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Angioglomoid Tumour of the Mandible

A Case Report

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Summary

A case of an angioglomoid tumour of bone occurring in the mandible is presented. The lesion is characterized by epithelioid and endothelioid cells that exhibit single and multilayered rings with a central signet ring-like cell suggesting vascular differentiation. These cells are arranged in strands and nests that are dispersed in a myxoid stroma. Radical resection of the involved mandibular bone was followed by a recurrence of the tumour in the interposed graft after 7 years. A second radical operation provided permanent cure as is demonstrated by a disease-free interval of 10 years. Histological features permitting the differentiation from other central jaw tumours are discussed.

Key-Words

Mandibular tumour – Vascular tumour of bone – Chondroid tumour of bone

Introduction

In 1976, *Tang* et al. reported a tumour in the right os pubis with a histological appearance not previously described. The unique features of this tumour were the clustering of epithelioid and endothelioid cells and the enclosure of solitary epithelioid cells in the lumina of single or multilayered endothelioid rings. Because of an invading and destructive growth pattern, the tumour was believed to be malignant. There was no recurrence six years after local excision and radiotherapy. *Tang* et al. (1976) presented presumptive evidence that the tumour was composed of angioblastoid cells that fail to organize in vascular networks, this opinion being based on the presence of capillary-like structures accompanied by epithelioid cells somewhat resembling glomus cells, and they used the term angioglomoid tumour of bone as a diagnostic label. However, they emphasized that their diagnosis was far from definite and that ultimate labelling awaited the description of additional similar cases. Such a similar case with involvement of the mandible is reported in the present paper.

Case History

A 43-year-old white male presented at the Department of Maxillofacial Surgery of the Utrecht University Hospital for treatment of a swelling in the right mandible. The swelling had been present for 4 years and had enlarged slowly during that time. Treatment elsewhere had consisted of extraction of several teeth and incision of the swelling without providing any relief.

Extra-oral examination revealed a firm-elastic, painless swelling of the right mandible extending from the 2nd molar to the ipsilateral canine. Trismus was not present nor was there disturbed function of the inferior alveolar nerve.

Inspection of the oral cavity revealed a swelling corresponding to that visible extra-orally. The tumour was bulging into the buccal as well as the lingual area. Firm-elastic portions alternated with fluctuant areas. The overlying mucosa exhibited hyperaemia but was not adherent to the tumour. There was no ulceration.

Radiographs (Fig. 1) showed a poorly circumscribed radiolucency in the body of the right mandible with considerable expansion and thinning of the inferior cortical border. A fine trabeculation was observed within the lesion. Pre-operative diagnosis was ameloblastoma.

Treatment consisted of resection of the involved jaw segment and reconstruction of the defect created with an iliac graft. The post-operative course was uneventful and no recurrence was observed during a period of 4 years after which the patient was lost to follow-up. Three years after his last follow-up visit, he returned with a swelling of the right mandible in the same area as before. Radiographs revealed apparent recurrent tumour extending from the bone graft into the distal part of the original mandibular bone. Again a radical resection of the affected bone was done, followed by reconstruction with an iliac graft. No recurrence has been observed for 10 years post-operatively.

Histological Features

The microscopic appearance of the tumour was the same in the original specimen and the recurrent lesion. At the

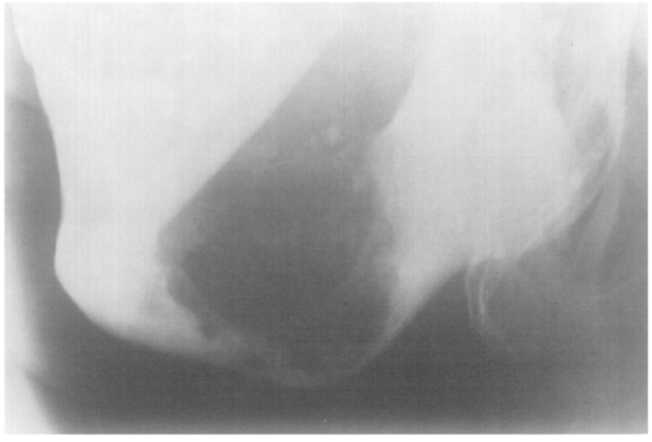

Fig. 1 Radiograph shows an irregular radiolucent lesion in the right mandibular body interspersed with fine bony septa and discrete calcifications.

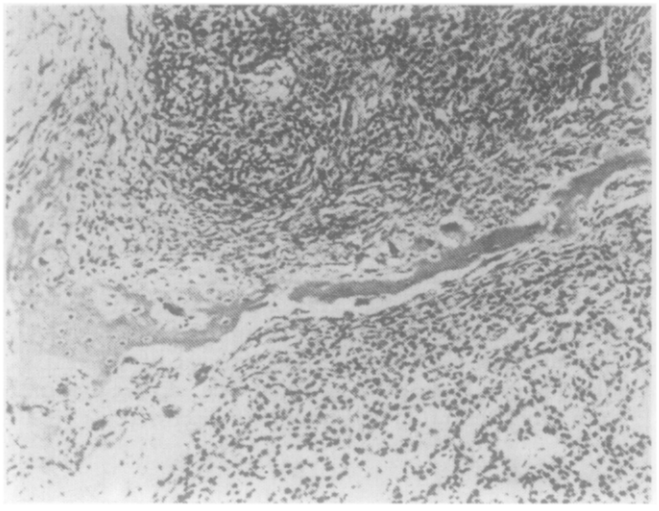

Fig. 2 Photomicrograph shows peripheral tumour area composed of closely packed cells. A rim of reactive bone projects from the overlying periosteum into the tumour. HE, × 36.

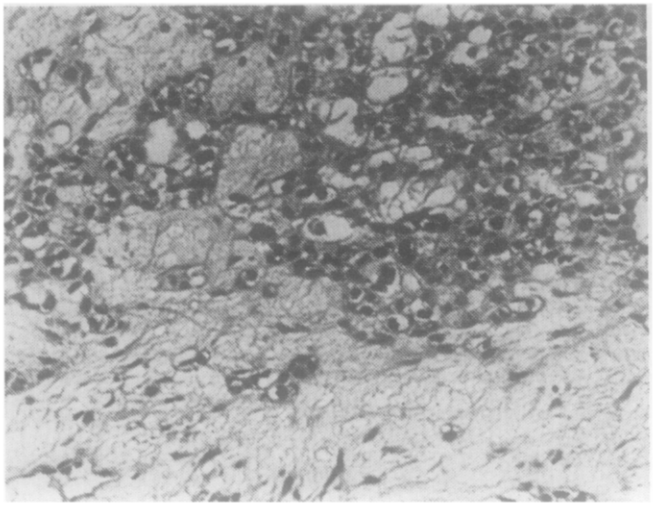

Fig. 3 More centrally, the compact tumour areas show a transition into strands and small clusters. HE, × 60.

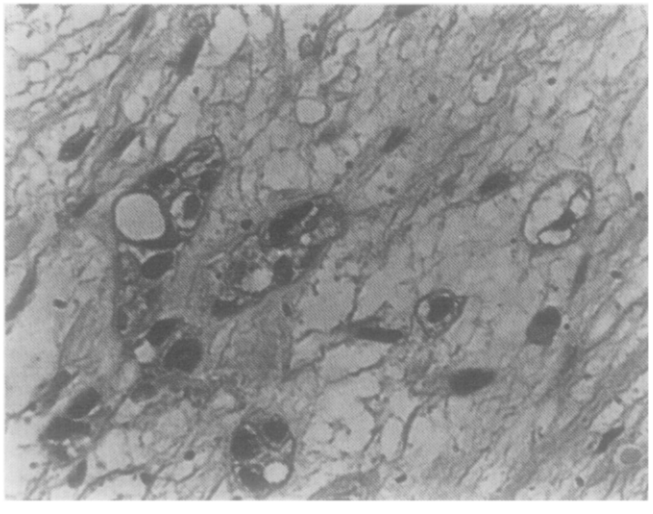

Fig. 4 Tumour cells with a vacuolated cytoplasm are dispersed in a loose-textured stroma. HE, × 150.

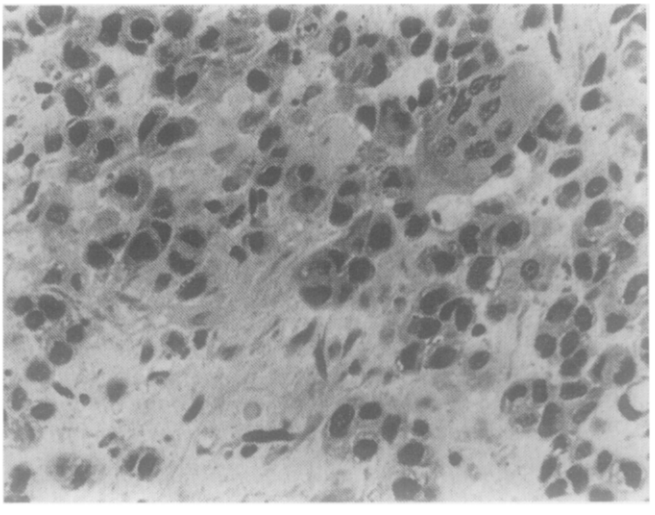

Fig. 5 An osteoclast-like giant cell is surrounded by tumour cells with vesicular nuclei and basophilic cytoplasm. HE, × 150.

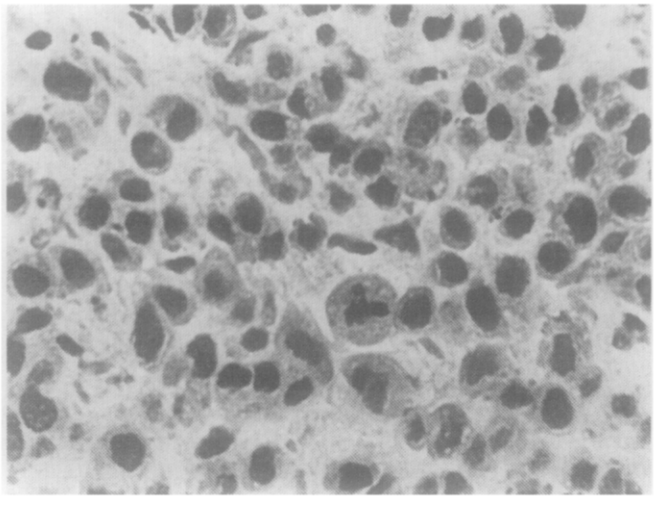

Fig. 6 High-power view shows poorly differentiated ovoid tumour cells with ample cytoplasm. An atypical mitotic figure is also present. HE, × 240.

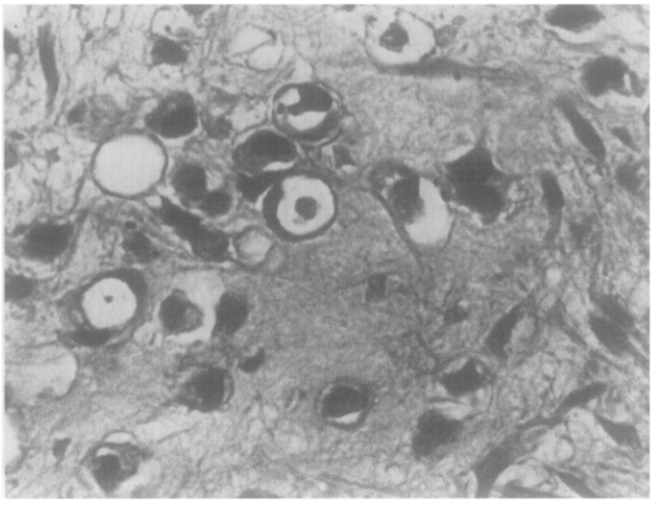

Fig. 7 Cytoplasmic vacuolation has resulted in the formation of several signet ring-like cells. Nuclear debris is present in some of the vacuoles. HE, × 240.

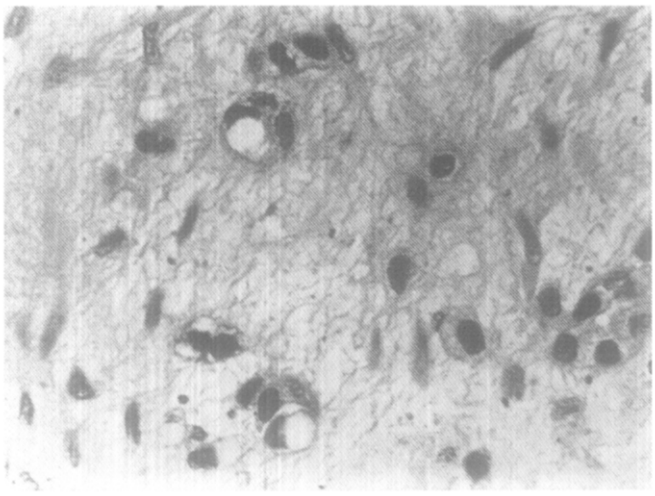

Fig. 8 Photomicrograph shows the typical multilayered structure that consists of a central signet ring-like cell that is embraced by two "navicular" cells. HE, × 180.

periphery of the tumour, the cells were arranged in extensive solid areas with little intervening stroma (Fig. 2). More centrally, these compact areas blended with anastomosing cords of tumour cells that in turn showed transition into small clusters or single cells (Fig. 3). The lesional stroma was relatively acellular (Fig. 4). Loose-textured fibrous areas alternated with myxoid, Alcian Blue positive zones as well as large haemorrhagic areas with many osteoclast-like giant cells (Fig. 5).

The tumour cells themselves exhibited a rather divergent morphology. In the solid areas they were plump and polygonal with a cytoplasm that varied from eosinophilic to basophilic. The nuclei were vesicular and mitotic figures were present in moderate numbers (Fig. 6). As the cells moved out of the compact areas to form strands and clusters, they exhibited an increasing cytoplasmic vacuolation. Single vacuoles coalesced, distending the cell body to form a vesicle; such cells appeared as signet ring-like cells (Fig. 7). Occasionally these cells were surrounded by one or several flattened cells and in this way a multilayered structure was created consisting of a central signet ring-like cell embraced by one or more "navicular" cells (Fig. 8). Nuclear debris was observed in some of the cytoplasmic vacuoles.

Staining for reticulin confirmed the epithelioid arrangement of the tumour cells. The stromal areas were rich in reticulin fibres but no reticulin fibres could be detected between individual cells within the tumour clusters. A condensation of reticulin reminiscent of a basement membrane bordered the clusters of tumour cells.

Immunoperoxidase staining for factor VIII-associated protein on paraffin sections was performed to substantiate the endothelial character (*Weiss* and *Enzinger*, 1982) but yielded negative results. Tissue for ultrastructural study was harvested from the paraffin blocks but the tissue ultrastructure has been destroyed by the previous paraffin embedding to such an extent that no reliable data could be obtained. As no fresh or formalin-fixed wet material was available for other additional studies, we could not obtain additional clues on histogenesis.

The tumour had invaded the surrounding cortical bone and overlying periosteum and had also infiltrated the adjacent intertrabecular spaces. No capsule could be discerned.

The tentative diagnosis was malignant bone tumour. Several pathologists were asked for advice; their diagnoses varied from chondrosarcoma to haemangio-endothelioma.

Discussion

In the current case, the diagnosis of angioglomoid tumour of bone was based on the light microscopy appearance that was similar to the case described by *Tang* et al. (1976). The striking features of this tumour are the co-existence of multilayered structures of vacuolated cells and a myxoid alcianophilic stroma. Some other tumour entities bear a superficial resemblance to this lesion. Firstly there is the possibility of a chondrosarcoma, especially the myxoid variant. Myxoid chondrosarcoma, however, mainly involves the soft tissues and moreover the tumour cells in this lesion are arranged in tiny anastomosing strand or sometimes in tubule-like formations with an ill-defined central empty pseudo-lumen (*Enzinger* and *Shiraki*, 1972; *Kindblom* and *Angervall*, 1983). The multilayered cell arrangement characterizing the present tumour is also absent in myxoid chondrosarcoma and as a consequence of these considerations, the possibility of a chondrosarcoma could be ruled out.

The second possibility is that the current case represents an epithelioid haemangio-endothelioma as defined by *Weiss* and *Enzinger* (1982). This tumour is composed of anastomosing strands of cells embedded in a myxoid matrix and cytoplasmic vacuolation is a prominent feature. Multilayered cell structures, however, are not described by these authors as a feature of this neoplasm and moreover all their cases involved soft tissues. The possibility that the angioglomoid tumour represents an intra-osseous example of an epithelioid haemangio-endothelioma cannot be discarded as prominent cytoplasmic vacuolation mimicking morphogenesis of a capillary is exhibited by both entities. Ultrastructural and immunohistochemical studies on fresh material from future cases may be helpful in deciding whether these tumours are identical.

The interpretation of the current case as malignant was based on the high cellularity in some parts, the prominent cellular and nuclear polymorphism, the mitotic activity and the infiltrative growth with invasion of adjacent bone and marrow spaces. In spite of this seemingly malignant histology, the clinical course was rather indolent; recurrence after 7 years and thereafter permanent cure by a second resection. The same favourable prognosis was exhibited by the case of *Tang* et al. (1976). It seems that local excision followed by radiotherapy or radical resection are succesful in eradicating this neoplasm.

The purpose of this report is for additional documentation of this rare tumour that couples a malignant histomorphology with an attenuated biological behaviour. Therefore we thought it appropriate to bring this entity to the maxillofacial surgery forum.

Acknowledgement

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