CASE REPORT

A delayed presentation of ameloblastic fibrosarcoma in an African patient

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Accepted 13 July 2017

SUMMARY

A 24-year-old woman presented with ameloblastic fibrosarcoma arising from ameloblastic fibroma. The delayed presentation accounted for the extensive destruction of the mandible and complete occlusion of her oral cavity. This resulted in an inability to eat and maintain oral hygiene. A multidisciplinary team management approach involved nutritional optimisation, segmental mandibulectomy, reconstruction with a reconstructive plate and a free anterolateral thigh flap to line the floor of mouth. Functional and aesthetic outcome was acceptable, and the patient is planned for secondary free fibular flap bony reconstruction.

BACKGROUND

Ameloblastic fibrosarcoma (AFS) is an extremely rare malignant tumour of odontogenic origin, with only 71 cases published until 2014.¹ None of these publications included African patients from Sub-Saharan Africa. The mean age of presentation is 27.3 years, with tumours occurring more frequently in the mandible compared with the maxilla.² Most cases arise de novo and one-third arise from benign lesions.³ This tumour displays both mesenchymal and epithelial constituents, with the mesenchymal component responsible for the sarcomatous nature of the lesion.³ Ameloblastic carcinosarcoma is different to AFS as it has a combination of carcinomatous and sarcomatous properties.¹ AFS has a high-reported recurrence rate of up to 37%, with two metastatic cases in the literature.⁴ Radical excision with long-term surveillance for recurrence is the recommended treatment of choice.⁴

CASE PRESENTATION

The patient is a 24-year-old woman, from a rural village in Limpopo province of South Africa, who presented to a Quaternary Academic Hospital with an oral tumour of 13 months duration. At the index presentation, the tumour was reported to be slow-growing and a biopsy, at a regional hospital, showed benign ameloblastic fibroma. Four months after the initial presentation, the patient noticed an

aggressive expansion of the tumour. Due to poor socioeconomic circumstances, the patient was only able to reach the regional hospital 10 months after presentation. A second biopsy confirmed malignant transformation of the tumour to ameloblastic fibrosarcoma. She reported weight loss of approximately 20 kg over 1-year period. She has no known co-morbidities and no personal or family history of any cancers. She does not drink alcohol, smoke or chew tobacco. She is unemployed, with grade 12 as the highest level of education obtained. The family is surviving on a disability grant.

Clinical examination revealed a large asymmetrical intraoral tumour that was extruding through the mouth (figure 1). The tumour surface was necrotic and bleeding intermittently. Her oral commissure, lips and cheek skin, although stretched, were not affected by the tumour. The left cheek was effaced up the subciliary level. The rest of the oral cavity examination was precluded by the size and extent of the tumour. The neck and supraclavicular area had no palpable lymph nodes. The rest of the systemic examination was normal.

INVESTIGATIONS

Haemoglobin was 7.3 g/dL. Renal and liver function tests were normal. The patient was negative on HIV testing. Her albumin was 23 g/L. CT scan of the neck, chest and abdomen were normal. CT scan of the head (figures 2 and 3) revealed extensive aneurysmal destruction of the left-lateral and medial segments of the mandible. Soft tissue CT views showed a large non-homogenous tumour, with cystic looking lesions and a fairly regular surface. Incisional biopsy was repeated and showed a poorly differentiated ameloblastic fibrosarcoma.

DIFFERENTIAL DIAGNOSIS

- ▶ Ameloblastic fibroma
- ▶ Ameloblastic fibrosarcoma
- ▶ Ameloblastic carcinosarcoma
- ▶ Ameloblastic fibro-odontoma
- ▶ Fibro-odontosarcoma
- ▶ Spindle cell carcinoma

TREATMENT

The tumour presented a mechanical obstacle to oral feeding, and her lack of nutrition was evidenced by her low albumin. After the repeat incisional biopsy was done, a 2-week lag period of definitive histological diagnosis presented an opportunity to

To cite: Chauke NY, Sofianos C, Liakos D, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220436

Figure 1 Presentation of the patient to our unit.

Figure 2 CT scan at presentation.

institute high-protein enteral feeds via a nasogastric tube. Preoperative optimisation also included blood transfusion.

The patient was staged as stage IIC. At the head and neck multi-disciplinary team (MDT) meeting, it was decided that a wide local excision without radiotherapy would be sufficient. Lower lip salvage segmental mandibulectomy of the central and left hemi-mandible with adequate margins would be initially reconstructed with a reconstructive plate based on a three-dimensional model of the resection (*figure 4*) and free anterolateral thigh (ALT) flap for intraoral lining.

Intraoperatively, a lip-splitting incision was chosen to allow adequate exposure, and a segmental mandibulectomy was performed. The specimen weighed 1 kg. The ALT flap was raised as per standard techniques, and the recipient vessels chosen were the left facial vein and artery. The large nature of the tumour had resulted in expansion of the adjacent soft tissues; external skin cover was, therefore, not necessary and the skin paddle of the flap was used as intraoral lining. At the conclusion of the procedure, the reconstruction was acceptable aesthetically (*figure 5*). The patient was admitted for monitoring to the intensive care unit for 48 hours postoperatively.

OUTCOME AND FOLLOW-UP

Healing in the postoperative period was problematic. Despite the preoperative feeding and optimisation, which was continued postoperatively, the patient experienced the following complications: lip commissure dehiscence, malocclusion, oral incontinence, reconstructive plate exposure and delayed decannulation of the tracheostomy. The commissure was successfully repaired without the need for any further flaps. The intraoral flap dehiscence recurred three times, and on the last attempt to repair the dehiscence, oral candidiasis was diagnosed; she was repaired and received a 2-week course of fluconazole. The repair remained intact thereafter. The patient was subsequently discharged when she could feed orally with soft foods. She was reviewed at 6 months with a good aesthetic result (*figure 6*); a result that the patient was satisfied with. Functionally, her feeding has remained limited to soft foods and her speech and swallowing were normal. A fibula osteocutaneous flap will provide dental rehabilitation once the patient has been deemed free of recurrence.

Figure 3 Three-dimensional reconstruction from CT scan.

Figure 4 Three-dimensional model and reconstructive plate used.

DISCUSSION

Ameloblastic fibrosarcoma (AFS) is a rare tumour that presents as a painful mass in the posterior mandible or maxilla that may ulcerate.⁵ Most of these tumours occur in the mandible.⁶ It is considered a low to intermediate grade malignancy, metastasises and involves lymph nodes infrequently but is locally aggressive.⁷ AFS occurs over a wide spectrum of ages (3–83 years) and shows a male preponderance.⁷

Imaging usually demonstrates a radiolucent, intraosseous lesion, with ill-defined borders.⁸ The CT scan of our patient showed a mass with a fairly regular border and was non-homogeneous with cystic spaces. Definitive diagnosis is histopathological, and several studies have shown that Ki-67 antigen, proliferative cell nuclear antigen (PCNA) and p53 protein are immunohistochemical markers that potentially aid in diagnosis of this tumour.⁵ Socioeconomic factors in our case that contributed to the delay in diagnosis include a low number of skilled personnel to diagnose and refer timeously, delayed/slow processing of biopsy specimens and the high number of poor patients who were unable to travel to nearest healthcare facility, which may be hundreds of kilometres away.

At present, a wide surgical resection with a margin of normal tissue is the best prognostic factor for AFS and recurrence, which leads to uniformly poor survival, if it occurs.⁹ The role, relevance and impact of adjuvant therapy for AFS has been debated. Goldstein *et al*, in 1976, presented a case of chemotherapy in treating ameloblastic sarcoma; however, a year after commencement, the patient developed a malignant melanoma and the chemotherapy was discontinued.¹⁰ More

Figure 5 Final inset of reconstructive plate (left) and patient at completion of procedure (right).

Figure 6 Six-month follow-up visit.

recently, two paediatric cases of chemosensitive AFS have been presented with a successful decrease in size and cell number of the tumour.¹¹ Adjuvant therapy has been previously reserved as a salvage treatment⁹; however, it may potentially be considered for advanced or large tumours. Healthcare personnel managing these patients should be cognizant of the high potential for airway compromise and the impact on feeding and nutrition when these tumours become very large.

Our patient had a reconstructive plate and anterolateral thigh flap (ALT) placed with a view to a second-stage bony reconstruction with a fibular flap, once she is deemed to be recurrence free. The ill-defined interval of recurrence and low number of reported cases of AFS make the decision for the timing of the second-stage reconstruction difficult. Khalili *et al* reported a case of maxillary AFS that had maxillectomy and postoperative radiation; the authors were satisfied with a 2-year follow-up period.⁶

The gold standard for secondary reconstruction of the bony defect to allow eventual dental rehabilitation, in our patient, would be the free fibula flap. Alternatively, Ferretti *et al* have convincingly shown that bone grafts may be used for mandibular reconstruction and allow osseointegration.¹² This approach is also a consideration in our patient as the ALT flap offers a well-vascularized bed for the bone grafts; adjuvant radiation is

not planned and the fibula and recipient vessels are preserved in the unfortunate eventuality of recurrence of the tumour.

In conclusion, definitive management of this locally aggressive tumour is difficult to establish due to its rare occurrence. The potential for ameloblastic fibromas to transform into AFS, mandates timeous referral and management by an MDT. Definitive bony reconstruction may be undertaken at the index extirpative procedure; however, a second-stage reconstruction after a recurrence-free interval is also acceptable. Future cases and further study will elucidate the role of adjuvant therapy in this condition.

Contributors NYC collected the data for the case report and prepared the first draft of the report. CS performed the subsequent write-up of the paper, referenced and prepared the images for inclusion. DL was the Consultant Plastic Surgeon performing the case and provided the articles and ultimate structure of the paper. EN provided support at all stages of the paper and his input was instrumental in the final revisions of the article.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- ▶ Early diagnosis, optimisation and management of such patients.
- ▶ There is a potential for malignant transformation of benign ameloblastic fibromas.
- ▶ A multi-disciplinary team approach is key to managing these patients.
- ▶ Stringent oral hygiene preoperatively and postoperatively is essential to minimise wound dehiscence.

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