AN AGGRESSIVE DESTRUCTIVE LESION OF THE MANDIBLE WITH INCONCLUSIVE HISTOPATHOLOGICAL DIAGNOSIS

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INTRODUCTION

This case highlights the challenges of managing a patient who presented with a pathological fracture from an aggressive lesion in the mandible, with no final histological diagnosis.

Initial clinical and histological results suggested chronic locally aggressive fibromatosis with treatment tailored around this working diagnosis.

Locally aggressive chronic fibromatosis is a rare benign tumour of the myofibroblasts which is:

- non-metastasising
- locally invasive¹

Fibromatoses are sub-divided into:

- Superficial; palmar, plantar and abdominal wall²
- **Deep**; intra and extra-abdominal tumours³

CASE DESCRIPTION

60 y.o. diabetic male was referred with a 5 month history of:

- Pain, buccal swelling, trismus, weight loss
- No improvement following extraction of LR7
- Extra-oral examination:
- Swelling the ramus of the right mandible, level II lymph node, mouth opening 25mm

Intra-oral examination:

- Palpable mass in the right buccal sulcus region of the grade III mobile LR8 tooth
- Exposed bone in the previous extraction socket of LR7
- Mobility of the mandible anterior segment

Due to the aggressive and suggestive malignant presentation urgent investigations and biopsies were performed.

INVESTIGATIONS

- Orthopantomogram: pathological fracture (figure 1)
- CT: large destructive soft tissue mass of right ramus
- MRI: mass consistent with possible malignancy and invasion of right masticator space
- Biopsies: oral and ultrasound transfacial open biopsies



Figure 1- Orthopantomogram showing pathological fracture of right angle and ramus of mandible with irregularity and destruction of the coronoid process **Authors** have no conflict of interest to declare References¹ Fielder C. Theory, K. derr

<u>HISTOLOGY</u>

Histopathology showed chronic inflammatory tissue but no evidence of neoplasia. Appearance were suggestive of a fibroproliferative lesion such as fibromatosis.

MANAGEMENT

Following MDT discussion with regional head and neck units a working diagnosis was made of:

Locally aggressive chronic fibromatosis (unknown aetiology) involving right condyle and angle of the mandible with pathological fractures of the mandible. Recommendation for:

Wide local excision of the tumour and mandibulectomy with composite fibular free flap reconstruction (figure 2) for treatment and further specimen for further histology.



Figure 2- Orthopantomogram following hemimandibulectomy and fibular free flap reconstruction

DIAGNOSTIC CHALLENGES + DISCUSSION

Final histology, with additional input from a specialist sarcoma unit was unable to confirm a definitive diagnosis.

Results were suggestive of an unusual reactive fibrosis with differentials including;

- Squamous cell carcinoma (arising from a salivary duct and/or oral mucosal epithelium)
- Odontogenic cyst
- IgG4 disease
- Inflammatory myofibroblastic tumour

Due to the inconclusive histology, close follow-up is essential due to local or distant recurrence of this unknown pathology.

In conclusion, this case presented a complex clinical presentation and pathogenesis making a standardised treatment pathway difficult. This highlights the importance of multi-disciplinary and multi-unit co-ordination.

Authors have no conflict of interest to declare References: Fisher C, Thway K. Aggressive fibromatosis. Pathology. 2014 Feb:46(2):135-40. doi: 10.1097/PAT.000000000000045. PMID: 24378386. - Walker, Eric A et al. "Imaging features of superficial and deep fibromatoses in the adult population." Sarcoma vol. 2012 (2012): 215810. doi:10.1155/2012/215810; "Steven D. Billings, John R. Goldblum, in