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Mesenchymal chondrosarcoma of the maxilla

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Abstract
We report, to our knowledge, the 10th recorded case of mesenchymal chondrosarcoma (MC) occurring in the maxilla. Our case is the youngest person reported with a tumour in this location. The prognosis for cure is poor with a high incidence of local recurrence as well as metastases. Treatment is based on radical surgery. Radiotherapy and chemotherapy have a adjuvant role but additional experience with this tumour is required to define the most efficacious treatment.

Key words: Paranasal sinus neoplasms; Surgery; Radiotherapy; Paediatrics

Case report

A 15-year-old girl presented to our clinic with a 10-month history of bilateral nasal obstruction. In addition she had noticed a watery rhinorrhoea and slight epistaxis. She had no significant past medical history.

On examination she looked well, there was no evidence of neck lymphadenopathy. Nasal examination revealed copious

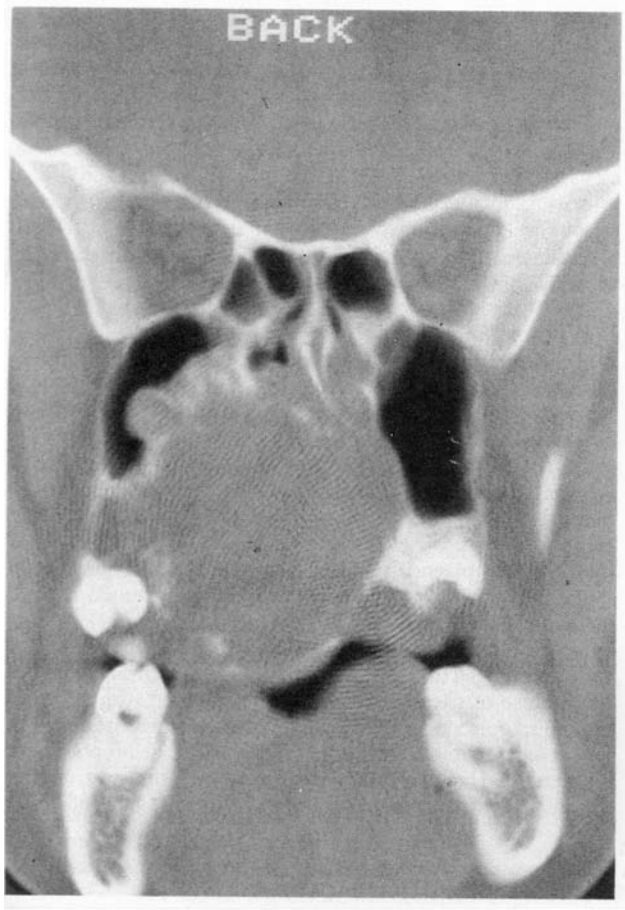

FIG. 1
Coronal CT scan of the head showing an extensive tumour occupying the nasal cavity and right antrum. Note the bony erosion of the hard palate and speckled calcification.

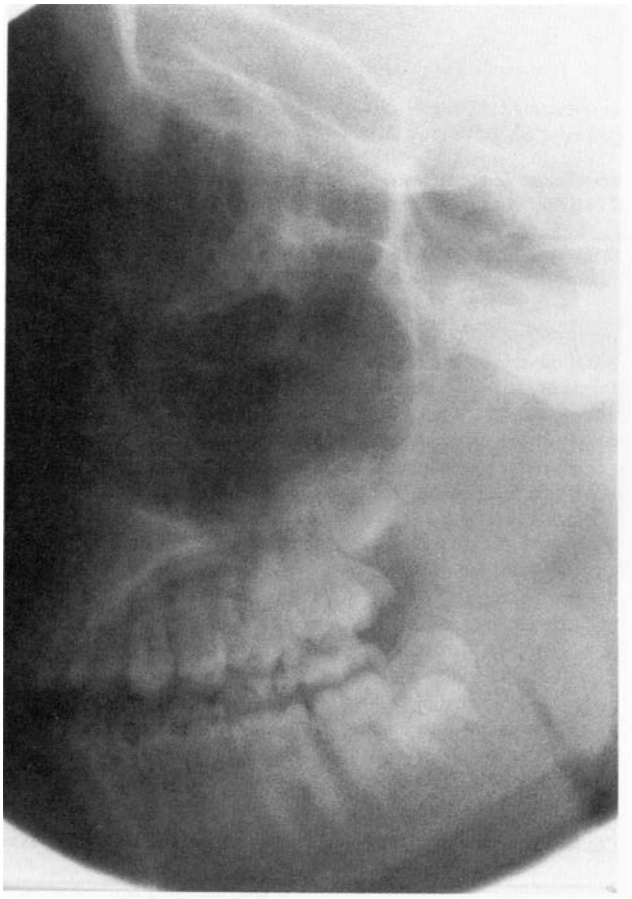

FIG. 2
Lateral X-ray of the facial skeleton showing a radiolucent region in the hard palate.

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TABLE I
RECORDED CASES OF MESENCHYMAL CHONDROSARCOMA (MC) OF THE UPPER JAW

Reference	Age (years)	Sex	Presenting symptoms	Duration[a]	Treatment	Status[b]
Bottrill *et al.* (1994)	15	F	Nasal Blockage Epistaxis	10/12	Maxillectomy Radiotherapy Chemotherapy	Dead: 10 months
Hiratsuka and Kohama (1991)	62	M	Swelling	?	Maxillectomy Orbital recurrence; Chemotherapy	Dead: 47 months
Salmo *et al.* (1988)	68	M	Swelling Pain	1	'Excision'	Patient lost to follow-up
Bloch *et al.* (1979)	18	M	Swelling Loose teeth	2	Radiotherapy – poor response Maxillectomy Orbital exenteration	Alive: disease-free at 7 years
	24	F	Nasal Blockage Epistaxis	7	Maxillectomy Radiotherapy Local recurrence at 15 years	Not stated
Salvador *et al.* (1971)	46	F	Swelling	?	Maxillectomy Retroperitonial metastases at 8.5 years Local recurrence at 9 years	Dead: 9.5 years
Dahlin and Henderson (1962)	30	M	Swelling	7	Maxillectomy Radium therapy	Dead: 6 years 'of cancer'
	23	F	Swelling	9	Curettage + radium Local recurrence at 3 years Roentgen therapy Lung metastases at 22 years Right pneumonectomy Breast metastases at 22 years Vulva, pelvis and scalp metastases at 23 years	Dead: 23 years

[a]The duration the symptoms were present prior to the first consultation (years).
[b]The time quoted is the duration of survival from first presentation.

quantities of mucus in both nasal cavities and a mass in the right nasal cavity. Intraorally there was a large mass occupying two thirds of the hard palate. It had a lobulated appearance, was firm and nontender. The overlying mucosa was normal and the cranial nerves were intact.

A CT scan was carried out (Figure 1) showing a large mass in the lower anterior part of the right antrum which appeared to be arising from the upper alveolus which was extensively eroded. The mass contained considerable peripheral calcification but showed minimal enhancement. It extended downwards into the mouth and across the midline into the left nasal cavity. The appearance suggested an odontogenic type of tumour.

The patient was admitted for further investigations. All routine haematological and biochemical investigations and a chest X-ray were normal. A biopsy was performed and the tumour was reported as a haemangiopericytoma (HPC). She therefore underwent a right maxillectomy via a mid-facial degloving approach. The tumour had breached the posterior wall of the maxilla and was present in the pterygopalatine fissure on the right. Post-operatively she made a rapid recovery. The diagnosis was revised on histological assessment of the operative specimen to mesenchymal chondrosarcoma.

In view of the extent of the disease she was given a course of post-operative radical radiotherapy and combination chemotherapy. The radiotherapy was given to the right side of the face using a wedged pair of fields to a total dose of 60 Gy in seven weeks. It was planned to give her six courses of cisplatin (100 mg/m^2: one four hourly infusion) and adriamycin (25 mg/m^2: three doses intravenously over three days) at three weekly intervals. She required several admissions for neutropaenic sepsis which prolonged the interval between courses. Ten days after her fifth course of chemotherapy she was admitted with a further prolonged episode of neutropaenic sepsis. She was treated with intravenous antibiotics and after seven days the neutrophil count recovered and her clinical condition stabilized. However, despite a normal neutrophil count, she became pyrexial again and blood

TABLE II
CLINICAL COURSE OF PATIENTS WITH MESENCHYMAL CHONDROSARCOMA (MC) AFTER VARYING TREATMENT MODALITIES (AFTER NAKASHIMA *ET AL.*, 1986)

No. of patients	Primary modality	No. with recurrence[a]	No. alive[a]	No. dead from MC[a]
41	Intralesional excision	33 (2.9 years)	12 (5.4 years)	27 (5.5 years)
22	Wide local excision	9 (3.6 years)	16 (7.8 years)	6 (5.9 years)
7	Primary radiotherapy and/or chemotherapy	7	0	6 (2.2 years)
37	Surgery and radiotherapy and/or chemotherapy	?	?	30

[a]Numbers in brackets refer to the mean duration of survival from initial treatment.

cultures grew *Candida spp.* Antifungal therapy was started but unfortunately her condition rapidly deteriorated despite vigorous cardiovascular and respiratory support and she died three weeks following her last admission. She had survived 10 months after her initial presentation. A postmortem was performed which confirmed septicaemia and adult respiratory distress syndrome as the cause of death but showed no evidence of local or distant recurrence.

Discussion

Mesenchymal chondrosarcoma is a very rare but characteristic malignant tumour first described by Lichtenstein and Bernstein (1959). Mesenchymal chondrosarcoma can occur in both osseous and extra-osseous sites in a ratio of 2:1 (Nakashima *et al.*, 1986). The total number of tumours recorded in the world literature for all sites was 51 by 1971 (Salvador *et al.*, 1971); more recent reviews report numbers in excess of 160 (Williams *et al.*, 1987). In the head and neck region the mandible is the commonest site of bony origin for the tumour. However there have only been nine cases reported arising from the maxilla; seven in a review of the literature by Nakashima *et al.* (1986) and two further case reports, one by Hiratsuka and Kohama (1991) and the other by Salmo *et al.* (1988).

Mesenchymal chondrosarcoma (MC) of the jaws has the highest incidence in the second and third decades. The sex incidence is approximately equal.

Radiographically the lesions show up as areas of radiolucency which may be well or poorly demarcated (Figure 2). Approximately 50 per cent may show diffuse calcification (Huvos *et al.*, 1983). Macroscopically these tumours are soft to firm and grey–white to red/tan and may contain calcific deposits. As a consequence of this a gritty texture may be noted.

The histopathological pattern of these tumours demonstrates a bimorphic pattern composed of abundant sheets or clusters of highly undifferentiated mesenchymal cells, with prominent nuclei and scanty cytoplasm, interspersed with small islands of well differentiated cartilaginous tissue in which calcification and areas of metaplastic bone formation may be observed. The histological picture of these tumours may be confused at first sight with haemangiopericytoma (HPC), especially if a biopsy is taken that does not show areas of cartilage formation as in this case and in other cases reported in the literature (Bloch *et al.*, 1979; Fu and Perzin, 1974). Immunohistochemical studies have demonstrated that the mesenchyme resembles that of embryonic cartilage (Swanson *et al.*, 1990). Histological grading is stated to be of little clinical significance and all mesenchymal chondrosarcoma tumours are considered to be high grade. If only the cellular area is included in the biopsy then the differential lies between MC, HPC, solid rhabdomyosarcoma and synovial sarcoma.

Table I shows data recorded from mesenchymal chondrosarcoma arising in the maxilla. The predominant presenting symptom is swelling, more rarely pain and epistaxis may occur. Neurological compression may be the presenting symptom in tumours arising from other sites. In their review, Nakashima *et al.* (1986) reported that the symptoms had been present from four days to seven years at the first consultation and three out of 111 patients had metastases at presentation.

Treatment for this condition is based on experience with relatively few cases and no one centre has sufficient experience to produce any form of scientific proposal. The basis of the treatment is one of initial radical surgery with, or without, adjuvant radiotherapy and/or chemotherapy. There is little evidence that the later improves the prognosis significantly. Nakashima *et al.* (1986) reviewed the outcome of patients treated in each of four broad groups. The numbers in each do not allow statistical analysis and the patients have a variety of tumour locations and 'stages'. Despite these flaws Table II demonstrates that radiotherapy and/or chemotherapy alone appear to be of little benefit. In addition there is no convincing advantage to be gained using any of the other treatment modalities.

Patients may succumb relatively quickly from their disease or may remain well for many years. Of 78 patients with adequate follow-up data the duration from primary treatment to first metastasis varied from 0 to 22 years with the lung being the most frequent site. However local recurrence usually occurred before distant metastases (Nakashima *et al.*, 1986).

The prognosis for mesenchymal chondrosarcoma is poor, but the clinical course may be protracted. Huvos *et al.* (1983) found a five-year survival rate of 42 per cent and a 10-year one of 28 per cent. Therefore prolonged follow-up is required.

Conclusion

In view of the rarity of this condition it would be valuable to collate data in one central registry. This would encourage continuous follow-up and monitoring of treatment protocols and might also allow for unified trials of therapy.

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