

Desmoplastic Fibroma: a benign neoplasm mimicking a malignancy

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CASE

A four-year-old African male patient presented to our clinic with a two-month history of a firm, warm lesion involving the right side of his face. The lesion presented as a bleeding papillary mass in the 85/46 region that extended to the floor of the mouth, causing displacement of the tongue (Figure 1A-B). Upon cone-beam computed tomography (CBCT) examination, an expansile homogenous intermediate density lesion was noted with complete destruction of the bucco-lingual cortical bone. Discernible extensions were identified in the anteroposterior plane from the region of the 84 to the posterior mandible involving the right corpus, developing 46, angle and ramus but spared the condyle and coronoid processes. The lesion extended inferiorly below the epiglottis and showed a reduction of the hypopharynx. Marked bucco-lingual expansion with soft tissue swelling was noted (Figure 2A-D). An incisional biopsy was performed under general anaesthesia, and histological examination rendered a diagnosis of desmoplastic fibroma (DF).



Figure 1: (A) Clinical photograph of the patient showing facial swelling and asymmetry on the right. (B) Intra-oral photograph of the patient showing a papillary, expansile lesion in the region of the 46 causing displacement of the tongue.

INTERPRETATION

Aetiology and pathogenesis

Desmoplastic fibroma is a rare benign tumour of connective tissue origin with low to variable cellularity. The tumour is a locally aggressive, invasive, benign neoplasm accounting for less than one percent of bone tumours.¹ The aetiology of DF is still unknown.² DF is usually similar to other intra-osseous fibrous lesions that mainly occur in the mandible (69.4%) followed by the femur (15%), pelvis (13%), radius (12%) and tibia (9%).^{1,3,4} The lesion commonly presents as a painless swelling that causes facial asymmetry, tooth displacement and mobility.²

Clinical presentation

DF generally occurs during the first two decades of life with a mean age of 16 and an equal male-to-female ratio.¹ It has been reported in patients as young as one year old. Clinically, most DFs present as painless, slow-growing swellings.⁵ A differential diagnosis of DF is difficult purely based on clinical and radiological examination and, therefore, histopathological examination is required for a definitive diagnosis.¹ Histologically, DF generally resembles a soft tissue desmoid tumour.⁵ According to the World Health Organization (WHO), the pathognomonic histological

features for DF are the presence of mature fibrous connective tissue, spindle-shaped fibroblasts (that are separated by abundant collagen fibres) and the absence of cellular mitoses or atypia.⁶ It is important to distinguish it from other benign spindle cell tumours such as non-ossifying fibroma, myofibroma, odontogenic fibroma, fibrous dysplasia and low-grade fibrosarcoma.¹

Diagnosis

DF is a very challenging tumour to diagnose due to the slow onset and unremarkable/variable findings. The radiographic features of DF are not very specific.¹ It may present as a uni- or multilocular lesion with well or ill-defined

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borders.⁵ Additionally, cortical expansion, perforation or destruction (because of rapid expansion) may cause new bone formation with a “sunburst” appearance. These features may then be mistaken for osteogenic sarcoma or Ewing’s sarcoma.² It commonly presents with a so-called soap bubble appearance and may be misinterpreted as an ameloblastoma, aneurysmal bone cyst or odontogenic myxoma.¹ Diagnostic imaging is a vital step in assessing bone lesions. Panoramic radiography is usually the first imaging modality, but three-dimensional imaging, like CBCT, CT and MRI, are the imaging modalities of choice if the lesion presents with suspected cortical perforation and extra-osseous growth.²

Treatment

Due to the aggressive nature and high recurrence rate, the treatment of choice is complete surgical resection with wide margins or enucleation in selective cases.⁴ The recurrence rate is between 40-47% if the lesion is treated via conservative methods such as curettage. Radiation therapy can be used if the lesion is close to vital structures but should be avoided in children due to post-operative complications.¹ Long-term follow-up will be needed for all patients with DF.⁴ Bone reconstruction is essential for patients’ quality of life.²

CONCLUSION

DFs can present very early in life, grow rapidly and have a destructive nature to the extent that they can mimic a malignancy; and thus it is important to manage appropriately. Early diagnosis, comprehensive clinical and radiographic examinations, adequate biopsy site selection, histological

evaluation and appropriate treatment can result in a better prognosis and expectation for the patients.²

DECLARATIONS

Conflict of interest

The authors declare that they have no conflict of interest.

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Ethics approval

This study was approved by the University of Pretoria Ethics Committee (Reference no: 456/2023). All procedures followed the ethical standards of the Helsinki Declaration of 1975, as revised in 2008.

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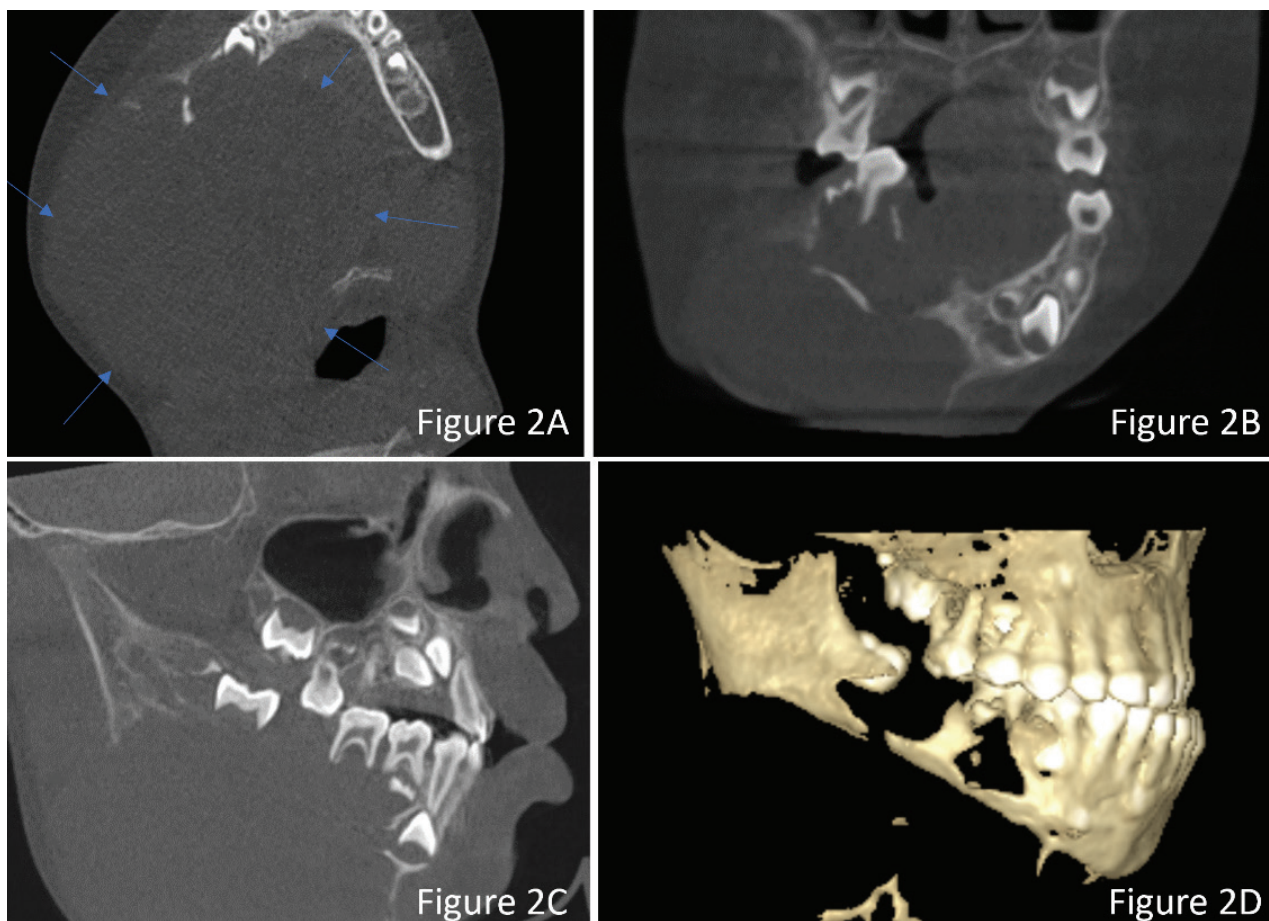


Figure 2: CBCT multiplanar reformatting showing the lesion in the right mandible with expansion, cortical perforation and destruction throughout the entire lesion, extending posteriorly into the corpus, angle and ramus but excluding the condyle and coronoid. (A) Axial view of the lesion with the blue arrows indicating the margins of the lesion. (B) Coronal, (C) sagittal and (D) three-dimensional reconstruction of the lesion indicating the extent of destruction.