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CASE REPORT

Large Paediatric Central Osteoma with Osteoblastoma-Like Features in the Mandible

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Abstract
The diagnosis of osteomas in the paediatric population can pose a challenge to pathologists in excluding malignant bony tumours. We present the case of a 10-year old male presenting with a large left mandibular radiopaque lesion. This paper discusses the case of a central osteoma with osteoblastoma-like features, literature review, differential diagnosis of radiopaque lesions of the maxilla and mandible as well as a detailed discussion of the pathology of the lesion. Although similar lesions have been described in the sino-orbital region, this is believed to be the first report of this pathological entity in the mandible.

Keywords Osteoma · Radiopaque · Osteoblastoma · Bone tumour · Mandible

Introduction

We present a case of a 10-year-old male presenting with an unusual and uncommon densely radiopaque lesion in the left body of the mandible. This case highlights the challenges of diagnosing maxillomandibular radiopaque lesions and bony tumours of the mandible. Although similar entities have been described in the sino-orbital region [1], this is believed to be the first reported case in the mandible. This paper discussed the clinical case, differential diagnosis of the radiological findings, the detailed pathology of this lesion and a review of the literature.

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Clinical Case

A 10-year-old boy was referred to the oral and maxillofacial department at Monash Medical Centre for a gradually increasing hard swelling of his left mandible. He had presented to his local dentist a few days earlier and an orthopantogram (OPG) was performed which revealed a large radiopaque mass in the left mandibular body extending to the angle of the mandible (Fig. 1).

The swelling was asymptomatic and noted to be increasing in size over the last 4 months from when it was first noted. He was systemically well with no constitutional symptoms. Extraoral examination revealed a firm non-fluctuant swelling from the left mandibular angle to the midpoint of the mandibular body. No crepitus or 'egg-shell cracking' was noted. He had full range of mouth opening and had adequate oral hygiene. The swelling extended in the submandibular region and the posterior extent could not be palpated due to its location. He had normal lower lip, chin and tongue sensation.

For further evaluation, a contrast CT scan was organised. The images revealed a well-defined, homogenous, sclerotic, lobulated mass measuring $3.6 \times 3.0 \times 3.9$ cm (AP × transverse × craniocaudal) (Figs. 2, 3). The periodontal ligament space of the first and second molar were preserved but the mass had displaced inferiorly and narrowed the inferior alveolar canal.

Fig. 1 Initial OPG. Demonstrates a child in the mixed dentition phase with a large well demarcated homogenously radiopaque mass in the left body of the mandible. There is associated displacement of developing lower left 2nd and 3rd molars. No obvious resorption of adjacent roots is seen

A panel of bloods including full blood examination, renal function, calcium, magnesium, phosphate, and liver function tests were all normal.

Family history was unremarkable with no history of gastrointestinal symptoms or tumours suggestive of Gardner's syndrome.

Differential diagnosis based on the clinical and radiographic appearance included all bone forming tumours of the jaws. The radiographic appearance suggested a benign lesion as it was well-defined and there was no surrounding soft tissue reaction. However, as bone tumours have such a wide variation in their presentation, malignancy could not be excluded. The differential list included odontogenic tumours such as odontoma or cementoblastoma, as well as

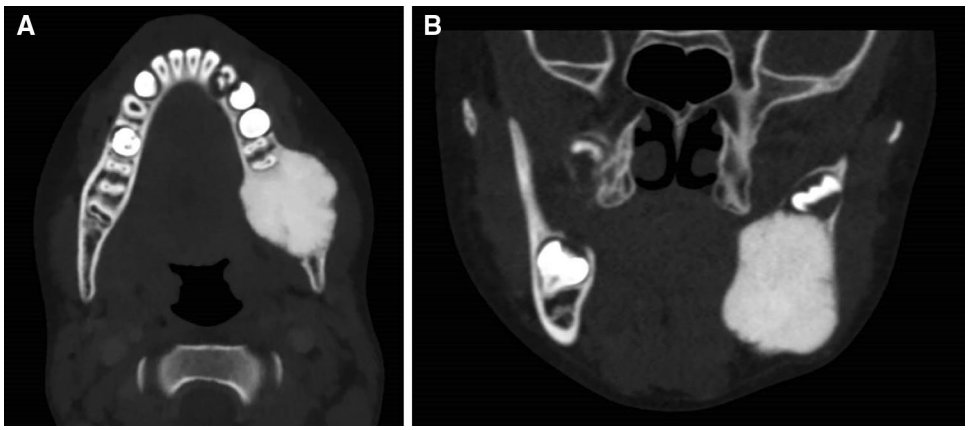

Fig. 2 Computerized tomography (CT) sections demonstrating a large homogenous expansile mass with obliteration of the inferior alveolar canal. **a** Axial slice at the level of C3. **b** Coronal slice showing degree of medial expansion of the lesion

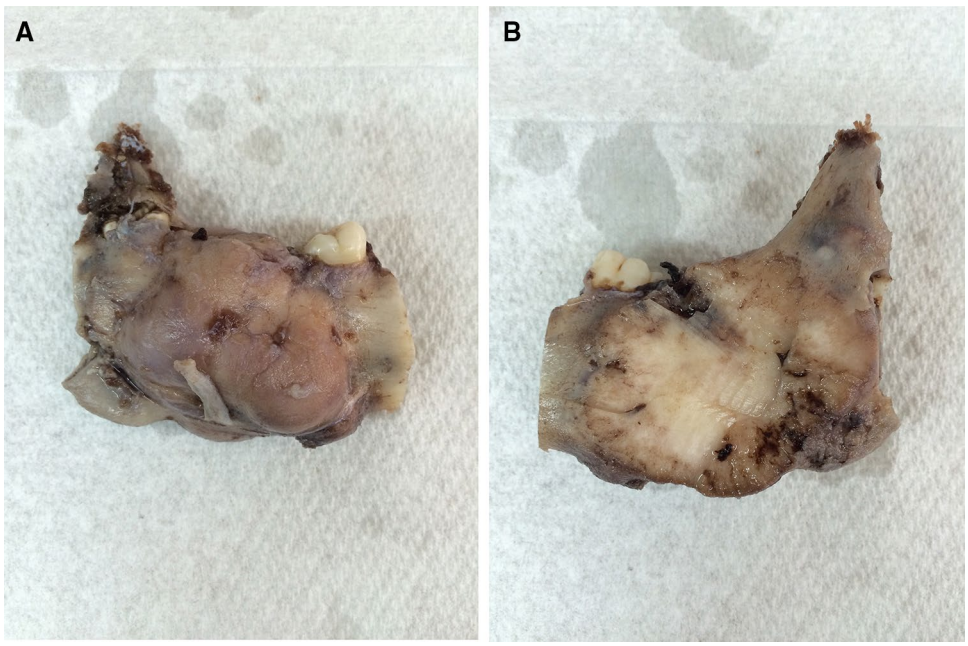

Fig. 3 Resected specimen-macro samples. **a** Lateral view of specimen. (**b**) Medial view with transection of area of tumour expansion showing the thick dense bone in the lesion

non-odontogenic tumours such as osteoma, osteoblastoma, osteosarcoma, and ossifying fibroma.

An initial incisional biopsy was performed initially. The histopathological appearance revealed an unclear picture due to the presence of islands of cells within a lesion of densely lamellar bone. The presence of large active osteoblasts also raised the possibility of a low-grade osteosarcoma. These findings were discussed with several consultants in the department and with the patient's family. Bearing in mind the size of the lesion, uncertainty of the exact pathology, and it being a potential malignancy, a decision was made to perform a segmental mandibulectomy with a 1 cm bony margin and next anatomical barrier soft tissue margin. The condylar head and neck would be spared to allow for continued growth and development of the mandible.

A segmental mandibulectomy was performed under general anaesthetic and the lesion was resected with its overlying capsule. The resected specimen is shown in Figs. 3 and 4 shows a radiograph of the specimen. A custom prebent reconstruction plate was placed from the body of the mandible anterior to the mental nerve to the neck of the condyle (Fig. 5). Due to the patient's age, the resection was aimed at maintaining as much of the condyle as possible to preserve the growth centre and minimise the surgical requirements in the future.

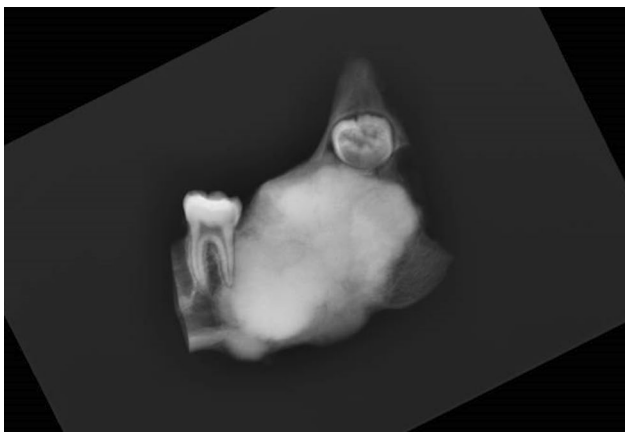

Fig. 4 Plain film of the intra-operative specimen

The resected specimen, along with clinical and radiographic information, was sent for further analysis at several local institutions including the Royal Children's Hospital (Victoria, Australia) and Monash Medical Centre (Victoria, Australia). Opinions ranged from osteoma, osteoblastoma, and low-grade osteosarcoma, with no consensus reached.

On gross examination, the biopsy consisted of a bosselated mass of grey-white bone. The surgical margins were 7 and 8 mm from the anterior and posterior aspects of the tumour respectively. Haematoxylin and eosin staining revealed a central portion that was composed almost entirely of densely sclerotic woven bone with scant stroma. The peripheral edges of the tumour revealed plump amphophilic osteoblastic cells with mildly pleomorphic enlarged nuclei, small nucleoli, and copious cytoplasm. Mitotic figures were infrequently seen. The tumour showed a relatively well circumscribed pushing margin, however occasional sections show some interdigitation of abnormal and adjacent normal bone (Fig. 6).

The specimen and radiographic data was forwarded to Mayo clinic Rochester, MN, USA for second opinion to rule out a low-grade osteosarcoma. These were analysed by a specialist pathologist and specialist musculoskeletal radiologist and the final diagnosis of "osteoma with osteoblastoma-like features" was made.

The patient has been followed up now for 12 months with no clinical or radiographic evidence of recurrence. The OPG at 6 months revealed some amorphous calcified tissue forming around the reconstruction plate representing spontaneous regeneration of bone.

Future reconstruction will be planned after on-going review of spontaneous regeneration and growth.

Fig. 5 Post-operative X-rays after reconstruction with reconstruction plate only. **a** OPG. **b** Towne's view

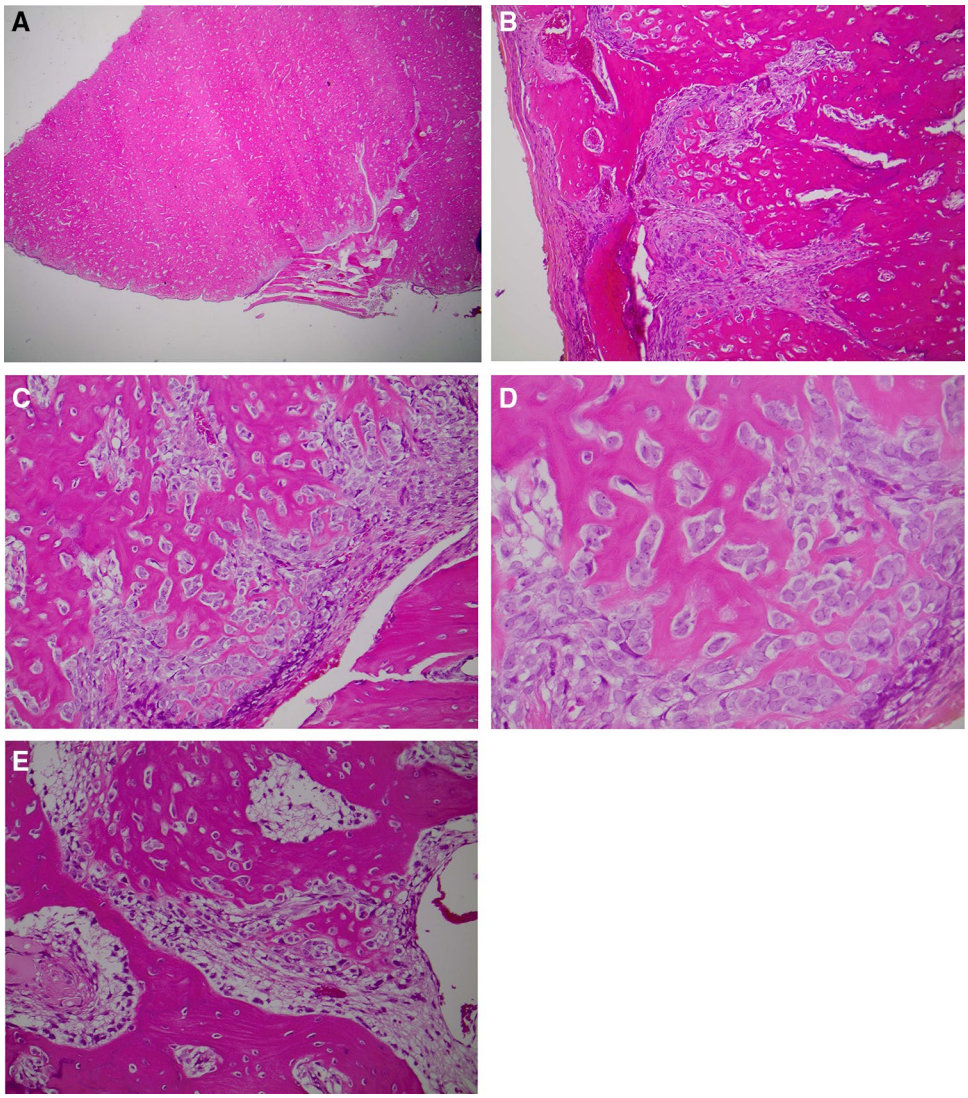

Fig. 6 Photomicrograhs of the final pathology. **a** Low power image demonstrating predominance of lesion is dense lamellar bone resembling an osteoma with a margin of plump osteoblast like cells. **b** Photomicrograph showing the cellular tumour front focally eroding into the normal overlying bone of the mandible. (Normal mandible on the left). **c** High power photomicrograph of periphery of the lesion showing a focal area of increased cellularity with plump osteoblast cells. Mildly atypical, but without mitoses and not obviously malignant. **d** High power view (×400) of the cellular area demonstrating the nature of the plump osteoblasts. Minimal atypia is seen, and no mitoses. **e** Low power view showing areas of plump osteoblasts within a fibrovascular stroma with osteoid deposition and woven bone formation. The normochromatic nature of the cells (minimal atypia, not anaplastic), and the predominant lamellar bone structure of the lesion was more in favour of a benign bony tumour

Discussion

Bone forming tumours are notoriously difficult to diagnose due to their radiographic and morphological heterogeneity. These tumours can be found within, or on the surface of, bone and contain neoplastic osteoblasts that have a spectrum of appearances. Benign tumours have inactive osteoblasts which are small and usually in a single layer adjacent to the surface of the mineralised tissues. In comparison, aggressive and malignant tumours show osteoblasts that are large, have prominent and irregular nuclei, increased mitotic figures, abundant eosinophilic nuclei, and form clusters and sheets of cells.

When considering the differential diagnosis for a radiopaque lesion in the maxillofacial skeleton, several lesions as considered.

Odontomas are mixed (epithelial and mesenchymal) odontogenic tumours that form tooth-like material in various states of histodifferentiation and morphodifferentiation. They are considered hamartomas. They are well defined radiopacities and are often incidental findings when examining for unerupted teeth. They are slow growing and rarely cause jaw expansion which is only seen in the larger complex odontomas. A radiolucent rim is often seen radiographically that aids in diagnosis.

Compared to osteoma, osteoblastoma is a rare bone forming tumour of neoplastic osteoblasts and uncommon in the facial skeleton. Clinically, they present with pain as the most common symptom. Diagnosis occurs in the fourth to sixth decade and have a 2:1 male predominance. Radiographically they are larger than 2 cm, a wide range of radiolucent-radiopaque appearance, and have a well-defined margin. Histologically, they appear as haphazard arranged woven bone with large osteoblasts in a loose fibro vascular stroma which can represent a healing callus. These lesions are very active and undergo constant remodelling so areas of Paget's

like bone (multiple reversal lines) are often seen. They are histologically indistinguishable from osteoid osteoma therefore an arbitrary radiographic criterion of 1.5–2 cm in size is used to differentiate these two entities. Osteoid osteomas have a unique clinical feature with night-time pain relieved by NSAIDs due to prostaglandin production within their central nidus. On immunohistochemistry, nerve fibres can be seen extending into the tumour.

Osteoblastomas have a spectrum of histological appearances and clinical behaviours. Compared to the typical osteoblastoma described above, multifocal/multinodular osteoblastoma have numerous growth centres/nidi with spindle cells that can be mistaken for an osteosarcoma. Pseudomalignant osteoblastoma have degenerative cytological atypia with large degenerated nuclei and smudged chromatin mimicking malignancy, however there is no evidence that they behave differently. Aggressive osteoblastoma are locally aggressive, contain large epithelioid osteoblasts with abundant eosinophilic cytoplasm, often arranged in sheets rather than the innocuous single layer rimming, and are very difficult to differentiate from osteosarcomas. Despite controversy regarding management of the aggressive form of osteoblastoma, they are benign tumours that do not metastasize and therefore can be managed with curettage alone. Recurrence following curettage is up to 20% [2], and therefore in expendable bone, en bloc resection can be considered and is usually curative [3].

Cementoblastoma also falls in the spectrum of osteoblastoma but being intimately associated with the roots of teeth. The mean age at diagnosis is 20 and there is no gender predilection [4]. A comparison of histological features reveal very little difference between these two entities. It was previously noted that cementoblastomas were unique in having spicules of radiating bone, however osteoblastomas have been noted with this appearance [5]. The only reliable difference appears to be their ability to invade through the intervening layer of periodontal ligament and being in direct association with the root surface. In the case described above, the periodontal ligament space of all the involved teeth was carefully examined and there did not appear to be any radiographic evidence of involvement with the tooth roots. This excluded the diagnosis of cementoblastoma where this is a defining criterion [5].

The patient's age and history of rapid growth suggested the possibility of osteosarcoma. In comparison to all other diagnoses considered so far, which are more common in an older population, more than 50% of osteosarcomas are diagnosed in patients under 20 years of age. Classic osteosarcomas, on histopathology, have large cells with striking pleomorphism, hyperchromia, prominent nucleoli, and frequent abnormal mitoses. Despite this, up to 10% display little anaplasia and can be confused for benign diagnoses [6]. The tumour matrix shows considerable variation from lacelike osteoid to dense sheets of woven bone. Areas which are intensely sclerotic, cells are small and scarce with no mitotic figures. Another pattern of histology is that which represents an osteoblastoma with large cells with bizarre hyperchromatic nuclei. The presence of these 'osteoblastomatous' lesions permeating the marrow spaces and trapping host lamellar bone represents an osteoblastoma-like osteosarcoma.

The radiographic location and appearance in our case was more in line with a benign osteoma. Osteomas are usually slow growing, benign lesions of well-differentiated mature bone. They have a slight male predominance and a mean age of diagnosis around 40–65, although age range varies widely. They are found in most areas of the skeleton but the vast majority are found in the craniofacial skeleton, especially the paranasal sinuses. They most commonly grow on the surface of bones (peripheral), and are termed exostoses and include maxillary and mandibular tori. Those that form within the medullary space are termed enostoses or central osteomas and are considered separate from peripheral osteomas as they rarely/very slowly cause expansion. They are often radiologically undistinguishable from dense bone islands (DBIs) or areas of focal bone sclerosis. Central osteomas are much rarer than their peripheral counterparts arising from the endosteum and can present significant diagnostic dilemmas. There are only 11 clearly documented, non-syndromic, cases of central osteomas and therefore their behaviour is not well understood and differentiated from peripheral osteomas [7–10]. They are usually asymptomatic but can cause local pressure effects such as pain, headache, disfigurement, especially when associated with the paranasal sinuses. Rarely, root resorption is seen with bone islands, however, these may be misdiagnosed osteomas. They are usually solitary and the detection of multiple lesions should raise the suspicion of Gardner's syndrome (GS). Gardner's syndrome is a subtype of familial adenomatous polyposis (FAP) characterised by the constellation of inherited adenomatosis, osteomas, epidermal cysts, and fibromas. It is caused by mutations in the adenomatous polyposis coli (APC) gene and is equally distributed between the genders. Up to 30% of new cases are diagnosed as new mutations (i.e. those not belonging to identified families). Osteomas were the first recognised extra-colonic manifestation of FAP and GS. Other dental anomalies related to GS include unerupted teeth, supernumerary teeth, and dentigerous cysts. Osteomas often precede the development of colonic polyps so the finding of multiple osteomas should prompt further investigation. When associated with GS, central radiopaque lesions rarely cause expansion and resemble enostosis rather than osteomas [11].

A variant of osteomas has been described in the sino-orbital region termed 'osteomas with osteoblastoma-like features'. These vary pathologically from traditional osteomas

in that they have more discrete areas of active osteoblastic activity [1]. In an analysis of 45 excised sino-orbital osteomas, 38% had areas indistinguishable from osteoblastomas. These were termed osteomas with osteoblastoma-like features. In this series, they appeared to behave no differently to other sino-orbital osteomas [1]. They were, however, noted to occur in younger patients, and to have a 3:1 M:F ratio with a male predominance compared to 1:1 for conventional osteomas. These features were noted at the base of the tumour adjacent to normal bone and therefore it has been suggested that these areas represent regions of active growth [3]. The large active osteoblasts laying down new osteoid in these areas are the same pathological process, rather than representing a transformative change to osteoblastoma or more aggressive behaviour. This entity has not been reported outside of the sino-orbital region previously.

Pathologically, osteomas are primarily composed of dense, compact bone and broad trabeculae of mature bone within a paucicellular fibrous stroma. The outer surface is often sharply demarcated and may have a single layer of overlying small osteoblasts. In the sino-nasal region, they are often lined by respiratory epithelium.

In this case, the final pathology was consistent with an osteoma with osteoblast-like features (Fig. 6). The tumour was densely sclerotic, and required 4 weeks of decalcification before sectioning. The lesion consists of irregular mildly cellular sclerotic bone with relatively small amounts of stroma. At the advancing front, the lesion is more cellular where enlarged plump cells with poorly defined cytoplasmic boundaries, enlarged nuclei and small nucleoli. These cells are mildly pleomorphic, but mitoses are not identified. There were also some patchy areas of centrally located cellular areas with woven bone, osteoid and a fibrovascular stroma. Most of the lesion is well circumscribed, but in occasional sections, there is interdigitation of abnormal and adjacent normal bone. The lack of mitoses, lack of necrosis and minimal pleomorphism were reassuring that this was not an osteoblastic osteosarcoma but an osteoma with osteoblastoma-like features.

In our case, based on the concern of osteosarcoma after the initial biopsy, and the rapidly enlarging lesion and involvement of the entire height of the mandible, a segmental resection was performed. This is very similar to a previous case of a rapidly expanding cementoblastoma which displayed radiographic and histological features of osteoblastoma and low-grade osteosarcoma [12]. As in that case, the lesion was treated aggressively to avoid undertreating the potentially malignant underlying pathology.

Conclusion

This case highlights the challenges with pathological diagnosis of radiopaque lesions in the maxilla and mandible. In this case, a large radiopaque lesion in the left mandible of a 10-year old male was eventually resected and final pathology confirmed an osteoma with osteoblastoma-like features. Twelve months follow up has demonstrated no evidence of recurrence. This pathology is more likely to be seen in younger patients, and should be considered as a differential diagnosis for radiopaque lesions in the maxillomandibular complex as well as in the sino-oribital region.

Compliance with Ethical Standards

Conflict of interest All authors declared that they have no conflict of interset.

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