Gnathic Desmoplastic Fibroma Mimicking a Dentigerous Cyst: A Case Report

Vimal Kalia · Supreet Kaur · Deepti Vashisht

Received: 24 December 2009 / Accepted: 24 April 2010 / Published online: 16 May 2012
© Association of Oral and Maxillofacial Surgeons of India 2012

Abstract Desmoplastic fibroma (DF) is a rare, well-differentiated fibrous tumor, which is slow in growth but has an aggressive potential. In its clinical and radiographic presentation it mimics many unusual pathologies of the jaws. This case report presents a DF in the mandible, which in its clinical presentation and radiographic appearance mimicked a dentigerous cyst highlighting the importance of detailed histopathological examination in the diagnosis of jaw pathologies.

Keywords Desmoplastic fibroma · Mandible · Dentigerous cyst

Introduction

Desmoplastic fibroma (DF) is a rare myofibroblastic, locally aggressive but non metastasizing tumor that was first described by Jaffe [1] in 1959 and that it histologically resembles a desmoid tumor of soft tissue.

Griffith and Irby [2] in 1965 first reported a case in the jaw and after that numerous individual cases have appeared in the literature.

Desmoplastic fibroma is a lesion which does not have specific clinical appearance and radiographic features.

Desmoplastic fibroma often mimics other common as well as unusual pathologies of the jaws including

ameloblastoma, odontogenic myxoma, aneurysmal bone cyst, chondromyxoid fibroma, central hemangioma, and eosinophilic granuloma [3].

Desmoplastic fibroma presenting as periapical pathosis in association with an endodontically treated tooth [4] and in an inter-radicular position at presentation [5] have also been reported.

The following is a report of a case of DF of mandible, which clinically and radio-graphically mimicked a dentigerous cyst.

This highlights the fact that a variety of pathologies occur in the jaws, which have similar clinical features and radiographic appearances, and histopathological examination alone reveals the true nature of the lesion.

Case Report

The patient was 17-year-old male who presented to the oral and maxillofacial clinic with a complaint of facial asymmetry. The facial asymmetry was because of a rapidly expanding, painless mass along the left angle and the body of mandible since 6 months.

On intraoral examination, there was a buccal and lingual cortical expansion of the retro-molar area, alveolar bone, and base of the mandible in the 37, 38 area. 37 and 38 teeth were absent and 36 was slightly mobile.

The overlying mucosa was of normal color and texture but was attached and gave a feeling of soft tissue fullness directly overlying the bony lesion.

Panoramic radiograph (Fig. 1) revealed an irregular radiolucent radio-opaque lesion with some trabeculation within and displayed a sclerotic border. There was no apparent resorption of roots. The CT scan (Fig. 2) revealed enlargement and ballooning of lower part of the left

V. Kalia · S. Kaur · D. Vashisht
Oral and Maxillofacial Surgery, BRS Dental College, Panchkula,
India

V. Kalia (✉)
H. No. 731, Sector 2, Panchkula, India
e-mail: vimal@kalia.in

Fig. 1 Panoramic radiograph showing lesion of left side

Fig. 2 CT scan showing enlargement and ballooning of lower left mandibular region

mandibular region with the incorporation of teeth and muscle/soft tissue within it. From the clinical presentation and radiographic analysis, location of the lesion, incorporation and displacement of the teeth, a provisional diagnosis of a dentigerous cyst associated with 37 tooth was made.

A biopsy was taken from the lesion, which revealed the presence of DF. After the histological confirmation of DF, under local anesthesia an incision was given extending from the third molar region up to the second pre-molar region and the fibrous non encapsulated lesion was

Fig. 3 The resected specimen

Fig. 4 The six month postoperative radiograph

enucleated (Fig. 3). The 37 and 38 teeth were also carefully elevated out from the lower border. The wound was closed with open packing. Patient was called for weekly follow up visits and radiographic examination was done after 6 months to evaluate bone formation (Fig. 4).

The histo-pathological examination (Fig. 5, 6) revealed parallel, inter-lacing bundles of fibroblastic proliferative growth of moderate cellularity and nuclear hyperchromatism. The spindle cells appeared plump in areas resembling myofibroblast. The lesion showed moderate vascularity and the tumor had no epithelial component. There was no evidence of anaplasia and tumor cells showed no mitotic activity. The findings were compatible with DF of the mandible.

Discussion

In the head and neck, fibromatosis may be intra osseous (DF) or more often, in soft tissue, with the highest incidence in the supra clavicular region of the neck [6].

Fig. 5 The photomicrograph showing parallel interlacing bundles of fibroplastic proliferation

Fig. 6 The histopathological picture

These tumors reside in a clinically gray zone between benign fibrous lesions e.g., Keloids and malignant tumors. This is reflected, in part, by various synonyms for desmoid fibromatosis: desmoma, aggressive fibromatosis, fibrosarcoma grade 1, desmoid type, and DF of bone [7].

Any bone may be involved with DF and in a study of 184 cases mandible (22 %) was the most common site of involvement [8].

Desmoplastic fibroma of the jaw presents in the same manner as its counter part in long bones. The age incidence being first, second, and third decade with no predilection for any sex [9], whereas the dentigerous cyst is by far the most common non-inflammatory cyst of the oral region [10].

Dentigerous cyst occurs most often in males in second decade of life and are associated with impacted third molars [11] as was seen in this 17-years-old male patient leading to a clinical diagnosis of dentigerous cyst.

The presentation of gnathic DF is generally non pathognomonic hence the diagnosis with certainty cannot be

reached. The DF may present as an ameloblastoma, odontogenic myxoma, aneurysmal bone cyst, chondromyxoid fibroma, central hemangioma, and eosinophilic granuloma [3].

On similar grounds dentigerous cysts are clinically asymptomatic but have a potential to become extremely large, cause cortical expansion, and erosion [12, 13] as was the clinical presentation of this case.

The radiographic investigation also revealed an osteolytic lesion of the mandible. But diagnosis could not be made accurately as in this case the appearance mimicked a dentigerous cyst due to the displacement of 37 and 38 to the lower border of the mandible.

Osteolytic lesion like gnathic DF needs to be differentially diagnosed from not only a dentigerous cyst but also giant cell reparative granuloma, aneurysmal bone cyst, odontogenic keratocyst, and fibrous dysplasia.

Dentigerous cyst typically appears as a well-circumscribed unilocular usually symmetric radiolucency around the crown of impacted tooth [13].

Giant cell reparative granuloma, like DF extends to the sub-chondral bone, but it is round rather than oval [14].

Aneurysmal bone cysts usually has an eccentric “blow-out” appearance as opposed to the fusiform in DF [14].

Odontogenic keratocyst is usually centrally located with a scalloped border and thin marginal sclerosis [14].

Fibrous dysplasia contains a mineralized matrix and a sclerotic rim [14].

In comparison DF has non-specific radiographic features including unilocular or multilocular well-demarcated or irregular radiolucency with variably expressed marginal sclerosis [15]. Hence it mimics common and uncommon jaw pathologies. Being an aggressive lesion it usually may disrupt the cortex as is evident in this case in the CT scan image. Bohn et al. [8] found the soft tissue extension in 50 % cases.

This particular case report is unusual as 38, 37 teeth were displaced by the DF to the lower border of the mandible and the radiographic picture simulated a dentigerous cyst.

Gross appearance of the tumor was that of a firm, rubbery, white, non-encapsulated, fibrous growth that made evident that it is not a dentigerous cyst. Another interesting feature was that the growth was enucleated enmasse and the teeth were not found included in the specimen. Rather they were found displaced to the lower border of mandible and had indented the fibrous mass again making the provisional diagnosis of a dentigerous cyst questionable.

Histological appearance is the only diagnostic criteria for DF, which shows mature fibrous tissue consisting of small sparse fibroblast with little or no mitotic activity. Absence of capsule

- Poor demarcation within the bone
- Absence of osteoid or osseous metaplasia which make it easy to distinguish from fibrous dysplasia or other fibro osseous lesion.

A variety of treatment modalities have been used for DF. Curettage, enucleation, wide local excision, peripheral ostectomy, and resection with a wide margin have all been included. In this case as the lesion was growing within the reactive bony rim, it was enucleated.

Iwai et al. [16] have reported 20–40 % recurrence in cases treated with simple excision or enucleation and 70 % in cases treated with curettage.

Radiation therapy [17] and chemotherapy with or without additional procedures [7] were also suggested as non-surgical modalities of treatment but are not successful.

Thus surgical approach is the preferred approach for DF and should be based on size, location, and the histological appearance. Lastly long term follow up is mandatory.

In conclusion, we reiterate that DF is a rare, well differentiated fibrous tumor with a slow but aggressive potential for growth which is incapable of metastasis and should always be included in the differential diagnosis of radiolucent pathologies of jaws which involve teeth because DF can mimic jaw pathologies like a dentigerous cyst.

References

1. Jaffe HL (1958) Tumors and tumorous conditions of the bones and joints. Lea & Febiger, Philadelphia, p 298
2. Griffith JG, Irby WB (1965) Desmoplastic fibroma: report of a rare tumor of the oral structures. *Oral Surg Oral Med Oral Pathol* 20:269–275
3. Hoipkins KM, Huttula CS, Kahn MA, Albright JE (1996) Desmoplastic fibroma of the mandible: review and report of two cases. *J Oral Maxillofac Surg* 54:1249
4. Valente G, Migliario M, Bianchi SD, Vercellino V (1989) Desmoplastic fibroma of the mandible: a case with an unusual clinical presentation. *J Oral Maxillofac Surg* 47:1087–1089
5. George DI, Gould AR, Miller RL, Strull NJ (1985) Desmoplastic fibroma of the maxilla. *J Oral Maxillofac Surg* 43:718–725
6. Batsakis JG, Raslan W (1994) Pathological consideration, extra-abdominal desmoid fibromatosis. *Ann Otol Rhinol Laryngol* 103:331–334
7. Ayala AG, Ro JY, Geopfert H, Cangir A, Khorsand J, Flake G (1986) Desmoid fibromatosis: a clinicopathologic study of 25 children. *Semin Diagn Pathol* 3:138–150
8. Bohm P, Krober S, Greschniok A, Loniado M, Kaiserling E (1996) Desmoplastic fibroma of the bone: a report of two patients, review of the literature, and therapeutic implications. *Cancer* 78:1011–1023
9. Kwon PH, Horswell BB, Gatto DJ (1989) Desmoplastic fibroma of the jaw: surgical management and review of literature. *Head Neck* 11:67–75
10. Daley TD, Wysocki GP, Pringle GA (1994) The relative incidence of odontogenic tumors: oral and jaw cysts in a Canadian population. *Oral Surg Oral Med Oral Pathol* 77:276–280
11. Bernick S (1949) Dentigerous cysts of the jaw. *Oral Surg Oral Med Oral Pathol* 2:914–921
12. Killey HC, Kay LW, Seward GR (1977) Benign cystic lesions of the jaw: their diagnosis and treatment, 3rd edn. Churchill Livingstone, Edinburgh, pp 93–103
13. Shafer WG, Hine MK, Levy BM (1983) A text book of oral pathology, 4th edn. WB Saunders, Philadelphia, pp 260–265
14. Shi H, Wang P, Wang S, Yo Q (2008) Desmoplastic fibroma of the mandible. *Dentomaxillofac Radiol* 37:408–411
15. Fisker AV, Philipsen HP (1976) Desmoplastic fibroma of the jawbones. *Int J Oral Surg* 5:100–103
16. Iwas S, Matsumoto K, Sakuda M (1996) Desmoplastic fibroma of the mandible mimicking osteogenic sarcoma: report of a case. *J Oral Maxillofac Surg* 54:1370–1373
17. Green TL, Gaffney E (1981) Desmoplastic fibroma of the mandible. *J Oral Med* 36:47–49