The Journal of Laryngology & Otology
July 2000, Vol. **114,** pp. 536–539

Mesenchymal chondrosarcoma of the maxilla

A. Chidambaram, F.R.C.S. Ed.(orl-hns), P. Sanville, M.R.C.P., F.R.C.R.*

Abstract
Mesenchymal chondrosarcoma (MC) is a rare tumour, with a predilection for the head and neck region. We describe a case of mesenchymal chondrosarcoma arising in the right maxilla extending to the basi-sphenoid. Its computed tomography (CT) and magnetic resonance imaging (MRI) and histopathological features and the management are presented. We also reviewed the literature of reported cases involving the maxilla.

Key words: Paranasal sinus neoplasms; Chondrosarcoma, mesenchymal; Radiology; Histology

Case report

A 14-year-old girl presented to the ENT clinic with right-sided nasal obstruction and snoring of 10 months duration. She had had one episode of nose bleeding three months prior to the consultation. There was no history of eye and ear symptoms. Clinical examination revealed marked obstruction in the right nasal cavity that prevented passage of a flexible nasendoscope. There was no swelling in the cheek or hard palate and no proptosis or middle-ear effusion. There was no cervical lymphadenopathy. Cranial nerve examination was normal.

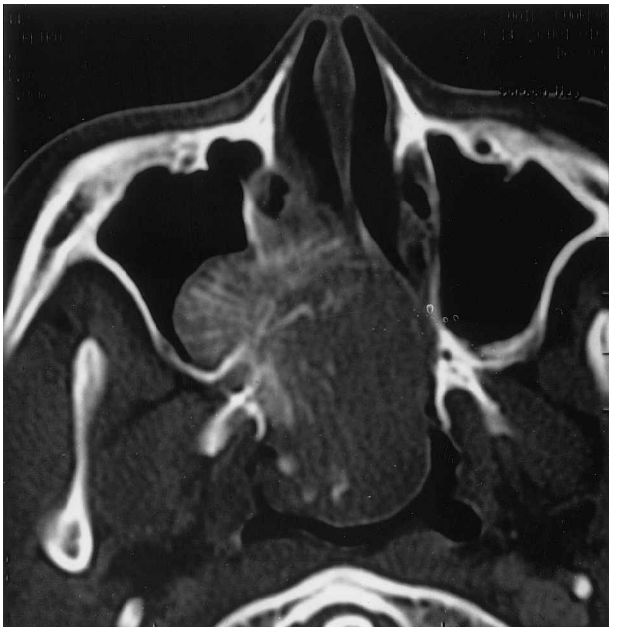

Fig. 1
Axial CT scan on bone settings showing the tumour with 'sun ray' spiculation arising from the medial wall of the right maxillary antrum.

High resolution coronal and post-contrast axial CT of the paranasal sinuses and MRI of the skull base were performed. CT revealed a soft tissue tumour measuring 7 cm × 5 cm with spiculated new bone formation arising from the medial wall of the posterior aspect of the right maxillary antrum (Figure 1). There was local bone destruction involving the ethmoid tracery, alveolar ridge, lateral aspect of the hard palate and posterior part of the nasal septum with the tumour crossing to the left side through the choana. MRI confirmed the CT findings. The tumour was of soft tissue density with destruction of bone and enhanced avidly but with some heterogeneity following intravenous contrast (Figure 2 and 3). There was no invasion of the cranial and orbital cavities or outside the lateral wall of the maxillary antrum.

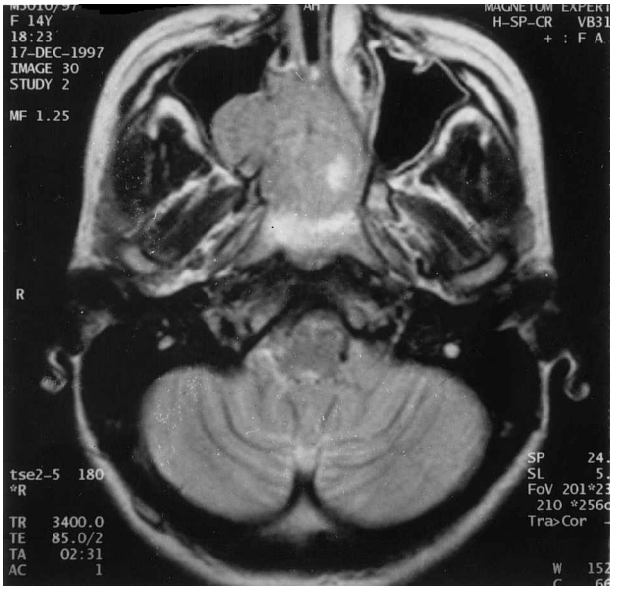

Fig. 2
Axial T_2 weighted MR scan showing heterogenous signal from the tumour.

From the Departments of Otolaryngology and Radiology*, Stepping Hill Hospital, Stockport, UK
Accepted for publication: 16 March 2000.

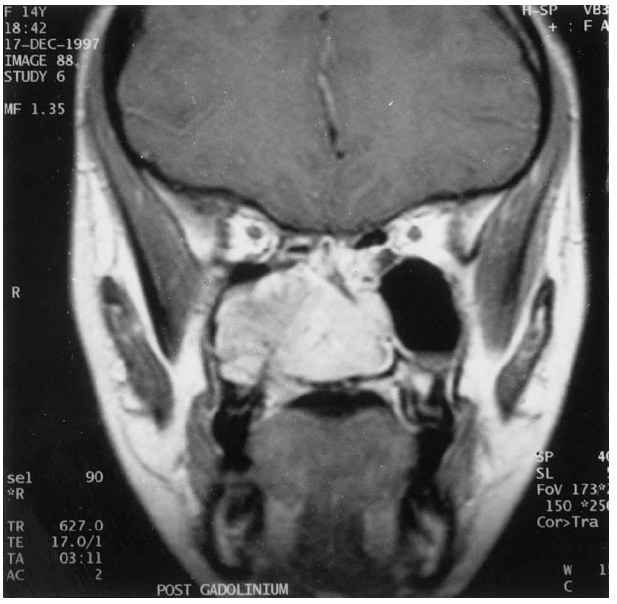

FIG. 3

Post-contrast coronal MR image showing superior boundary of the tumour.

On examination under general anaesthetic the lesion was found to be a firm to hard mass with a lobulated mucosal surface occupying the right nasal cavity and extending into the post-nasal space. Multiple biopsies were taken. The histopathologists reported the tumour as mesenchymal chondrosarcoma showing features of small monomorphic tumour cells and islands of immature cartilage (S100 protein positive) with blood vessels exhibiting a haemangiopericytoma-like pattern (Figures 4 and 5). The patient was referred to the Regional Paediatric Oncology unit, where metastasis was ruled out by CT scan of the lungs and an isotope bone scan. She underwent two cycles of pre-operative chemotherapy but there was no regression of the tumour size. The tumour was resected using a mid-facial degloving approach. A right partial medial maxillectomy and osteotomies through the posterior septum and the anterior wall of the sphenoid were performed and good macroscopic clearance was obtained. The post-operative period was uneventful. Radiotherapy has been reserved as a possible future option for any local recurrence and metastasis. She has been reviewed regularly and remains disease-free at 12 months after the surgical treatment.

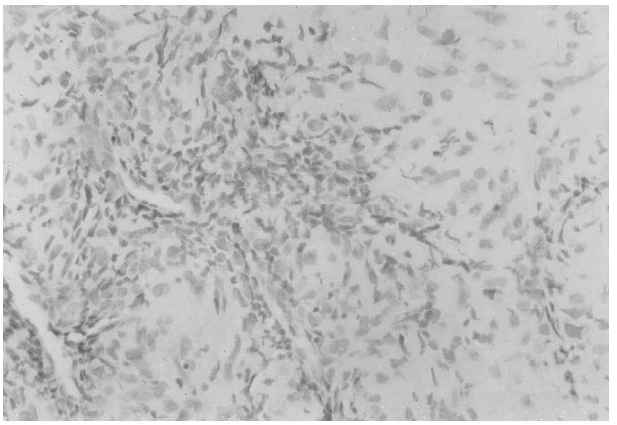

FIG. 4

Mesenchymal chondrosarcoma showing immature chondroid matrix and undifferentiated mesenchymal cells exhibiting haemangiopericytoma-like arrangement. (H&E; original magnification ×45).

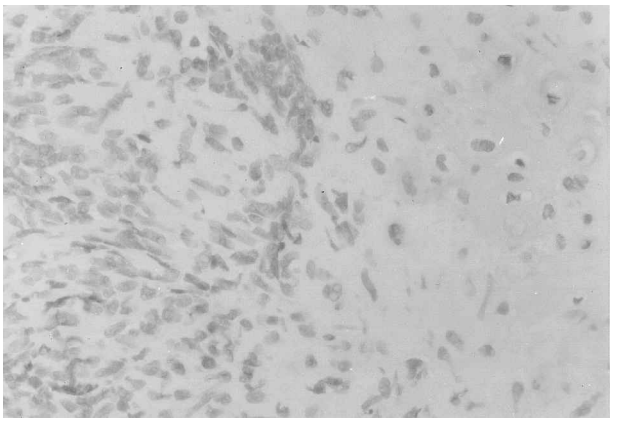

FIG. 5

Chondroid matrix (right) and mesenchymal cells (left) demonstrating clear transition zone. (H&E; original magnification ×250).

Discussion

Mesenchymal chondrosarcoma is a rare clinico-pathological entity first described by Lichtenstein and Bernstein.[1] These tumours arise both from skeletal and extraskeletal sites with a predilection for the head and neck region.[2] Maxilla and mandible are common sites of skeletal origin. Extraskeletal tumours commonly arise from the orbit and meninges.[3] Nasal and sinus mucosa,[4] parapharyngeal space[5] are among the other extraskeletal sites reported in the literature. Most patients are in their second and third decades when the diagnosis is first made.[2] The youngest age at presentation was a one-day-old infant[4] and the oldest was 68 years old.[6] The sex incidence is approximately equal.[7]

Table I summarizes the details of the cases of mesenchymal chondrosarcoma of maxilla reported in the world literature. Their presenting symptoms were swelling, pain, nasal blockage, epistaxis, loose teeth, diplopia and hard of hearing in the order of their frequency. The duration of the symptoms varied from one to 10 months. There was no metastasis at presentation. All patients underwent surgery and some of them had adjuvant chemotherapy and/or radiotherapy. Local recurrence occurred from 12 months to nine years after the initial surgical treatment. Metastases were noticed as late as 23 years after treatment and common sites were lung, bone and lymph nodes.

Imaging plays an important role in the management of these tumours. Conventional radiographs may show calcification and bony erosion. Cross sectional imaging with CT and MRI shows the soft tissue nature and extend of the lesion. Imaging features of mesenchymal chondrosarcoma, although non-specific, may reflect its dual histopathological morphological characteristics of differentiated cartilage islands interspersed within vascular undifferentiated mesenchyme.[8] The vascular anatomy is best demonstrated by angiography[5] that may be useful when radical surgery is considered. CT and MRI are also useful in the follow up. When disseminated bone metastases are suspected or the presenting tumour occurs in an unusual soft tissue location an isotope bone scan should be considered.[5]

Histology of these tumours show characteristic bimorphic pattern. Regardless of the site of origin, histological features are consistent from case to case.[9] It

https://doi.org/10.1258/0022215001906084 Published online by Cambridge University Press

TABLE I
SUMMARY OF MESENCHYMAL CHONDROSARCOMA OF THE MAXILLA REPORTED IN THE LITERATURE

Study	No.	Age (yrs)/ Sex	Site	Duration (months)	Treatment	Recurrence	Metastasis*	Outcome
Present study	1	14/F	Right maxilla	10	Maxillectomy + CH	No	No	Alive: 12 months, NED
Vencio *et al.*[12]	2	9/M	Maxilla	1	Maxillectomy	No	No	Dead: 6 years
	3	26/M	Maxilla	6	Maxillectomy + CH	Yes, 1 year	Yes (lung, T11) 10 years	Dead: 13 years
	4	27/F	Maxilla	?	Maxillectomy + CH + RXT	Yes, 4 years	Yes (cervical lymph nodes) 4 years	Dead: 5 years
	5	35/F	Maxilla	?	Maxillectomy	No	No	No follow-up
	6	20/M	Maxilla	?	No information	?	?	No follow-up
Mateos *et al.*[13]	7	32/F	Left maxilla	48	Maxillectomy	No	No	Alive: 9 years, NED
Crawford *et al.*[14]	8	9/F	Left maxilla	6	Maxillectomy + CH	No	No	Alive: 2 years, NED
Bottrill *et al.*[7]	9	15/F	Right maxilla	10	Maxillectomy + CH + RXT	No	No	Dead: 10 months
Riethdorf *et al.*[15]	10	26/M	Rigth maxilla	?	Maxillectomy + RXT Orbital exenteration	Yes, 1 year	No	Dead: 18 months
Ito *et al.*[16]	11	62/M	Right maxilla	?	Maxillectomy + CH	Yes (orbital)	Yes (multiple sites)	Dead: 47 months
Salmo *et al.*[6]	12	68/M	Premaxilla	1	Excision	?	?	No follow-up
Bloch *et al.*[3]	13	18/M	Right maxilla	2	Maxillectomy + RXT Orbital exenteration	No	No	Alive: 7 years, NED
	14	24/F	Left maxilla	?	Maxillectomy + RXT	No	Yes (axillary lymph nodes) 15 years	Not stated
Mikata *et al.*[17]	15	30/M	Left maxilla	4	Maxillectomy + CH + RXT	No	No	Alive: 18 months, NED
Salvador *et al.*[2]	16	46/F	Maxilla	?	Maxillectomy	Yes, 9 years	Yes (retroperitoneum) 8.5 years	Dead: 9.5 years
Dahlin and Henderson[18]	17	30/M	Maxilla	7	Maxillectomy + RXT	No	No	Dead: 6 years
	18	23/F	Maxilla	9	Curettage + RXT	Yes, 3 years	Yes (lung, breast) 22 years (pelvis, vulva, scalp) 23 years	Dead: 23 years

M = male; F = female; CH = chemotherapy; RXT = radiotherapy; NED = no evidence of disease.
*No case had metastasis at presentation.

is composed of variable amounts of well differentiated or less mature cartilage with masses of undifferentiated small, round or bluntly fusiform mesenchymal cells. The presence of a haemangiopericytoid vascular pattern is a well recognized feature.[2,9] Immunohistochemical studies have demonstrated that the immunophenotype of mesenchymal chondrosarcoma resembles that of embryonic cartilage and may indicate that this tumour is an embryonic cartilage analogue.[9] In the absence of chondroid areas in the biopsy specimens, that may be due to superficial or inadequate sampling, its separation from Ewing's sarcoma, 'solid' rhabdomyosarcoma, haemangiopericytoma or synovial sarcoma may be problematic.[9]

Radical surgery is the mainstay of treatment. Radiotherapy or chemotherapy (or both) may be indicated for lesions not amenable to ablative surgical treatment.[10] These treatment modalities may also be used as adjuvant to primary surgical treatment and also for recurrence and metastasis. Prognosis for mesenchymal chondrosarcoma is poor and the clinical course may be protracted. Huvos *et al.*[11] found a 10-year survival rate of 28 per cent in their series of 35 patients. Local recurrence and distant metastases can occur many years after treatment making long-term follow-up essential.

Conclusion

We report a case of skeletal mesenchymal chondrosarcoma arising from the maxilla which is a rare malignant tumour and present the imaging and histopathological features that distinguish it from common chondrosarcoma. The literature review of 18 cases involving the maxilla is reported. This rare tumour has a poor prognosis. It may lead a protracted course and long-term follow-up is essential.

Acknowledgements

We would like to thank Mr N. J. Kay, Dr H. R. Gattamaneni and Mr A. D. Cheesman for allowing us to report a case under their joint care. We also would like to thank Dr D. Vickers and Dr S. Banerjee for their help with the histopathological interpretation, and Dr K. S. Vasudev for the photomicrographs.

References

1 Lichtenstein I, Bernstein D. Unusual and malignant chondroid tumours of bone. *Cancer* 1959;**12**:1142–57
2 Salvador AH, Beabout JW, Dahlin DC. Mesenchymal chondrosarcoma – observation of 30 news cases. *Cancer* 1971;**28**:605–15
3 Bloch D, Bragoli A, Collins D, Batsakis J. Mesenchymal chondrosarcoma of the head and neck. *J Laryngol Otol* 1979;**93**:405–12
4 Roland NJ, Khine MM, Clarke R, Van Velzen D. A rare congenital intranasal polyp: mesenchymal chondrosarcoma of the nasal region. *J Laryngol Otol* 1992;**106**:1081–3
5 Gomersall LN, Needham G. Case report: Mesenchymal chondrosarcoma occurring in the parapharyngeal space. *Clin Radiol* 1990;**42**:359–61
6 Salmo NA, Shukur ST, Abulkhail A. Mesenchymal chondrosarcoma of the maxilla: Report of a case. *J Oral Maxillofac Surg* 1988;**46**:887–9

7 Bottrill ID, Wood S, Barrett-Lee P, Howard DJ. Mesenchymal chondrosarcoma of the maxilla. *J Laryngol Otol* 1994;**108**:785–7
8 Shapeero LG, Vanel D, Couanet D, Contesso G, Ackerman LV. Extraskeletal mesenchymal chondrosarcoma. *Radiology* 1993;**186**:819–26
9 Swanson P, Lillemoe T, Manivel C, Wick M. Mesenchymal chondrosarcoma: an immunohistochemical study. *Arch Pathol Laboratory Med* 1990;**114**:943–8
10 Nakashima Y, Unni KK, Shives T, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue. A review of 111 cases. *Cancer* 1986;**57**:2444–53
11 Huvos AG, Rosen G, Dabska M, Marcove RC. Mesenchymal chondrosarcoma: A clinicopathologic analysis of 35 patients with emphasis on treatment. *Cancer* 1983;**51**:1230–37
12 Vencio EF, Reeve CM, Unni KK, Nascimento AG. Mesenchymal chondrosarcoma of the jaw bones. Clinicopathologic study of 19 cases. *Cancer* 1998;**82**:2350–5
13 Mateos M, Forteza G, Gay-Escoda C. Mesenchymal chondrosarcoma of the maxilla. A case report. *Int J Oral Maxillofac Surg* 1997;**26**:210–1
14 Crawford JG, Oda D, Egbert M, Myall R. Mesenchymal chondrosarcoma of the maxilla in a child. *J Oral Maxillofac Surg* 1995;**53**:938–41
15 Riethdorf L, Lorenz G, Werner E, Karnitzki G. Mesenchymales Chondrosarkom des Oberkiefers. *Laryngorhinootologie* 1993;**72**:621–3
16 Ito T, Hiratsuka H, Kohama G. Mesenchymal chondrosarcoma of the maxilla. Report of a case. *Int J Oral Maxillofac Surg* 1991;**20**:44–5
17 Mikata A, Iri H, Inuyama Y. Mesenchymal chondrosarcoma – a case report with an ultrastructural study and review of Japanese literature. *Acta Patholog Jap* 1977; **27**:93–109
18 Dahlin DC, Henderson ED. Mesenchymal chondrosarcoma: further observations on a new entity. *Cancer* 1962;**15**:410–7

Address for correspondence:
Mr A. Chidambaram,
9 Alder Court,
Sutton-in-Ashfield NG17 4NZ, UK.

E-mail: achidam@usa.net

Mr A. Chidambaram takes responsibility for the integrity of the content of the paper
Competing interests: None declared