CASE REPORT

Desmoplastic ameloblastoma of mandible

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SUMMARY

Desmoplastic ameloblastoma is one of the six histopathological subtypes of ameloblastoma. The age and gender groups affected by desmoplastic are similar to those affected by the conventional ameloblastoma. It usually presents as a painless enlargement of the jaw. Owing to its deceptive radiological appearance as a mixed radiopaque–radiolucent lesion, it is often mistaken as a fibro-osseous lesion. Histologically, desmoplastic ameloblastoma has a densely collagenised and hypocellular stroma, where the epithelium tends to proliferate in the form of cords and nests instead of cellular islands. Most desmoplastic ameloblastomas display occasional classic islands of follicular ameloblastoma among the predominant strands and cords. Studies have shown that desmoplastic ameloblastoma shows a tendency to recur. We present a rare case of a tumour occurring in the anterior mandibular region in a 60-year-old man over a period of 11/2 months.

BACKGROUND

In 1984 Eversole *et al* described a new variant of ameloblastoma which had a clinicoradiographic and histopathological picture different from the conventional ameloblastoma.^{1 2} The histological picture demonstrated extensively collagenised stroma containing nests and strands of odontogenic epithelium. This was later known as desmoplastic ameloblastoma. The desmoplastic variant differs from other variants of ameloblastoma in that it is located in the anterior region of jaw and its radiographic appearance is often more representative of a fibro-osseous lesion.² Ameloblastomas of 0.9–12.1% have been reported to be desmoplastic ameloblastomas.³ Owing to this presenting uniqueness and an aggressive course, diagnosis of this rare entity becomes a challenge. This disease should be considered in the list of differential diagnoses of any mixed radiopaque–radiolucent lesion occurring in the anterior region of jaw.

CASE PRESENTATION

A 60-year-old man reported to the outpatient department with a symptom of swelling in the lower jaw for the past 11/2 months. History revealed that the swelling had gradually increased in size over a period of 11/2 months. Initially the swelling was small but increased rapidly to the present size. On physical examination, facial asymmetry due to an extraoral swelling was found, which obliterated the labiomental sulcus (figure 1). Thickening of the inferior border of the mandible at the symphysis was felt. A single well-defined dome-shaped swelling was present in the midline of

the mandibular residual ridge extending from the region of 33 to 44, crossing the midline. The swelling measured 5×6 cm in dimensions (figure 2). There was extensive buccal and lingual cortical plate expansion. The overlying mucosa was stretched, smooth but was ulcerated at the point where the opposing tooth 13 impinged on the swelling (figure 3). On palpation, the swelling was found to be soft-to-firm in consistency and slightly tender but only around the area where the ulcer was present. A provisional diagnosis of ameloblastoma was given.

INVESTIGATIONS

A mandibular anterior topographic occlusal radiograph showed a single well-defined multilocular homogeneously radiolucent lesion with corticated border (figure 4). The panoramic image revealed a single, well-defined mixed radio-opaque radiolucent lesion in the midline of the mandible extending from the region of 35 to 46. The lesion had a corticated and scalloped inferior border. The internal structure was of varying radiodensities with more radiodensity in the region of 33 (figure 5). Aspiration yielded blood-tinged fluid and

Figure 1 Swelling of the mandibular symphysis region.

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Figure 2 Dome-shaped swelling on the mandibular residual ridge with a traumatic ulcer on the superior surface.

examination of the same turned out to be infected cystic fluid (figure 6). Blood picture showed no abnormality except slightly raised levels of serum alkaline phosphatase, 150 IU/L, suggestive of a bone forming lesion. Incisional biopsy confirmed the histopathological diagnosis of desmoplastic ameloblastoma. The report showed odontogenic cells lying in a background of fibrous tissue in the form of islands and the plexiform arrangement was appreciated (figure 7). The islands exhibit peripheral lining of darkly staining cuboidal cells with central polygonal cells. A few islands showed cystic degeneration and squamous metaplasia.

DIFFERENTIAL DIAGNOSIS

A list of differential diagnosis was given which included a central giant cell granuloma, aneurysmal bone cyst and primary intraosseous squamous cell carcinoma and metastatic bone tumour.

TREATMENT

Resection with wide margins was performed and the patient is regularly recalled for follow-up to check for any signs of recurrences.

Figure 3 Upper canine impinging on the mandibular swelling causing an ulcer.

DISCUSSION

Desmoplastic ameloblastoma (DA) is a subtype of the conventional ameloblastoma which has predilection to occur in the anterior regions of the jaws and shows a mixed radiolucent–radiopaque pattern radiographically, similar to fibro-osseous lesions.⁴ Studies have shown that 13% of the ameloblastomas are of the desmoplastic variety⁴ while according to Kishino *et al*⁵ the DA accounts for about 5.3% of all ameloblastomas. This mixed radiographic appearance is due to osseous metaplasia within the dense fibrous septa that characterise the lesion, and not because of the tumour itself producing a mineralised product.¹ Reactive osteogenesis may occur in desmoplastic variants. This makes it difficult to differentiate it from an adenomatoid odontogenic tumour, ameloblastic fibro-odontoma and odontoameloblastoma.⁶ It has been suggested that DA is derived from the periodontal ligament tissues.⁵

DA affects men as well as women equally and usually occurs in a wide age range of 20–80 years.⁷ There is no proven racial predilection but few authors suggest that this variety of ameloblastoma may have a predilection for the oriental population.⁸ Most of the literature suggested that DA is a small lesion, size usually limited to 2 cm or less. Macroscopically, DA has a gritty texture and has a solid cut surface.² In this case the lesion was located in the anterior region of the mandible measuring 5×6 cm in diameter and palpatory findings showed that the swelling had a doughy consistency. Radiographically, about 50% of DA show a mottled, mixed radiolucency/radiopacity with diffuse margins, making it difficult to differentiate it from a fibro-osseous lesion, this being the reason of including fibro-osseous entities in the differential diagnosis.^{2–9} It was hypothesised that this maybe due to the infiltrative nature of DA to involve the trabeculae.⁷ Three radiological presentations of DA are mentioned in the literature as follows: type I (osteofibrosis type) which has radiolucent as well as radiopaque appearance; type II (radiolucent type) which has a completely radiolucent appearance; and type III (compound type) which has radiolucent as well as radiopaque appearance combined with a large radiolucent change. Radiographic features of our case showed mixed radiodensities which were consistent with that of osteofibrosis type which is the most common pattern and the compound type is the least common.¹⁰ Histologically, DA does not show capsule formation and interstitium is abundant with bundles of collagen fibres. The stromal components dominate over the epithelial components of the tumour. The cellular components of the tumour consist of whorls of epithelial

Figure 4 Mandibular topographic occlusal view showing a single multilocular radiolucency with a corticated border.

Figure 5 Panoramic radiograph showing a single multilocular lesion with varying degrees of radiodensities affecting the midline of the mandible.

islands with cuboidal epithelial cells at the periphery containing hyperchromatic nuclei. There are whorls of spindle-shaped cells towards the centre of the islands. Formation of metaplastic osteoid trabeculae may be present.² It is distinctive due to an abundance of collagen with a moderate amount of cellular components of connective tissue.⁴ The palisading pattern of follicles as observed in conventional ameloblastoma is absent.¹¹ Immunohistochemistry suggests a marked expression of transforming growth factor β .¹² Various immunohistochemical studies have reported DA tumour cells as showing expression of S-100 protein and desmin.¹³ There may be increased expression of caspase-3 and Fas, cell surface receptor protein of tumour necrosis factor receptor family,¹⁴ p63¹⁵ decreased expression of

Figure 7 Nests of odontogenic cells lying in a background of dense fibrous stroma.

cytokeratin19.¹⁶ Prognosis of this disease is dependent on the treatment modality instituted. High recurrence rates have been reported with curettage and excision. Curettage is an inappropriate treatment for ameloblastomas because recurrence becomes inevitable and difficult to treat. A better prognosis is observed when the tumour is removed by extended resection.⁹

Desmoplasia of the stromal connective tissue in DA be thought to be a maturation change in a solid ameloblastoma, as it has been observed that dense collagenisation may be observed in tumours with a long history.¹⁷ This logic can be applied to cases of hybrid tumours. A combination of DA with any other histological type of multicystic ameloblastoma is called a hybrid ameloblastoma.² So it may be hypothesised that a hybrid tumour may be a transitional phase in the maturation of a solid multicystic ameloblastoma to the desmoplastic variety.

To eliminate the delay in diagnosis of recurrent lesions, close follow-ups with radiographic imaging modalities are advised.

Figure 6 Blood tinged aspirate.

Learning points

- ▶ Desmoplastic ameloblastoma has a predilection to occur in the anterior region of the lower jaw.
- ▶ It often mimics fibro-osseous lesions.
- ▶ Desmoplastic ameloblastoma should be included in the list of differential diagnosis of a mixed radiolucent–radiopaque lesion.
- ▶ Fine needle aspiration cytology may mislead to the diagnosis of a cystic lesion.
- ▶ Biopsy is mandatory for confirmatory diagnosis.
- ▶ Association of desmoplastic component in a hybrid tumour is not rare.

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Patient consent Obtained.

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