Rare disease

Ameloblastic carcinoma: a rare entity

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Summary

Ameloblastic carcinoma is a rare aggressive malignant epithelial odontogenic tumour seen in a wide range of age group with no sex predilection. Patient usually presents with a rapidly enlarging swelling. It usually involves the posterior portion of the mandible. Treatment of choice is surgical removal of tumour followed by radiotherapy. We present a case of ameloblastic carcinoma in a 35-year-old man.

BACKGROUND

Ameloblastic carcinoma is a very rare tumour and fewer than 35 cases have been reported till now. Literature on the treatment modalities is limited.

CASE PRESENTATION

A 35-year-old man presented with painless, rapidly enlarging swelling at the left side of the lower jaw for 3 months. Swelling was 5×5 cm in size, firm in consistency and non-tender. Overlying skin was normal. No lymphadenopathy was observed. Systemic examination was unremarkable. CT scan of head and neck showed a multiloculated, expansile lytic lesion with associated soft-tissue mass within the left mandible, typical of ameloblastic malignancy (figure 1).

No other lesion was noticed. Chest X-ray was unremarkable. Incisional biopsy showed densely packed hyperchromatic spindle cells along with few mitotic figures. On the basis of clinical, radiological and histopathological examination, a tentative diagnosis of ameloblastic carcinoma was made and hemimandibulectomy was advised. Hemimandibulectomy was done and tissue sent for histopathological examination, which showed nests of cells having distinctive features of ameloblastic differentiation (figure 2), that is, peripheral palisading of basaloid cells coupled with the dyscohesiveness of the cells in the middle of the nests created the typical stellate reticulum arrangement and hyperchromatic nuclei (figures 3, 4A and 4B). Areas of necrosis along with nuclear features of malignancy were

Figure 1 CECT scan showing a multiloculated, expansile lytic lesion with associated soft-tissue mass within the left mandible.

Figure 2 Section showing variably sized nests of basaloid cells (H&E×50).

Figure 3 Section showing hyperchromatic nuclei (H&E×500).

found (figures 5A and 5B). Mitotic figures were also seen and no evidence of keratinisation was noted. A final diagnosis of primary ameloblastic carcinoma was rendered. One surgical margin showed tumour cell infiltration. As one of the tumour margin was positive for tumour cells, postoperative radiotherapy was advised. Patient did not

have any distant metastasis or local recurrence till now (14 months after surgery).

DIFFERENTIAL DIAGNOSIS

- Squamous cell carcinoma (basaloid variant).
- Squamous odontogenic tumour.

Figure 4 (A and B) Section showing dyscohesiveness of the cells at the centre of the nests created the stellate reticulum formation (H&E×500).

Figure 5 (A) Section showing areas of necrosis (H&E×50). (B) Section showing hyperchromatic nuclei and focal necrosis (H&E×125).

- ▶ Acanthomatous ameloblastoma.
- ▶ Kerato-ameloblastoma.
- ▶ Malignant ameloblastoma.

TREATMENT

Hemimandibulectomy was done followed by radiotherapy.

OUTCOME AND FOLLOW-UP

Patient is well till now.

DISCUSSION

The term ameloblastic carcinoma was first described by Shafer *et al*¹ in 1983. Ameloblastic carcinomas are extremely rare malignant odontogenic epithelial neoplasm, and, to our knowledge, fewer than 35 cases have been reported worldwide. It may arise de novo or from a pre-existing benign lesion.^{2–3} The term malignant ameloblastoma is used for the lesions that metastasize despite their benign histology.² The tumour can affect any age group with a mean age of 30 years and has no sex predilection.⁴ It is commonly seen in the posterior portion of the mandible⁴ as in our case but rarely maxilla can be affected.^{5–8} The most common complaint is rapidly enlarging swelling, our patient also presented with the same complaint. Other complaints are pain, rapid growth with trismus and dysphonia.⁴

The differential diagnosis includes squamous cell carcinoma (basaloid variant) arising in the lining of odontogenic cyst,^{9–10} squamous odontogenic tumour,¹¹ malignant ameloblastoma, acanthomatous ameloblastoma and keratoameloblastoma. The features that distinguish ameloblastic carcinoma from squamous cell carcinoma are jigsaw puzzle type of nests of tumour cells, lack of keratinisation and the presence of stellate reticulum cell appearance in the former. On the other hand, squamous odontogenic tumour is composed of islands of squamous epithelium that lack stellate reticulum-like zones and peripheral palisading and also the epithelium of squamous odontogenic tumour lacks any evidence of malignancy, which helps in differentiating it from ameloblastic carcinoma. Acanthomatous ameloblastoma and keratoameloblastoma lack cellular features of malignancy and later consist of prominent keratinous cysts. Presence of atypical nuclear features and absence of any foci of metastasis rule out the possibility of malignant ameloblastoma.²

As this is a rare tumour, there is no consensus on treatment. In most of the reported cases, surgery followed by radiotherapy is the treatment of choice,^{12–13} however, few authors doubt the effectiveness of radiotherapy.^{14–15} Philip *et al*¹⁶ suggested to give radiotherapy in patients with positive resection margins and also in positive lymph nodes, extracapsular spread and perineural invasion. In our case, one of the resection margin showed tumour cell infiltration so postoperative radiotherapy was also given. Role of chemotherapy is not clear.¹⁷ Local recurrences may appear 5–11 years after definitive treatment.¹² Distant metastasis can be seen as late as 12 years postoperatively.¹² Lung is the most common site for distant metastasis followed by bone, liver and brain.^{12–13–17–18} So meticulous follow-up is

essential as chances of local recurrence and distant metastasis are very high.

Learning points

- Ameloblastic carcinoma is a rare neoplasm.
- Ameloblastic carcinoma and malignant ameloblastoma are two different entities.
- Patient of ameloblastic carcinoma should undergo a life-long follow-up in order to detect early local recurrence and distant metastasis.

Competing interests None.

Patient consent Obtained.

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