DESMOPLASTIC FIBROMA OF RAMUS OF MANDIBLE – A RARE CASE REPORT

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ABSTRACT

Desmoplastic fibroma (DF) is a rare, benign fibrous tumor of the bone which is locally aggressive. Desmoplastic fibroma forms 0.3% of the benign osseous tumors, which most commonly occurs in the tibia, scapula, and femur. Most commonly affected site in the head and neck region is Mandible. Desmoplastic fibroma causes bone destruction and has a high tendency for local recurrence. In this case report, we present desmoplastic fibroma of mandible of 5 year old female patient with imaging, histopathology, treatment and discussion about prognosis.

Key words: Locally aggressive, Fibroma, Non metastatic, Desmoplastic, Recurrence

INTRODUCTION

Desmoplastic fibroma is a rare locally aggressive non metastatic benign fibrogenic tumor of the bone. Desmoplastic fibroma of the bone is considered to be the intraosseous counterpart of the common soft-tissue desmoid or fibromatoses. In 1958 Jaffe reported five cases occurring in the tibia, scapula and femur. This tumor constitutes less than 1% of the bone tumors and 0.3% of benign osseous tumors, which usually involve the tibia, scapula, and femur. Desmoplastic fibroma causes bone destruction with a propensity to invade the soft tissues if untreated and has a high tendency for local recurrence if inadequately treated was reported in the year 2013. In the head and neck region, the most commonly affected site is the mandible. Desmoplastic fibroma of the mandible was first reported by Griffith and Irby in 1965.

CASE REPORT

A female patient aged about 5 years reported to the outpatient Department of Oral medicine and radiology, with a complaint of swelling in right side of face for past 6 months. Patient gives history of painful swelling in the same region two years back which subsided spontaneously within two days without taking any medications. Presently the patient noticed swelling recurred on the same site which was initially smaller in size and gradually increased to present size within 6 months period. On extra oral examination a diffuse swelling seen in the right side of lower part of face approximately measuring about 4×3 cm in size which is extending superiorly 4 cm from lower eyelid, inferiorly it crosses the inferior border of the mandible and extends to submandibular region, anteriorly 2.5 cm away from the commissure of the lip and posteriorly 1 cm anterior to tragus of the ear. On palpation the inspectsory findings were confirmed. The swelling is non tender, firm to hard in consistency. On intra oral examination a single diffuse swelling seen in relation to 85 obliterating buccal vestibular region, which is approximately measuring about 3×3 cm in size. Overlying mucosa appeared to be normal in color. On palpation it was firm to hard in consistency with mild tenderness and no secondary changes noted. Correlating the patient history and clinical findings a differential diagnosis...
of ameloblastoma, odontogenic keratocyst, and aneurysmal bone cyst was given. Further investigations like complete hematological and radiographical evaluation were performed. OPG revealed multilocular radiolucency involving right ramus and body of mandible and the radiolucency extended superiorly upto sigmoid notch, inferiorly it extended to the angle of mandible, also revealed break in the continuity of inferior border mandible. Tooth crypt of 47 displaced superiorly. Root resorption of erupting 46 evident (Fig-1). CT revealed 4.4x2.9x3.7cm expansile, multiloculated soft tissue density evident in right ramus with cortical bone discontinuity involving the inferior, buccal and lingual cortex (Fig-2). 3D reformatted image showed buccal cortical expansion with buccal and inferior cortical breach (fig-3). Working diagnosis of ameloblastoma was arrived and considering age of patient curettage of the lesion was performed under G.A and the specimen was sent for histopathological evaluation (fig-4).

Fig 1: Preoperative orthopantomogram

Fig 2: Contrast enhanced Computed Tomography showing expansile, soft tissue density lesion in right ramus with cortical bone discontinuity in lingual side

Fig 3: CT 3D Reformatting

Fig 4: Photograph of surgical specimen

On histological examination revealed the lesion showed plump spindle shaped fibroblasts cells arranged in short and long fascicles, focal storiform pattern and intervening bands of collagen bundles, there is no atypia or increase in mitosis suggestive of Desmoplastic fibroma (fig-5).

Fig 5: Photomicrograph showing plump spindle shaped fibroblasts cells (40X)
Fig 6: Postoperative orthopantomogram

DISCUSSION

In greek the term desmos means band or ligament. In 1938, the German physiologist and anatomist Johannes Muller characterized the term desmoids. Desmoid tumors were localized in the abdominal wall and the intraosseous variant is the desmoplastic fibroma. Jaffe in 1958 reported a similarity between the intraosseous lesions and the desmoid tumor of the abdominal wall [6]. Desmoplastic Fibroma is a non-metastasizing, often locally aggressive neoplasm with normal appearing fibroblasts. In 2002 Desmoplastic Fibroma was defined as a rare benign bone tumor consisting of spindle-shaped cells along with minimal cytological atypia and excess collagen production[7]. Possible etiologies were related to endocrine [8], genetic factors [9] and trauma[10,8]. Dahlin and Unni recorded only 9 case of Desmoplastic Fibroma in a series of 8542 primary bone tumours. Bohme et al reviewed 191 cases of Desmoplastic Fibroma reported in 80 publications. In their review, the age of patients ranged from 15 months to 75 years, with a reported mean age around 23 years [11,12]. The age incidence is in the first, second or third decade. There is no specific gender predilection [13]. In jaw bones desmoplastic fibroma occurs predominantly in the mandible and the maxilla is rarely affected. The ramus, angle and molar area of the posterior mandible are frequently involved. Less frequently affected areas are the premolar area and the anterior segments [14]. Radiographically, the tumor presents as a well-defined, expanding, osteolytic, radiolucency, either unilocular or multilocular, and the cortex is perforated in some areas, with an associated soft tissue mass. Our patient had similar features of a multilocular osteolytic lesion with corticated borders with cortical perforation in the inferior, Buccal and lingual cortex [15]. The differential diagnosis of an osteolytic lesion in the mandible includes ameloblastoma, odontogenic keratocyst and fibrous dysplasia, aneurysmal bone cyst, Ameloblastoma occur at 40 years of age, radiographically may appear as soap bubble, honey comb or tennis–racket pattern. Odontogenic keratocyst is usually centrally placed, with a scalloped border and thin marginal sclerosis. Aneurysmal bone cyst is a false cyst which occurs as unilocular or multilocular radiolucency which frequently balloons out of the cortex as opposed to the fusiform expansion usually seen with desmoplastic fibroma [16]. Fibrous dysplasia is similar but would also show typical patterns of woven bone and contains a mineralized matrix and often has a sclerotic rim [6]. Low-grade fibrosarcoma is a main concern to be ruled out, because of its aggressive, malignant nature with spindle cells, increased mitotic activity and pleomorphism [17]. Desmoplastic fibroma can be diagnosed only from tissue evaluation. If cortical expansion is present, a few other lesions can be included such as eosinophilic granuloma, arteriovenous malformations and hemangiomas. Central hemangioma may cause loosening and migration of teeth and teeth demonstrate rebound mobility when depressed into sockets. Diverse medical and surgical treatment options have been recommended for desmoplastic fibroma, which are simple curettage, segmental resection, en block Resection, radiotherapy, and chemotherapy, with or without additional surgical procedures [4]. The major characteristic of desmoplastic fibroma is increased rate of local recurrence. Recurrence rate is at least 40% if treated by curettage or intralossional resection [18,19]. Depending on the affected area and its aggressive nature, the management is decided. In cases of intraosseous lesions without any evidence of extension to the adjacent soft tissue and also when there is high risk associated with resection because of anatomical conditions, risk-benefit-analysis can be carried out and curettage is considered as adequate management because it reduces operation time along with lower risk of infection and hence facilitates faster recovery. Radiation therapy is not recommended because of its potential for transformation of this lesion into fibrosarcoma [6]. In our case, treatment options were discussed and curettage was decided. Due to higher risk of
recurrence, postoperative observation is mandatory, including clinical and radiographic examinations, considering the child’s age and also as the condyle is one of the main sites in post natal growth of mandible curettage was performed and the patient was kept under observation. Patient reviewed 2 months after surgery and also 6 months once regular follow up was done (fig-6).

CONCLUSION

Desmoplastic fibroma is a rare, benign, locally aggressive, intraosseous lesion, with a high local recurrence rate. The tumor should be resected when ever feasible or curettage done with regular postoperative follow up.

Conflict of Interest: Nil

REFERENCES