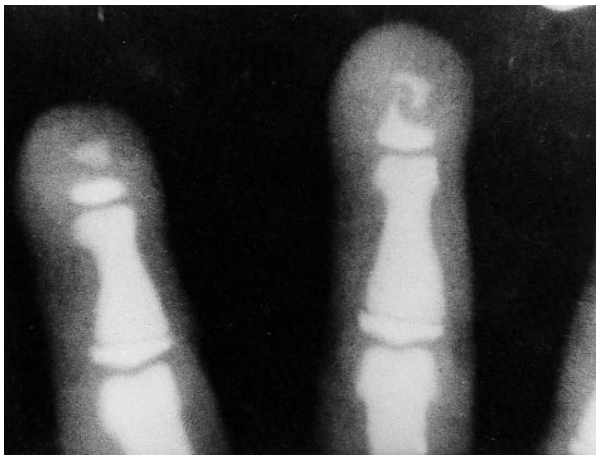

Fig. 1 – Hand X-ray showing digital acro-osteolysis.

DOI: 10.1054/bjom.1999.0020

ASSOCIATION OF CENTRAL GIANT-CELL GRANULOMA OF THE MAXILLA WITH PYKNODYSOSTOSIS

Sir,

We are reporting here, for the first time, the association of central giant-cell granuloma of the maxilla in a patient with classical clinical features of pyknodysostosis.[1] The granuloma showed partial regression with systemic corticosteroid therapy over a period of six weeks, indicating that systemic corticosteroids may be useful in some giant-cell granulomas of the maxilla.

A 13-year-old girl born to consanguinous parents presented with short stature and recurrent fractures after trivial trauma over a period of 3 years and a swelling in the palate in the preceding 2 months. Three siblings of this patient had died of obscure causes.

On examination, this young girl was of average intelligence, short statured (height was 125 cm – height standard deviation score: minus 3) and had normal body proportions. She was pale and bad koilonychia and acro-osteolysis (Fig. 1). Malunited fractures of both femora and tibiae were observed. She had a small face with micrognathia, retrognathia, a pinched nose with anteverted nares, frontal bossing and absent mandibular angles. A lesion arising from the palate was seen protruding from the oral cavity.

Intraorally, the lesion, which measured 8 × 4 × 3 cm involved the whole of the maxillary bone and was firm to hard in consistency. There was expansion of the alveoius in the medial and lateral aspects and the palatal arch was obliterated and maxillary dentition markedly displaced. Ulceration was seen on the anteroinferior aspect. The teeth consisted of permanent and deciduous molars, hypoplastic premolars, impacted canines bilaterally and anterior dentition resembling deciduous teeth. Partial anodontia was present in both jaws.

Examination of the abdomen revealed a soft liver 4 cm below the costal margin and a just palpable spleen. The rest of the physical examination was normal and genital development was appropriate for age.

Investigations revealed anemia (PCV of 19%), normal total and differential white-cell counts, and features of iron-deficiency anemia. Liver-function tests, creatinine, fasting plasma glucose, serum electrolytes, acid and alkaline phosphatase and creatine phosphokinase were normal. The girl's serum calcium was 8.4 mg%, serum phosphate 5.9 mg% and serum intact parathormone was 68 pg/ml (normal < 72 pg/ml). Her 24 h urine calcium was: 71 mg/1770 ml, suggestive of nutritional calcium deficiency, but urinary phosphate excretion 283 mg/24 h and renal phosphate threshold (3.4 mg%) were normal.

X-rays revealed generalized osteosclerosis of all bones, with acro-osteolysis of the distal phalanges. There was a radiolucent lesion involving the whole of the maxilla with floating dentition. The mandible, seen in the lateral skull film, showed elongated condyles and obtuse mandibular angles.

Bone biopsy from the posterior superior iliac spine showed marked sclerosis of bone with narrowing of the marrow spaces without significant osteoclastic proliferation. Biopsy of the maxillary swelling on histopathological evaluation (Fig. 2) showed large giant cells with up to 60 vesicular nuclei containing prominent nucleoli and eosinophilic cytoplasm and the stroma was composed of nondescript spindle-shaped cells and thin-walled vessels. There was inflammation, mainly at the periphery of the section where the lesion was seen to ulcerate through the oral mucosa. There was no evidence of vasculitis.

Since excision of such a large mass was not feasible, the patient was started on prednisolone 1 mg/kg/day and, on re-evaluation at four weeks, a marked reduction in the size of the lesion to more than half the original size was observed.

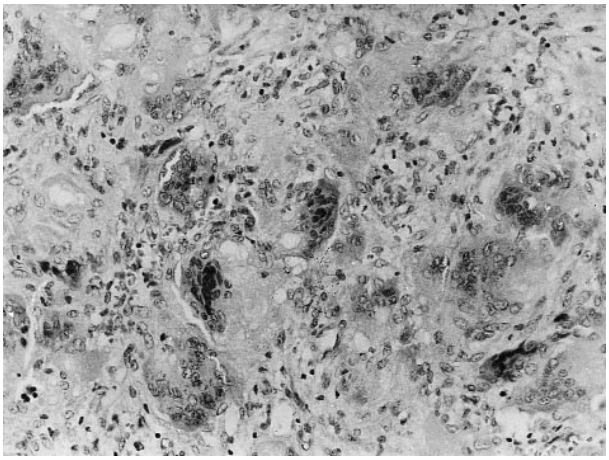

Fig. 2 – Histology of giant-cell tumour.

At six weeks, the regression bad ceased and the adjacent palatal mucosa had softened; a decision for curettage was made and surgery was carried out under general anaesthesia.

At surgery, the lesion was well circumscribed and less vascular than at the initial biopsy. It was completely excised and the underlying bone curetted. Postoperatively, the dosage of corticosteroids was tapered. One year later, the child is doing well without any recurrence of the tumour and repeat X-rays showed partial bone filling in the maxilla.

Our patient had typical clinical features of pyknodysostosis, normal serum levels of CPK and acid phosphatase, and lack of typical histological changes in bone biopsy excluded osteopetrosis.

Central giant cell granuloma[2] is predominantly a paediatric disorder and is twice as common in females. Mandibular involvement is more common. It is locally aggressive, and extensive tissue destruction can occur in some cases. The standard therapy of giant-cell granuloma involves curettage or surgical excision. We considered systemic corticosteroids in this patient because of the large size of the lesion, extension into both maxillae and vascularity of the lesion, and previous reports which indicate that intralesional steroids may be useful.[3] We surmised that steroid therapy may reduce the size of the lesion and make it more amenable to excision. We found an impressive reduction in the size of the lesion after 6 weeks of corticosteroids and the lesion could be totally excised.

In summary, our patient, who had classic features of pyknodysostosis, presented with a giant-cell granuloma arising from the palatal process of the maxilla. Systemic corticosteroid therapy led to useful reduction in the size of the tumour and made surgical excision easier. We suggest that the role of corticosteroids in giant-cell granuloma should be prospectively evaluated.

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Acknowledgments

We thank Mr Umakanth and Mr Seethapathy for the photography.

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DOI: 10.1054/bjom.1999.0030

VISUALIZATION OF METASTATIC LYMPH NODES IN ORAL CANCER BY ANGIOGRAMS

Sir,

Oral squamous cell carcinomas often metastasize to the submandibular and upper cervical lymph nodes. In addition to clinical assessment, helpful methods for clarifying presence of metastatic cervical lymph nodes (MCLs) are computed tomography (CT), ultrasonography and magnetic resonance imaging. On contrast-enhanced CT, a MCL appears as hyperdense components with a low attenuation necrotic area[1] (Fig. 1A). Hypodense areas correlate well with necrosis on pathologic slices, iso- or hyperdense areas with tumor or lymphoid component.[1] In this case (a 25-year-old woman, tongue cancer, well-differentiated squamous cell carcinoma, T4N2bMO), a MCL in the subdigastric nodes measuring 4×2 cm was fixed to the right internal jugular vein (arrow head) and made contact with the right internal and external carotid arteries. Suspecting invasion to the carotid arteries, we performed carotid angiography to disclose the relationship. A lateral view of a right common carotid angiogram revealed mild deviation of the right internal carotid artery without encasement. A lateral view of an external carotid angiogram disclosed tumor staining of the MCL in the subdigastric nodes. This correlated with our finding of hyperdense area on the contrast-enhanced CT (Fig. 1B, arrow). Visualization of the cervical lymph nodes was a rare finding on carotid angiograms,[2] but we could demonstrate tumor staining in other MCLs of head and neck carcinomas with superselective angiograms. The branches of the facial artery, superior thyroid artery and/or occipital artery gave off branches to MCLs. The