Case Report

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A Challenging Mandibular Tumour – Desmoplastic Fibroma: Case Report

Parimala Kumar*, Laxmish Kumar, Patrick J. McCann***, Andrew Burns******

Abstract

Desmoplastic Fibroma is a rare Mandibular tumour, only a few cases have been reported in the literature. It is a locally aggressive tumour with a high rate of local recurrence if not completely excised. A 21 yr old male consulted our department with a six month history of slowly growing swelling left mandible. He also had a complaint of complete numbness of the left lower lip and chin. He had a large radiolucent lesion on the left angle and ramus of the mandible measuring 100x 80 mm in dimension. Further investigations included biopsy and CT scans.

It was a solid fibrous tumour perforating the cortical plates. Histopathological examination confirmed Desmoplastic Fibroma. He had resection of the tumour by hemimandibulectomy and resection of the left condyle. It was reconstructed with a left fibula graft and temporomandibular prosthesis.

Keywords: Aggressive tumour; Bone; Desmoplastic fibroma; Mandible; Numbness to lip.

Introduction

Desmoplastic fibromas are benign but locally aggressive neoplasm of the bones.[1,2] Intraosseous Desmoplastic fibromas are even rarer (far less than 1% of all bone tumours). In 22% of these cases the mandible was involved. The incidence is equal in male and female patients.[2] On average, the age was 15.1 years at the time of diagnosis.[2]

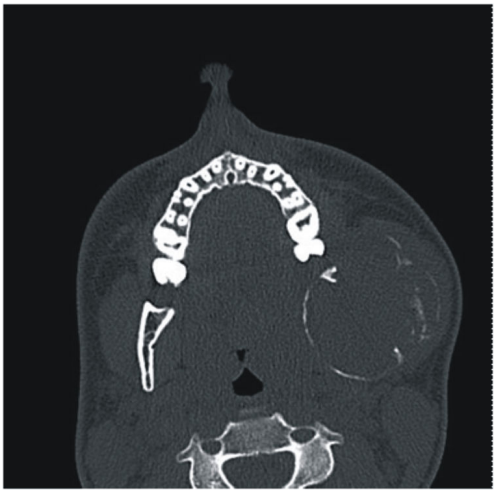
Surgical resection and radiotherapy have been suggested for treatment, with wide surgical resection being the most favoured option.[2,3,4,5]

In this report we highlight the challenge these cases present to the clinician for diagnosis and to decide on the appropriate treatment for the best outcome for these young patients.

Case Report

A 21 year old Caucasian male was referred by his general medical practioner with a slow growing swelling of the left mandible. He was a healthy patient with no significant past

Figure 1: CT Scan Showing the Expansion of Desmoplastic Fibroma in the Left Mandible

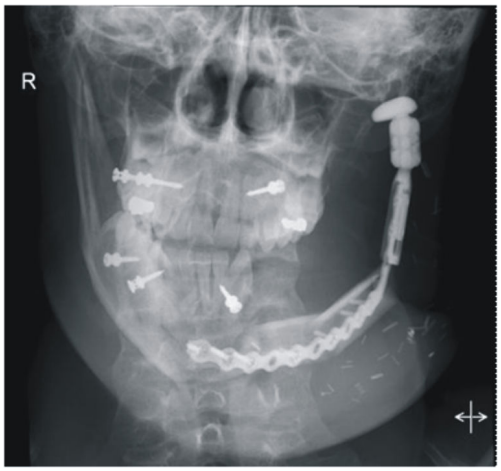


Author's Affilation: *Assistant Professor, Dept. of Periodontics, Post Graduate, Oral and Maxillofacial Dept., AJ Shetty Dental College, Mangalore, Karnataka, India, ***Consultant Oral and Maxillofacial Surgeon, University Hospital, Galway, Ireland, ****Consultant Oral and Maxillofacial Surgeon, Sunderland Royal Hospital, United Kingdom.

Reprints Requests: Dr. Parimala Kumar, Flat 302, Third floor, Manjunath Residency, Kapikad–Kottara Cross, Mangalore, Karnataka, India.

E-mail: raodrparimala@rediffmail.com

Figure 2: Radiograph of the Mandible Showing the Reconstruction of the Left Mandible Using TMJ Prosthesis and Recon Plate



medical history. He was a hurling player and there was a history of trauma to the left side of the jaw 6-8 months prior to his first consultation with us.

This swelling was noticed by the patient immediately after trauma, but the patient decided to wait and see if it would subside. He also had complete anaesthesia in the territory of the left inferior alveolar nerve. On clinical examination he had good mouth opening, his occlusion was unaltered. The swelling was non tender and hard on palpation. It was the size of a tennis ball centred in the left ramus. The overlying skin was normal.

There was no crepitus of bone felt on palpation. He had definite numbness in the distribution of the left inferior alveolar nerve, this was tested by pin prick and light touch. There was no evidence of swelling intraorally and he had no problems with his swallowing or airway.

Panoramic radiography revealed a unilocular radiolucent lesion on the left angle and ramus of the mandible measuring 100x 80 mm. There was a carious lower left second molar in association with the swelling. This was another factor to be considered in the differential diagnosis. The CT scans revealed an expansile lesion in the left mandible resulting in cortical plate expansion both buccally and lingually with areas of perforation.

Initial management involved aspiration of the swelling to rule out any vascular lesion. Exploration of the swelling and biopsy under general anaesthetic was then carried out.

The lower left second molar was extracted at this time. The tumour was a solid firm pink lump with perforation of the cortical plate. Initial histopathology report suggested Neurolemmoma. A second opinion was sought from a tertiary centre as the tumour had a peculiar presentation and grew rapidly unlike a neurolemmoma which is typically slow to grow. It was reported as Desmoplastic fibroma the second time after being discussed in the histopathology multidisciplinary meeting.

Treatment

Based on the literature review and multidisciplinary meeting discussions it was decided to resect the mandible enbloc to obtain good clearance of the tumour. As the patient was young the mandible was reconstructed with a left fibula graft and the joint reconstructed with an artificial TMJ prosthesis. His facial nerve function was intact after the operation and he retains near normal movements and function of the mandible.

Histology

Histological sections showed a well circumscribed round lesion having thin shell like bone trabeculae at the periphery. The lesion was composed of closely packed fascicles of fibroblasts showing no atypia or mitotic activity. Between the fibroblasts, variable amount of collagen fibres could be seen and scattered tiny bone trabeculae were also present. No histological signs of malignancy were noted. Excision was complete.

Results of immunohistochemistry

- S100: Negative

- Desmin: Negative
- Actin: Approximately 15% of the cells were positive.
- H-caldesmon: Negative
- Ki-67: <2%

Elastica Van Gieson special stain highlights the large amount of collagen fibre within the lesion. The immunohistochemistry and the special stain confirmed the diagnosis of Desmoplastic fibroma.

Discussion

The present case was unique in the sense that it was a large nonpainful, unilocular lesion that did not lead to any migration or loss of teeth. It was however similar to other cases reported in that it presented in a young person, typical site and was locally aggressive.

The use of adjuvant radiotherapy prior to surgery to reduce the size and in turn reduce morbidity from temporomandibular joint loss is debateable, as is the use post surgery which may lead to secondary neoplastic lesions in this age group. However as there is a high chance of recurrence a wide resection with good margins is the best treatment to render the patient tumour free.

The challenges in diagnosing these lesions accurately due to the similarity in clinical and radiological pictures to ameloblastoma, odontogenic fibroma, aneurysmal bone cyst, haemangioma and histologically to fibrous dysplasia, low grade intraosseous sarcoma, fibrosarcoma is considerable. A multidisciplinary approach is the best way forward for such rare cases.

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