete (+ RT)	70%	40%	36%	30%	
Surgery					p <0.01
Surgery ± RT	81%	52%	40%	33%	
No surgery (RT/CT)	50%	25%	12%	12%	
Local failure					p <0.01
Local failure present	66%	31%	14%	7%	
No local failure	90%	90%	80%	80%	

p <0.05 was considered statistically significant; NS = not statistically significant ($p \geq 0.05$); RT = radiotherapy; CT = chemotherapy; no RT = no radiotherapy; + RT = adjuvant radiotherapy.

adult patients (p >0.05; Table III). The disease-free survival at two and three years in paediatric patients was 80 and 50 per cent respectively. Only one child was alive and free of disease after surgical control of locally recurrent CS, whereas the remaining five children succumbed with local failure (Table IV).

Female patients had a significantly better survival rate at three years than males (50 *versus* nine per cent; p<0.01; Table III). Disease-related mortality occurred in 15 out of the 17 (83 per cent) male and 16 out of the 24 (67 per cent) female patients (Table IV). There was a significant difference in the mean disease-free survival rate of female and male patients (73 months in female *versus* 26 months in male patients; p = 0.03). Nine female patients were disease-free for five years or more and six of them were alive NED beyond 10 years. Of 18 male patients, only one survived beyond three years (NED at 15 years).

TABLE IV
DISEASE MORTALITY

Characteristics	CS	OS	FS	Total
Age				
≤ 15 years	0/1	3/3	2/2	5/6
> 15 years	7/11	10/13	9/12	26/36
Sex				
Male	3/4	8/8	4/6	15/18
Female	4/8	5/8	7/8	16/24
T stage				
T_1	0/1	1/1	0/0	1/2
T_2	2/4	3/3	0/2	5/9
T_3	1/1	0/0	0/0	1/1
T_4	4/6	9/12	11/12	24/30
Grade				
I	1/5	0/2	2/3	3/10
II	2/3	1/1	1/2	4/6
III	4/4	12/13	8/9	24/26
Treatment				
Surgery*	3/5	4/4	1/3	8/12
Surgery + RT*	1/4	5/8	8/8	14/20
Radiotherapy*	3/3	2/2	3/3	8/8
Chemotherapy	0/0	2/2	0/0	2/2

CS = chondrosarcoma; OS = osteosarcoma; FS = fibrosarcoma; total = CS + OS + FS; RT = adjuvant radiotherapy; *some of the patients (see Table II) had also neoadjuvant chemotherapy.

The patients with grade I tumours had a significantly better survival rate than those with grade III tumours (56 *versus* 14 per cent at five years; p <0.05; Table III). The patients with grade II tumours had intermediate survival rates (33 per cent at five years). The disease related mortality was at least 30 per cent with grade I and highest 80 per cent with grade III tumours (Table IV). Only two patients (eight per cent) with grade III tumours are currently alive and disease-free compared to half of those with tumours of other grades.

The influence of histological grade on survival was obvious in patients with CS. Four out of five (80 per cent) patients with grade I tumour (CS) were alive and free of

TABLE V
PATTERN OF FAILURE

Site	CS	OS	FS	Total
Local	6	12	10	28
Distant	1	1	1	3
Local + regional	0	1	1	2
Local + distant	2	0	0	2

CS = chondrosarcoma; OS = osteosarcoma; FS = fibrosarcoma; total = CS + OS + FS.

https://doi.org/10.1017/S0022215100128609 Published online by Cambridge University Press

TABLE VI
FACTORS AFFECTING LOCAL FAILURE

Factors	Local failure CS	OS	FS	Total	p value
Age					NS
≤ 15 years	1/1	3/3	2/2	6/6	
> 15 years	7/11	10/13	9/12	26/36	
Sex					$p = 0.016$
Male	4/4	8/8	5/6	17/18	
Female	4/8	5/8	6/8	15/24	
T stage					NS
T_1	1/1	1/1	0/0	2/2	
T_2	2/4	2/3	1/2	5/9	
T_3	1/1	0/0	0/0	1/1	
T_4	4/6	10/12	10/12	24/30	
Grade					$p = 0.02$
I	2/5	1/2	2/3	5/10	
II	2/3	1/1	1/2	4/6	
III	4/8	11/13	8/9	23/26	
Treatment[+]					NS
Surgery*	4/5	4/4	1/3	9/12	
Surgery + RT*	1/4	5/8	7/8	13/20	
Radiotherapy*	3/3	2/2	3/3	8/8	
Chemotherapy	0/0	2/2	0/0	2/2	

CS = chondrosarcoma; OS = osteosarcoma; FS = fibrosarcoma; total = CS + OS + FS. The factors were analysed in the total group by chi-square test; p value <0.05 was considered as statistically significant; NS = statistically nonsignificant ($p > 0.05$); [+] $p = 0.04$ when treatment was studied in two categories: surgery ± RT and RT or CT; *Some of the patients in the category had neoadjuvant chemotherapy.

disease at the last follow-up between two and 10 years (24, 40, 60 and 120 months respectively) and the fifth patient died of a local recurrence at five years. Three of the four patients (75 per cent) with grade III tumour (CS) died of the disease within two years after the initial treatment. There was no significant relationship between survival rate and tumour extent. The survival was 42 per cent at five years in patients with T_1 or T_2 tumours and 27 per cent in patients with T_3 or T_4 tumours ($p < 0.1$).

The quality of resection did not show significant correlation with survival ($p > 0.1$). The overall survival at five years in patients with complete resections was 26 per cent with a disease-free survival rate of 58 per cent at one year, 33 per cent at three years and 16 per cent at five years. The median disease-free interval in these patients was 13 months. The overall survival at five years in patients with incomplete resections was 35 per cent, with an estimated disease-free survival of 50, 34 and 27 per cent at one, three and five years respectively. The median disease-free interval following incomplete resections was 14 months.

The incidence of incomplete excisions was significantly influenced by the extent of the tumour (T stage). The T_1 and T_2 stages had 25 per cent incidence of incomplete excisions (positive margins) compared to 74 per cent incidence for T_3 and T_4 tumours ($p = 0.033$). In this study, only the patients with positive margins or macroscopically incomplete resections had adjuvant radiotherapy. There was a significant survival benefit of adjuvant radiotherapy for T_3 and T_4 tumours. The five-year survival in patients with T_3 and T_4 tumours was 30 per cent in the adjuvant radiotherapy group compared to 16 per cent in the surgery alone group ($p<0.05$). The two and three-year survival for patients with grade III tumours was 50 and zero per cent respectively in the surgery alone group and 50 and 33 per cent respectively in the adjuvant radiotherapy group ($p>0.05$).

TABLE VII
DISTANT METASTASIS

Site	Type	Grade	Survival (months)	Follow-up status
Scalp	CS	I	60	NED
Lung	CS	II	24	DM
Bones	CS	III	14	DM
Lung	OS	III	120	NED
Lung	FS	III	12	DM

CS = chondrosarcoma; OS = osteosarcoma; FS = fibrosarcoma; NED = no evidence of disease; DM = died of metastasis.

The survival rate was significantly better in patients who underwent surgery than those who had no surgery (40 *versus* 12.5 per cent at three years; $p < 0.01$; Table III). The overall survival for eight patients who received radiotherapy with or without neoadjuvant chemotherapy was 12.5 per cent at three and five years with a disease-free survival of 12 per cent at one year. Only one remained disease-free for 92 months following irradiation alone for a grade III OS, but subsequently died of local recurrence at 16 years. The two patients who received adjuvant chemotherapy prior to irradiation died of disease at 24 and 36 months respectively. The survival for the two patients who had chemotherapy exclusively with a palliative treatment for an unresectable primary was five and 10 months respectively. All these 10 patients (who had had no surgical treatment) died of the disease whereas one third of those who had surgery are currently alive and free of disease (Table IV).

In our series, a total of seven patients received adjuvant chemotherapy pre-operatively (Table II) with no tumour regression. The overall survival was 86 per cent at one year and 29 per cent at two years with a disease-free survival of 42 per cent at one year and 29 per cent at two years. The disease-free period and overall survival for the four patients who received adjuvant chemotherapy, surgery with incomplete excisions and post-operative radiotherapy was 50 per cent at two years. The median disease-free and overall survival rates were 24 and 30 months respectively. One of these four is currently alive and free of disease after a follow-up of 10 years but the other three died of local or locoregional disease at eight, 14 and 45 months respectively). The median disease-free period and overall survival rate in three patients who had had pre-operative chemotherapy and complete excisions,

TABLE VIII
CLINICAL STATUS AT LAST CONTACT

Status	CS	OS	FS	Total
Living NED	5 42%	3 19%	2 14%	10 24%
NF	3	2	2	7
LC	1	0	0	1
MC	1	1	0	2
DOD	7 58%	13 81%	11 86%	31 74%
LF	5	12	9	
L+R	0	1	1	
L+M	2	0	0	
M	0	0	1	
DIC	0	0	1	1 2%

NED = no evidence of disease; NF = no failure; LC = local failure controlled; MC = metastasis controlled.
DOD = died of disease; LF = local failure alone; L+R = locoregional failure; L+M = local failure and metastasis; M = metastasis alone.
DIC = NED but died of intercurrent cause.

https://doi.org/10.1017/S0022215100128609 Published online by Cambridge University Press

but no adjuvant radiotherapy was 10 and 20 months respectively. None of these three patients survived beyond two years (died of disease at 16, 20 and 20 months respectively).

The pattern of treatment failure in 42 patients is shown in Table V. Local failure was the major cause of treatment failure and occurred in 32 patients (76 per cent). The local failure significantly affected the overall survival ($p<0.01$; Table III). The overall survival at three and five years in patients with local failure was 14 and seven per cent respectively compared to 80 per cent in those who had no local failure.

The median time for local recurrence was seven months (range two to 92 months; mean 13 months). The pattern of local failure is shown in Table VI. The incidence of local failure was significantly higher in male patients ($p<0.01$) and in patients with grade III tumours ($p<0.02$). The recurrence rate was significantly less in patients who had surgery than in those who had radiotherapy and/or chemotherapy without surgery (69 *versus* 100 per cent; $p<0.04$). There was no significant correlation of histological type, tumour extension (T stage), adequacy of resections with the overall incidence of local recurrences. However, when the results of treatment modalities were studied among histological types, the local recurrence rate was noted to be less when the excisions were complete rather than incomplete for FS, even though the latter received postoperative radiotherapy (67 *versus* 87 per cent). In contrast, the recurrence rate was higher in patients who had complete excisions than in those who had incomplete resections and adjuvant radiotherapy for CS (80 *versus* 25 per cent) and OS (100 *versus* 62 per cent).

Twenty-six patients underwent attempts at salvage for a locally recurrent disease, including surgery, radiotherapy and chemotherapy, or a combination of these modalities. Local recurrence was controlled only in two patients whereas the remaining 28 died of the disease (Table V). Distant metastasis occurred in five patients (12 per cent) and two of them also had local recurrence (Tables V and VII). The site of the metastasis was the lung (three patients), bones (one patient) and scalp (one patient). The incidence of metastasis was more with CS (three out of 12; 25 per cent) rather than with FS (one out of 14; seven per cent) or OS (one out of 16; six per cent). The histological grade of the primary was grade III in three out of five cases (60 per cent; Table VII). The median time to metastasis was 10 months (range: seven to 36 months; mean 16 months). The median survival rate in these five patients was 24 months (range: 14–120 months; mean 46 months). Two patients were alive and disease-free with metastases surgically controlled whereas the remaining three succumbed to disease (Table VII).

Regional lymph node metastasis was associated with extensive local recurrence in two patients in whom the primary was a grade III (OS) in one and a grade II (FS) in the other (Table V). Both the patients died of uncontrolled locoregional failure within seven and eight months respectively.

The clinical status of the 42 patients at last contact (according to histological type) are shown in Table VIII. At last follow-up 10 patients were alive NED and seven of them (two patients CS, three patients OS and two patients FS) were aliive NED at 10 years. One patient died of another cause but without evidence of disease (NED) at 15 years. Thirty-one patients (74 per cent) died of disease; 26 (62 per cent) died of local failure alone, one (two per cent) died of metastasis alone, two (four per cent) died of locoregional recurrence and two (four per cent) died of relapses at both local and distant sites.

Discussion

Sarcomas of the head and neck area are rare and constitute 10 to 15 per cent of all sarcomas (Burkey *et al.*, 1990; Farhood *et al.*, 1990; Mark *et al.*, 1991a). Fibrosarcoma is the commonest type of soft tissue sarcoma in the head and neck region (Figueredo *et al.*, 1988) and involves paranasal sinuses more frequently (Mark *et al.*, 1991b). The bone sarcomas constitute approximately 20 per cent of head and neck sarcomas and were evenly divided between OS and CS (Wanebo *et al.*, 1992). Osteosarcomas are more common in the mandible than in the maxilla (Vener *et al.*, 1984) whereas CS frequently involve maxilla (El-Silimy *et al.*, 1987).

These tumours tend to occur more frequently during the third and fourth decade of life (Batsakis *et al.*, 1980). Fibrosarcomas commonly occur in the paediatric age group (Weber *et al.*, 1986) but primary nasal chondrosarcomas in children are extremely rare (Finn *et al.*, 1984; Lacovara *et al.*, 1992).

Previous local irradiation was reported to be most often associated with OS (Arlen *et al.*, 1971) and FS (Mindell *et al.*, 1977) and rarely with CS (Feintusch, 1973). In our series previous local irradiation was noted in seven per cent of patients; one developed CS following irradiation for ameloblastoma (Feun *et al.*, 1991) and the remaining two developed OS after irradiation for a recurrent fibrous dysplasia (Mark *et al.*, 1991a) and for a lymphoepithelioma respectively.

The overall survival rate for patients in our series was 42 per cent at two years, 28 per cent at three years and 21 per cent at five years. The five-year survival in patients with CS was 36 per cent, with OS was 28 per cent and with FS was 21 per cent. The survival rate has been reported to be influenced by the type of sarcoma (Weber *et al.*, 1986; Wanebo *et al.*, 1992), grade (Evans *et al.*, 1977; Finn *et al.*, 1984; Farhood *et al.*, 1990; Frankenthaler *et al.*, 1990), anatomical site (de Fries *et al.*, 1979) size (Weber *et al.*, 1986; Farhood *et al.*, 1990) and quality of resection (Farhood *et al.*, 1990; Frankenthaler *et al.*, 1990; Wanebo *et al.*, 1992). The sinonasal region has a poorer prognosis than the skin and soft tissue for two reasons: firstly the more indolent nature of presentation and secondly the greater difficulty in obtaining tumour-free margins (Frankenthaler *et al.*, 1990). The local recurrence rate was 76 per cent in our series and we found that sex ($p<0.01$), grade ($p<0.02$) and the modality of treatment ($p<0.04$) were the most important prognostic factors for local recurrences. In our series of patients, the survival was significantly affected by sex ($p<0.01$), grade ($p<0.05$) and local recurrences ($p<0.01$).

Surgery remains the main stay for the treatment of choice of these tumours. Wide local excision with radical margins was recommended (Conley *et al.*, 1967; Swain *et al.*, 1974). Despite radical surgery, Mark *et al.* (1991a) reported a recurrence in four out of five patients treated for OS in the head and neck region. Similarly we noted high

local recurrence rates when radical surgery was employed alone without radiotherapy for CS or OS. We used adjuvant radiotherapy only when excisions were incomplete and found a decrease in the recurrence rate in case of CS or OS, even though the resections were inadequate. The positive surgical margins and/or macroscopically incomplete excisions occurred most commonly with advanced stage lesions and adjuvant radiotherapy in these situations improved the survival rate significantly. However, adjuvant radiotherapy had no significant impact on grade III tumours.

Harwood *et al.* (1980) stated that CS were potentially radiocurable and recommended radiotherapy for unresectable disease or in cases of positive surgical margins and reported a complete remission with a local control for more than five years in three patients following irradiation with a curative intent for head and neck chondrosarcoma. In our study the results of irradiation as the sole modality were discouraging. Irradiation alone provided a long-term control in only one patient. However adjuvant radiotherapy following incomplete excisions provided a significant survival benefit for patients with advanced stage (T_3 or T_4) tumours ($p < 0.05$). The data from recent series (McKenna *et al.*, 1987; Mark *et al.*, 1991a) supports the role of aggressive multimodality therapy for the sarcomas of head and neck. In our series we noted a longer median disease-free interval for patients who received adjuvant chemotherapy, surgery (incomplete excisions) and post-operative radiotherapy than for those who had pre-operative chemotherapy and complete tumour resections but no radiotherapy (24 *versus* 10 months; $p > 0.05$).

The role of adjuvant chemotherapy in the treatment of OS comes from studies on extremities. The natural history of OS of extremities was well summarized by Friedman and Carter (1972). Although local control was assured by amputation, the majority of patients treated nonadjuvantly developed metastasis within a year of amputation. Because the major obstacle for cure of OS of the extremities is distant metastasis, adjuvant chemotherapy had been employed in the treatment of non-metastatic OS to increase the relapse-free survival by delaying the development of overt metastatic disease. Link *et al.* (1985) reported from a multi-institutional osteosarcoma study, on 114 patients eligible for randomization, the two year actuarial relapse-free survival of 56 per cent in those who received adjuvant chemotherapy compared to 18 per cent in the surgery alone group. The development of adjuvant chemotherapy also permitted a limb sparing surgery without compromising the survival or the risk of local recurrences (Eilber *et al.*, 1984; Simon *et al.*, 1986). Kalifa *et al.* (1993), from our Institut Gustave Roussy, reported a study on 76 paediatric patients treated by 'Rosen's T_{10} protocol' (Rosen *et al.*, 1982) for OS of limbs, who had a relapse-free survival of 67 per cent at seven years for 70 patients who received pre-operative chemotherapy which included seven courses of high doses of methotrexate, one course of a combination of bleomycin, actinomycin and cyclophosphomide and one course of adriamycin. Fifty-eight of the 70 patients had limb sparing surgery.

The pattern of failure of sarcoma of the nose and paranasal sinus region is different from that of the extremities. There is a high risk of local recurrences and distant metastasis is very rare. The biological behaviour of the tumour in this region is similar to that of carcinoma and death usually results from intracranial extension of a persistent or a recurrent tumour. Because it is difficult to carry out complete excisions in the nasal and paranasal sinus area, surgery alone frequently results in local recurrence. From the experience gained in the past with sarcomas in the extremities and the head and neck, aggressive multimodality regimen may be employed for the sarcomas of the nose and paranasal sinuses to obtain a long-term local control.

Conclusions

Chondrosarcoma, osteosarcoma and fibrosarcomas of the nasal cavity and paranasal sinuses are rare and the survival rate was very poor. Histological grade was the important predictive factor for local recurrences. The local failure rate was very high and distant metastasis was less frequent. The survival rate was significantly affected by sex, grade of the tumour and local recurrences. Surgery remains the main stay of treatment. Radical surgery with adjuvant radiotherapy and chemotherapy may provide long-term local control.

References

Arlen, M., Higinbotham, N. L., Huvos, A. G. (1971) Radiation-induced sarcoma of the bone. *Cancer* **28:** 1087–1099.

Batsakis, J. B., Solomon, A. R., Rice, D. H. (1980) The pathology of head and neck tumours: neoplasms of cartilage, bone and notochord. *Head and Neck Surgery* **3:** 43–57.

Burkey, B. B., Hoffman, H. T., Baker, S. R., Thornton, A. F., McClatchey (1990) Chondrosarcoma of the head and neck. *Laryngoscope* **100:** 1301–1305.

Conley, J., Stout, A. P., Healey, W. V. (1967) Clinico-pathologic analysis of eighty-four patients with an original diagnosis of fibrosarcoma of the head and neck. *American Journal of Surgery* **114:** 564–569.

de Fries, H. O., Perlin, E., Leibel, S. A. (1979) Treatment of osteogenic sarcoma of the mandible. *Archives of Otolaryngology* **105:** 358–359.

Eilber, F. R., Morton, D. L., Eckardt, J., Grant, T., Weisenburger, T. (1984) Limb salvage for skeletal soft tissue sarcomas: multidisciplinary preoperative therapy. *Cancer* **53:** 2579–2584.

El-Silimy, O. E., Harvey, L., Bradley, P. J. (1987) Chondrogenic neoplasms of nasal cavity. *Journal of Laryngology and Otology* **101:** 500–505.

Evans, H. L., Ayola, A. G., Romsdahl, M. M. (1977) Prognostic factors in chondrosarcoma of bone, a clinicopathologic analysis with emphasis on grading. *Cancer* **40:** 818–831.

Farhood, A. I., Hadju, S. I., Shiu, M. H., Strong, E. W. (1990) Soft tissue sarcomas of the head and neck in adults. *American Journal of Surgery* **160:** 365–369.

Feintusch, T. A. (1973) Chondrosarcoma arising in a previously irradiated fibrous dysplasia. *Cancer* **31:** 877–881.

Feun, L. G., Albores-Saavedra, J., Savaraj, N., Fla, M. (1991) Osteogenic sarcoma arising adjacent to a long-standing ameloblasoama. A case report. *Oral Surgery, Oral Medicine and Oral Pathology* **71:** 77–79.

Figueiredo, M. T. A., Marques, L. A., Campos-Filho, N. (1988) Soft tissue sarcomas of the head and neck in adults and children: experience at a single Institution and review of the literature. *International Journal of Cancer* **41:** 198–200.

Finn, D., Goepfert, H., Batsakis, J. (1984) Chondrosarcoma of the head and neck. *Laryngoscope* **94:** 1539–1544.

Frankenthaler, R., Ayala, A. G., Hartwick, R. W., Goepfert, H. (1990) Fibrosarcoma of the head and neck. *Laryngoscope* **100:** 799–802.

Friedman, M. A., Carter, S. K. (1972) The therapy of osteogenic sarcoma: current status and thoughts for the future. *Journal of Surgical Oncology* **4:** 482–510.

Freidman, A. M., Reinman, H. M., Woods, J. E. (1989) Soft-tissue sarcomas of the head and neck. *American Journal of Surgery* **158:** 367–372.

Fu, Y-S., Pezin, K. H. (1974) Nonepithelial tumours of the nasal cavity, paranasal sinuses, and nasopharynx: a clinicopathologic study. III. Cartilaginous Tumors (chondroma, chondrosarcoma). *Cancer* **34:** 453–463.

Harwood, A. R., Krajbich, J. I., Fornasier, V. L. (1980) Radiotherapy for chondrosarcoma of bone. *Cancer* **45:** 2769–2777.

Kaplan, E. L., Meijer, P. (1958) Non-parametric estimation from incomplete observations. *Journal of the American Statistical Association* **53:** 457–481.

Kalifa, C., Razafindrakoto, H., Vassal, G., Contesso, G., Vanel, D., Edeline, V., Valteau, D., Lemerle, J. (1993) Chemotherapy in osteogenic sarcoma: the experience of the paediatric Department of the Gustave Roussy Institute. In *Osteosarcoma in Adolescents and Young Adults* (Bennett-Humphry, G., ed.), Kluwer Academic Publishers, Boston, pp 347–349.

Lacovara, J., Patterson, K., Reaman, G. H. (1992) Primary nasal chondrosarcoma. The paediatric experience. *American Journal of Paediatric Hematology/Oncology* **14 (2):** 158–162.

Link, M., Goorin, A., Miser, A., Green, A., Pratt, C., Belasco, J., Pritchard, J., Baker, A., Kirkpatrick, J., Ayala, A., Shuster, J., Abelsone, H., Simone, J., Vietti, T. (1985) The role of adjuvant chemotherapy in the treatment of osteosarcoma of the extremity; preliminary results of the Multi-Institutional Osteosarcoma Study. *Proceedings of the American Society of Clinical Oncology* **4:** 237.

Malawer, M. M., Link, M. P., Donaldson, L. L. (1989) Sarcoma of bone. In *Cancer: Principles and Practice of Oncology* (Devita, V. T., Hellman, S., Rosenberg, S. A., eds). J. B. Lippincott, Philadelphia; 1442–1453.

Mandard, A. M., Petiot, J. F., Marnay, J., Mandard, J. C., Chasle, J., de Ranieri, E., Dupin, P., Herlin, P., de Ranieri, J., Tanguy, A., Boulier, N., Abbatucci, J. S. (1989) Prognostic factors in soft tissue sarcomas. A multivariate analysis of 109 cases. *Cancer* **63:** 1437–1451.

Mark, R. J., Sercarz, J. A., Tran, L., Dodd, L. G., Selch, M., Calcaterra, T. C. (1991a) Osteogenic sarcoma of the head and neck. The UCLA experience. *Archives of Otolaryngology, Head and Neck Surgery* **117:** 761–766.

Mark, R. J., Sercarz, J. A., Tran, L., Selch, M., Calcaterra, T. C. (1991b) Fibrosarcoma of the head and neck. The UCLA experience. *Archives of Otolaryngology, Head and Neck Surgery* **117:** 396–401.

McKenna, W. G., Barnes, M. M., Kinsella, T. J., Rosenberg, S. A., Lack, E. E., Glastein, E. (1987) Combined modality treatment of adult soft tissue sarcomas of the head and neck. *International Journal of Radiation Oncology, Biology and Physics* **13:** 1127–1133.

Mindell, E. R., Shols, N. K., Webster, J. H. (1977) Post-radiation sarcoma of bone and soft tissues. *Orthopaedic Clinics of North America* **8:** 821–834.

Peto, R., Pike, M. C. (1973) Conservatism in the approximation $\Sigma (O-E)^2/E$ in the log–rank test for survival data or tumor incidence data. *Biometrics* **29:** 579–584.

Rosen, G., Caparros, B., Huvos, A. G., Kosloff, C., Nirenberg, A., Cacavio, A., Marcove, R. C., Lane, J. M., Mehta, B., Urban, C. (1982) Preoperative chemotherapy for osteogenic sarcoma: selection of postoperative chemotherapy based on the response of the primary tumour to preoperative chemotherapy. *Cancer* **49:** 1221–1230.

Simon, M. A., Achliman, M. A., Thomas, N., Mankin, H. J. (1986) Limb salvage treatment *versus* amputation for osteosarcoma of the distal end of the femur. *Journal of Bone and Joint Surgery. American volume (Boston)* **68:** 1331–1337.

Swain, R. E., Sessions, D. G., Ogura, J. H. (1974) Fibrosarcoma of the head and neck: a clinical analysis of fifty cases. *Annals of Otolaryngology* **83:** 439–444.

UICC (1987) International Union against Cancer. In *TNM Classification of Malignant Tumours*, 4th Edition (Hermanek, P., Sobin, L. H., eds). Springer-Verlag, Heidelberg.

Vener, J., Rice, D. H., Newman, A. N. (1984). Osteosarcoma and chondrosarcoma of the head and neck. *Laryngoscope* **94:** 240–242.

Wanebo, H. J., Koness, J. R., MacFarlane, J. K., Elber, F. R., Byers, R. M., Elias, G., Spiro, R. H. (1992) Head and neck sarcoma: report of the head and neck sarcoma registry. *Head and Neck Surgery* **14:** 1–7.

Weber, R., Benjamin, R., Peters, L. (1986) Soft tissue sarcomas of the head and neck in adolescents and adults. *American Journal of Surgery* **152:** 386–392.

Address for correspondence:
Dr G. Schwaab,
Département de Chirurgie Cervico-Faciale,
Institut Gustave Roussy,
Rue Camille Desmoulins,
94805 Villejuif cedex,
Paris,
France.

Fax: (33 1) 45 59 63 76

https://doi.org/10.1017/S0022215100128609 Published online by Cambridge University Press