The Journal of Laryngology and Otology
February 1988. Vol. **102.** pp. 177–181

Maxillary chondrosarcoma
(Report of two cases)

GIOVANNINO MASSARELLI,* LEONARDO GANDOLFO,† FRANCESCO TANDA,* FRANCO GHISELLI† and VINCENZO MANUNTA* (Sassari, Italy)

Abstract

Two cases of chondrosarcoma of the maxilla are reported together with the clinical and histological differential diagnosis. The origin of this rare tumour in the maxilla, a bone with exclusive membranous ossification, is discussed in accordance with the new concept of cell differentiation in neoplasia.

Introduction

Chondrosarcoma is a malignant tumour of soft tissues showing cartilaginous differentiation. The most frequent localizations are in the pelvis, ribs, and long bones (Spjut *et al.*, 1971). In the head and neck region its incidence has been estimated at about 10 per cent of all chondrosarcomas (Jones, 1973); a maxillary localization is particularly uncommon, amounting only to 0.7 per cent (Smatt, 1977). Up to 1974, Fu and Perzin (1974) had found only 25 well-documented cases in the literature; 40 cases were reported by Smatt in 1977. The unusual occurrence of chondrosarcoma in the maxilla is still under debate.

Report of cases

Case 1

A 32-year-old man was admitted to the ENT Department in 1965, complaining for one year of a painless swelling of the upper lip, just below the nostrils. The tumour mass was firm in consistency and extended to the left ascending ramus of the upper jaw. A nodular bulge was also present along the hard palate, in the midline. X-rays showed erosion of the base of the septum and of the floor of the left nasal fossa by a translucent mass containing scattered foci of calcification. A tumour with chondromatous features was resected and the histological diagnosis was that of chondroma.

One year later, the tumour recurred with clinical and radiographic patterns similar to the previous ones. The patient underwent further surgical excision and histological examination revealed a tumour formed by cartilaginous cells with plump nuclei. A suspicion of malignancy was raised.

Three years later, a second recurrence took place. A left subtotal maxillectomy, extending to the septum and to the medial portion of the right maxilla, was performed. The histological diagnosis was: poorly differentiated chondrosarcoma. The patients was lost to follow-up 12 years later without further recurrence.

Case 2

A 35-year-old man was admitted to the ENT Department in 1984, complaining of epiphora and a hard swelling of the left cheek of three months' duration. A round mass, 3–4 cm. in diameter, arising from the anterior wall of the left maxillary sinus, involved the lateral wall of the left nasal fossa, dislocating the inferior turbinate medially. X-rays and CT-scans showed a cloudy left maxillary sinus, containing disseminated islets of calcification; areas of erosion were also present in the medial and anterior walls of the sinus (Fig. 1 A–B). A transvestibular biopsy was performed. Histological examination revealed chondromatous tissue arranged in nodular masses, intermingled with fibroblastic proliferation, suggesting a chondromyxoid fibroma.

The patient underwent subtotal left maxillectomy and the microscopic picture of the specimen showed chondromatous proliferation with formation of nodules. The nuclei of the cartilaginous cells were plump, with prominent nucleoli and few mitoses. The diagnosis was chondrosarcoma, grade II according to Evans *et al.*, 1977. The patient is free of recurrence two years after surgery.

Pathological Findings

Case 1

The sections show immature cartilaginous cells embedded in a chondromatous matrix, rich in acid mucopolysaccharides. The cellular nuclei are irregular and many of these show prominent mitoses (Fig. 2A–B).

Case 2

Biopsy specimen: The tumour is surrounded by a pseudocapsule and is composed of cartilaginous cells arranged haphazardly or in continuous sheets. The presence of trabecular bone suggests endochondral ossification. At high power, cartilaginous cells display nuclear hyperchromatism with a few mitoses.

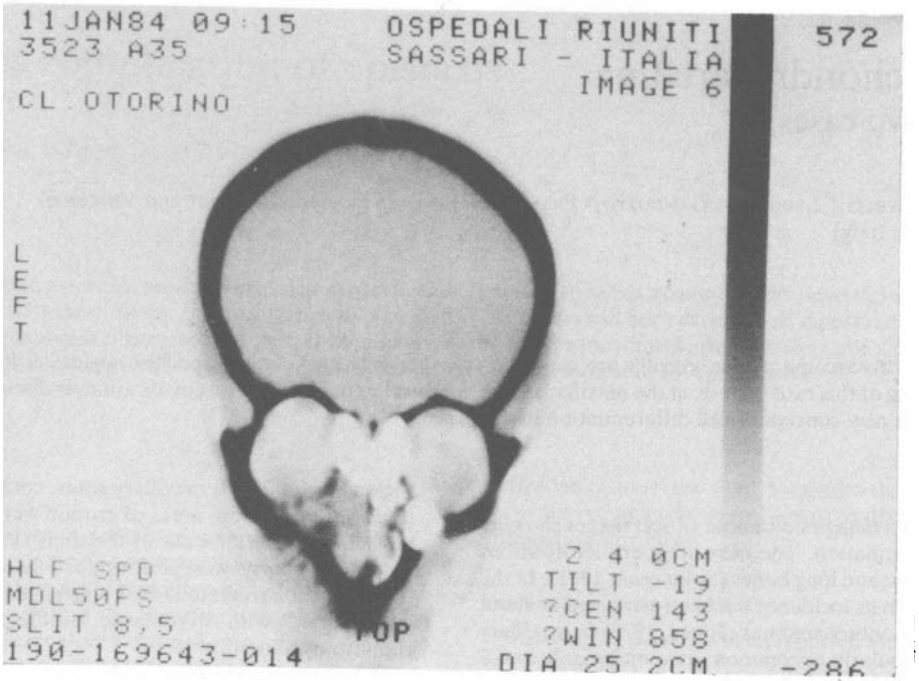
Surgical specimen: The tumour is formed by small nests of cartilaginous tissue surrounded by a fibrous capsule; the cartilaginous cells are intermingled with spindle-shaped fibroblast-like cells, and intermediate degrees of differentiation are noted up to unicellular calcification (Fig. 3 A–B).

Discussion

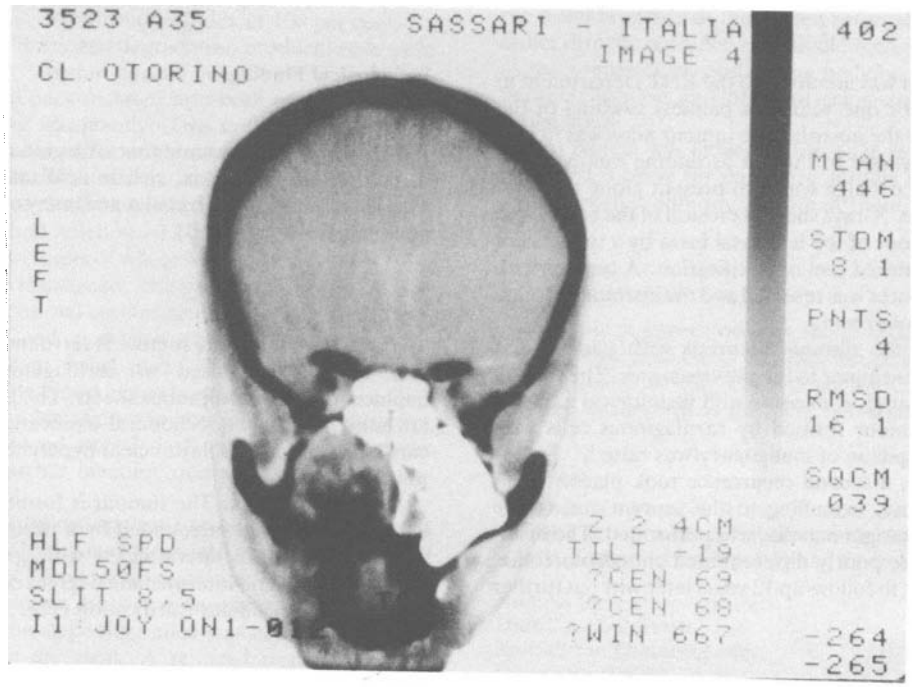
Chondrosarcoma has been considered to be a malignant tumour histogenetically derived from mature cartilaginous tissue (Jaffe, 1958); at present it is interpreted rather as a connective tissue tumour with morphologic differentiation towards a chondromatous pattern (Brocheriou and Payen, 1975). Such a view seems in better agreement with current

*Institute of Morbid Anatomy and Histopathology, †ENT Department, University of Sassari, Italy.

https://doi.org/10.1017/S0022215100104451 Published online by Cambridge University Press



A

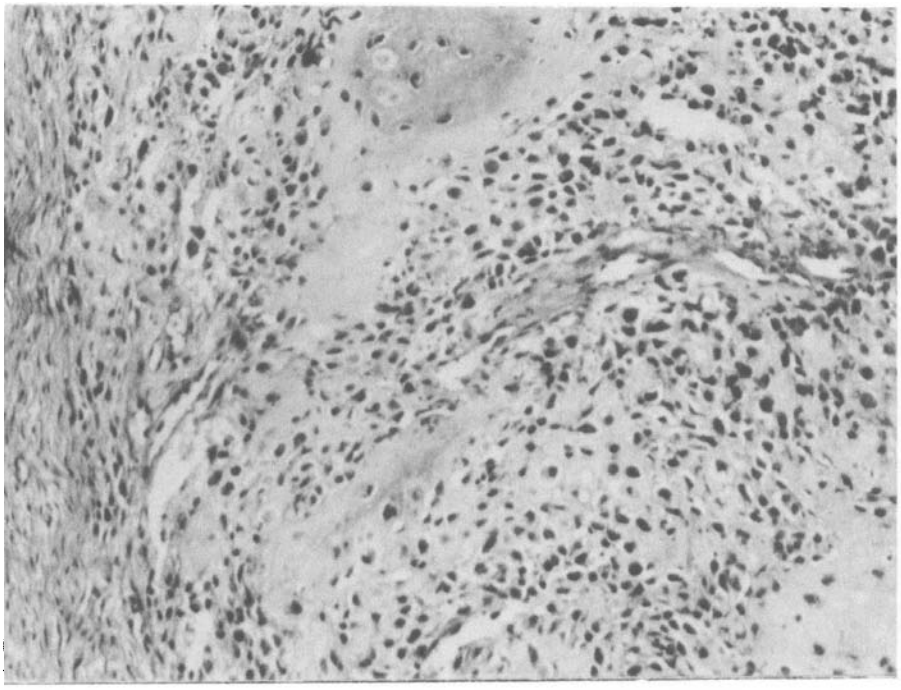


B

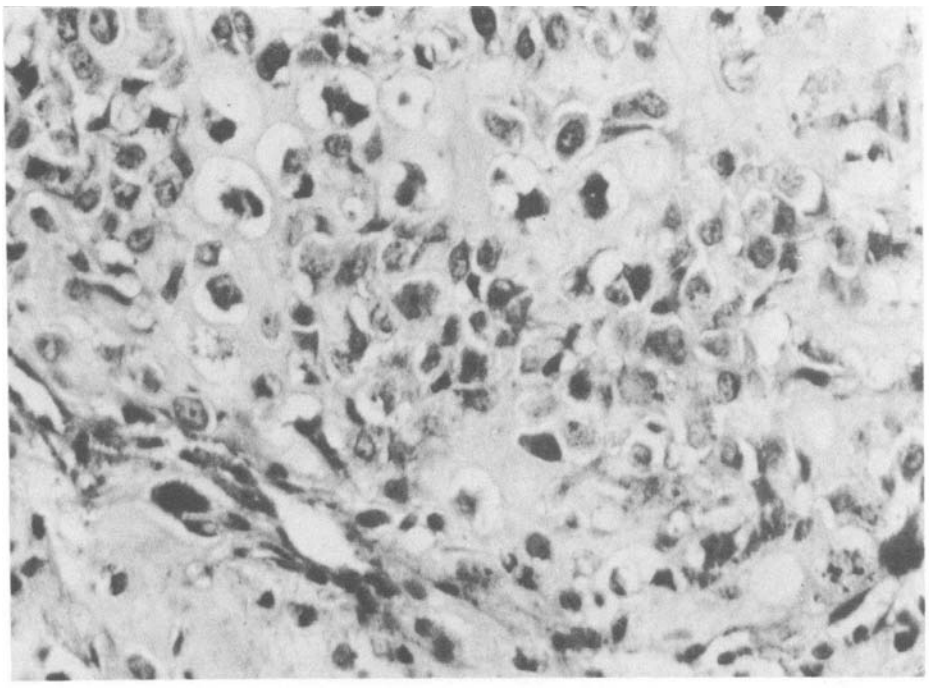
FIG. 1

Case 2. The CT-scan reveals a large mass involving the left nasal fossa and maxillary sinus and dislocating the inferior turbinate medially.

https://doi.org/10.1017/S0022215100104451 Published online by Cambridge University Press



A



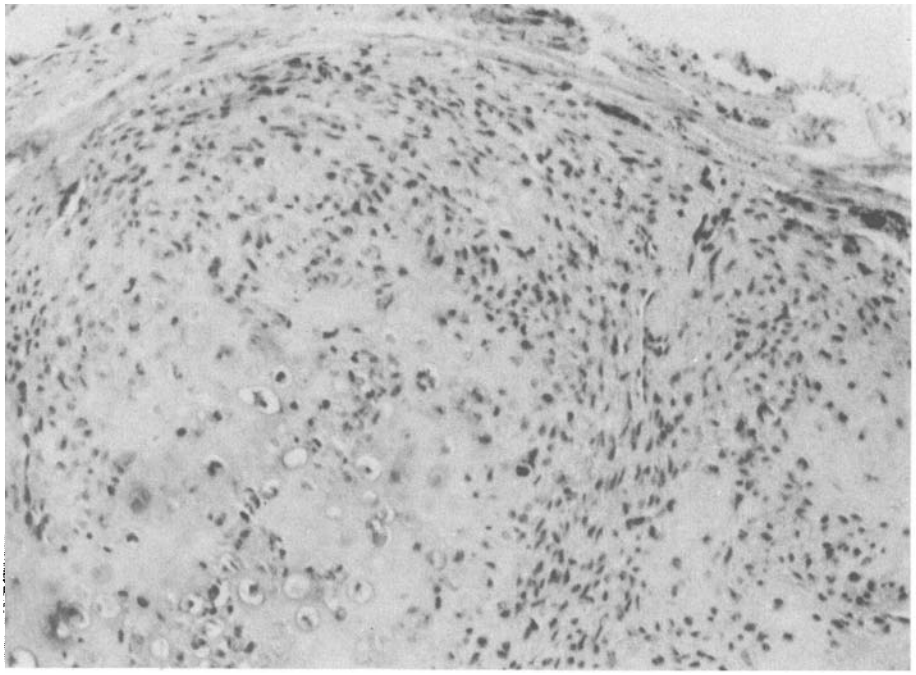
B

FIG. 2

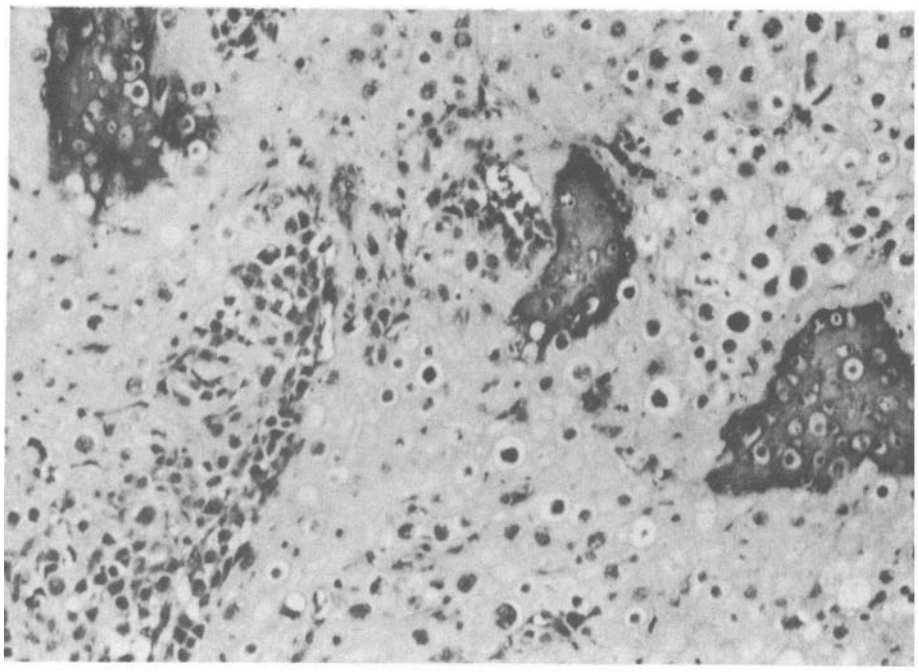
A. Case 1. Chondrosarcoma appears to be surrounded by a fibrous capsule. (Hematoxylin and eosin 145×).

B. Case 1. Undifferentiated chondrosarcomatous cells are actively proliferating, with atypical mitoses (bottom right corner). (Hematoxylin and eosin 580×).

https://doi.org/10.1017/S0022215100104451 Published online by Cambridge University Press



A



B

FIG. 3

A. Case 2. Chondrosarcoma appears as a well demarcated, multilobulated mass. (Hematoxylin and eosin 145×).

B. Case 2. Chondrosarcomatous cells undergoing scattered foci of calcification. (Hematoxylin and eosin 362×).

https://doi.org/10.1017/S0022215100104451 Published online by Cambridge University Press

knowledge on neoplastic growth thought to originate from reserve or stem cells undergoing incomplete or abnormal differentiation (Pierce and Cox, 1978). In other words, chondrosarcoma may derive either from cartilaginous tissue reserve cells (primary chondrosarcoma), or from primitive mesenchymal cells displaying chondromatous differentiation (so-called mesenchymal chondrosarcoma) (Brocheriou and Payen, 1975).

This issue is more than academic, because it helps to explain the rarity of these tumours in the maxilla—bone of exclusively membranous ossification—from mesenchymal cells rather than from embryonal cartilaginous nests.

Therefore, **mesenchymal chondrosarcoma** can be regarded as an ubiquitous tumour.

Both from a diagnostic and from a prognostic point of view, chondrosarcoma is an intriguing tumour. Clinically it does not present specific symptoms and radiologically it does not display any peculiar findings, since the small foci of calcification often found within the mass can also be observed in other tumours, such as osteosarcomas. Therefore, the diagnosis is based on histology alone.

However, from a pathological standpoint also the differential diagnosis is often difficult. The presence of either well differentiated chondroid tissue or of myxoid areas could suggest a benign chondroma, or a chondromyxoid fibroma at first proposed in our own cases.

It should be appreciated that both benign chondroma and chondromyxoid fibroma are even rarer in this area than chondrosarcoma (Brocheriou and Payen, 1975); therefore chondromatous tumours of the maxilla should always be suspected of malignancy (Dahlin and Henderson, 1956; Myers and Thawley, 1979). Maxillary chondrosarcomas have a poor prognosis (Fu and Perzin, 1974).

Radical surgery is mandatory to avoid recurrences which are extremely frequent (Batsakis and Dito, 1962).

References

Batsakis, J. G. and Dito, W. R. (1962) Chondrosarcoma of the maxilla. *Archives of Otolaryngology,* **75:** 55–61.

Brocheriou, C. and Payen, J. (1975) Tumeurs cartilagineuses des Maxillaires: a propos de 11 observations. *Annales d'Anatomie Pathologique,* **20:** 23–24.

Dahlin, D. C. and Henderson, E. D. (1956) Chondrosarcoma, a surgical and pathological problem. *Journal of Bone and Joint Surgery,* **38A:** 1025–1038.

Evans, H. L., Ayala, A. G. and Romsdahl, M. M. (1977) Prognostic factors in chondrosarcoma of bone. *Cancer,* **40:** 818–831.

Fu, Y. and Perzin, K. H. (1974) Non-epithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx: A clinicopathological study. *Cancer,* **34:** 453–463.

Jaffe, H. L. (1958) Tumors and tumorous conditions of the bones and joints. Henry Kimpton, London, pp 314.

Jones, H. M. (1973) Cartilaginous tumours of the head and neck. *Journal of Laryngology and Otology,* **87:** 135–151.

Myers, E. M. and Thawley, S. E. (1979) Maxillary chondrosarcoma. *Archives of Otolaryngology,* **105:** 116–118.

Pierce, G. B. and Cox, W. F. jr. (1978) Neoplasms as Caricatures of Tissue Renewal. In "Cell Differentiation and Neoplasia" (Saunders, G. F., ed.), pp. 57, Raven Press, New York.

Smatt, V. (1977) Chondrosarcome du Maxillaire Superieur: a Propos d'un Cas Clinique. *Revue de Stomatologie (Paris),* **78/1:** 45–51.

Spjut, H. J., Dorfman, H. D., Fechner, R. C. and Ackerman, L. V. (1971) Tumors of Bone and Cartilage, 2nd edn., fasc. 5. Armed Forces Institute of Pathology, Washington D.C.

Address for correspondence:
Prof. G. Massarelli,
Istituto di Anatomia ed Istologia Patologica,
via G. Matteotti 58,
07100 Sassari, Italy.

https://doi.org/10.1017/S0022215100104451 Published online by Cambridge University Press