Case report

"BENIGN FIBROUS HISTOCYTOMA OF SOFT TISSUE : A CASE REPORT AND REVIEW OF LITERATURE".

ABSTRACT :

Fibrous histiocytoma is a heterogenous group of mesenchymal tumors ,composed of fibroblasts & histiocytes. The common age of occurrence is fifth decade with male predilection . The most common site is on skin of extremities. Intraorally BFHs are commonly seen in soft tissues of buccal mucosa, gingiva, lips, soft palate & floor of mouth. Radiographically BFH shows buccolingual expansion with multilocular radiolucency with sclerotic rim. Microscopically lesion is composed of elongated, spindle-shaped fibroblasts arranged in fascicles and whorls pattern . Considering unusual site of tumor in this case, immunohistochemistry was performed to confirm diagnosis.

Keywords: Benign fibrous histiocytoma, mandible , storiform pattern

INTRODUCTION :

Benign fibrous histiocytoma (BFH) is a mesenchymal tumor composed of fibroblasts and histiocytes . Arises in cutaneous and non cutaneous soft tissues . Cutaneous BHF commonly originate in sun exposed areas of skin. Non cutaneous BFH comprises 1 % of all BFH.[7] It was first described by Stout and Lattes in the year 1967 as a soft-tissue neoplasm, most commonly seen in the skin as solitary,

slow-growing nodule, which targets mid-adult life age group.[1] We report one such case of BFH occurring in left mandibular region in a 9-year-old female which was asymptomatic since 6 months .[4]

CASE REPORT :

A 9 year old female patient was reported to the outpatient department (OPD) with chief complaint of painless swelling in the left lower back gum region for 5-6 months. The swelling was of gradual onset, slowly progressive and was not associated with pain or discharge. Medical history revealed congenital acynotic heart disease. Family history was noncontributory. On general physical examination no other abnormalities detected . No history of trauma to the jaw, any recent dental procedures.

Extraoral examination revealed no abnormality . Intraorally , a well−defined swelling of size 2 cm × 1 cm on the left side of the mandible , extending from the mesial side of mandibular first premolar to the retromolar pad . The mucosa over swelling appears stretched, shiny and pale with patches of erythematous areas . Also , obliteration of lingual vestibule was noted (Fig.1). On palpation, swelling was slightly tender, firm in consistency, sessile which is nonreducible, noncompressible and nonfluctuant. The involved tooth did not show signs of mobility or tenderness. A provisional diagnosis was epulis , pyogenic granuloma .

Cone beam computed tomography (CBCT) radiograph which showed bone loss in 34 , 35 and 36 regions displaced and incomplete root. (Fig.2) Incisional biopsy was advised for diagnosis . Histopathology showed , H&E stained section of the single soft tissue exhibited a fibrocellular stroma with cellular proliferation of spindle shaped cells which are arranged in intersecting fascicles ,whorled and storiform pattern with few hyalinised areas . Moderate degree of chronic inflammatory cell infiltrate with few foamy histocytes . Hence , final diagnosis of “Benign Fibrous Histiocytoma” was given (Fig.3). Considering the unusual site of the tumor in this case, immunohistochemistry was

performed to confirm the diagnosis. It revealed positivity for CD 68, SMA & negativity for S 100 (Fig. 4). This indicated fibroblastic & histiocytic cell origin . Less aggressive behavior excluded the neural lesion , thus confirmed final diagnosis as Benign Fibrous Histiocytoma . After surgical treatment and follow up revealed no signs of local recurrence.

DISCUSSION

Benign Fibrous Histiocytoma (BFH) represents a diverse group of neoplasms which exhibit both fibroblastic and histiocytic differentiation. The term "Benign Fibrous Histiocytoma" was first described by Stout and Lates in 1967 .[2] It is a mesenchymal tumor and most commonly occurs on the skin of the extremities as a solitary, slowly growing nodule in early to mid-adult life.[4] Primary BFH of the bone is rare, 1% of all benign bone tumors.[5] As per review of literature of BFH involving oral and maxillofacial region, occurrence of this tumor in the jaw bone is rare as only 2 cases of maxilla and 7 cases of the mandible has been reported.[2]

The age of the patients in the cases reviewed ranged from 8 to 62 years, with the average of 33.8 years. Among the eighteen case reports summarized in Table 1, the incidence was slightly more in males (n = 10) compared to females (n = 8), with a male: female ratio of 1.25:1. In our case, the patient was a 9-year-old female. According to the reports, the most common location for BFH was the mandibular posterior region followed by the maxillary posterior region and the mandibular anterior region.[13] In the present case, the affected site is mandibular posterior region. In six cases, it had affected the left side, whereas right side was affected in three cases . Thus, it appears that BFH has affinity for the left side, which was also evident in the present case. Swelling alone was the most common symptom in the reviewed cases, followed by pain , while in two cases, the patients were asymptomatic. While painless swelling was evident in present case . Radiographic appearance in the

cases reviewed varied from well-defined,ill-defined unilocular radiolucency to well-defined multilocular radiolucency with or without reactive hyperostotic border. In the present case, it appeared as a ill -defined multilocular radiolucency without hyperostotic border .

Immunohistochemical staining was done in six cases which showed that the tumor cells were positive for vimentin , CD68 , α-1-antitrypsin and α-1-antichymotrypsin and negative for SMA , S-100 protein, cytokeratin , desmin , and CD34. The positivity for CD68 and vimentin indicated that the lesion was composed of histiocytic cells and fibroblast-like cells , and negative for SMA and S-100 showed that the lesion could be differentiated from leiomyosarcoma and neurogenic tumors.[13] In the present case immunohistochemical staining demonstrated strong positive for CD68 in moderate number of cells . Positive for SMA indicated that the lesion was composed of apha actin smooth muscle cells due to location of SFH and the neurogenic tumors could be differentiated due to S-100 negativity.[4]

According to the WHO histological classification of the tumors, primary BFH of the bone is defined as a benign lesion composed of spindle-shaped fibroblasts arranged in a storiform pattern with a variable admixture of small, multinucleated osteoclast-like giant cells. Foamy cells (xanthoma), chronic inflammatory cells, stromal hemorrhages, and hemosiderin pigment are also commonly present.[13] In our case, it showed spindle-shaped fibroblasts arranged in a storiform pattern, histiocytes, and giant cells .

According to Cale et al., BFH and the metaphyseal fibrous defect together constitute the benign fibrohistiocytic lesions of the bone. On the contrary, though BFH lesions may occur at any age, they are more common in adults and are frequently associated with pain even in the absence of fracture. BFH occurs in non-long bones, or even if in a long bone there is lack of metaphyseal involvement.[5] In our case, it was a painless lesion occurring in non-long bone (mandible) without bone fracture.

Differential diagnosis of oral BFH includes : MFH , Fibrosarcoma , Solitary fibrous tumor , Angiomatoid fibrous histiocytoma , Leiomyoma.[1]

MFH was excluded due to lack of pleomorphic sarcomatous cells, lack of typical and atypical mitotic figures, and lack of bizarre giant cells, necrosis, and prominent areas of haemorrhage. Fibrosarcoma was excluded by the lack of malignant features, invasive margins, and the characteristic herring-bone pattern. SFT was excluded as they pathologically appear as monomorphic spindle cells organised into interlacing fascicles and 'pattern-less' pattern with hemangiopericytoma-like vascular spaces. AFH was excluded as it is characterized predominantly by sheets of histiocytic cells arranged in serpentine pattern having cystic areas of haemorrhage. Leiomyoma was excluded it has distinct fascicular arrangement with blunt-ended plumper nucleus and cytoplasm showing longitudinal striations corresponding to myofilamentous material.[12]

The prognosis for BFH seems to be excellent and the recurrence rate of BFH is low.[16] The follow-up period in the reviewed case ranged from 1 to 3 years and no recurrence had been found. In our case, follow up for 1 years with no recurrence found . Normally, the treatment plan for BFH consists of a wide surgical resection as mentioned in the literature, but considering the age of the patient and location of the lesion . It was decided to curette out the lesion instead of resection of the affected area for the best outcomes.

CONCLUSION :

Benign fibrous histiocytoma is a mesenchymal tumor composed of cells with fibroblastic and histiocytic characteristics. The tumor is rare and presents a clinical and histopathological challenge. Proper diagnosis and treatment plan with longterm follow-up is of utmost importance in the management of these tumors.[7]

Table 1: Summary of case reports of benign fibrous histiocytoma in either jaw in chronological order

First author	Year	Age/ sex	Site	Duration	Side	Symptoms	Radiographic appearance	Immunohistochemical findings
Cale	1989	13/M	Post maxilla	Unknown	L	Asymptomatic	Ill-defined unilocular radiolucency	Not done
Dardo et al. [14]	1998	44 year/M	Floor of mouth					
Dardo et al. [14]	1999	34 year/M	Tongue					
Ertas[8]	2003	13/F	Ant mandible	NR	L	Swelling	Well-defined unilocular radiolucency	Not done
Heo[1	2004	42/M	Post mandible	8 years	L	Swelling	Well-defined multilocular radiolucency with hyperostotic border	Positivity for CD68, vimentin Negativity for SMA, S-100
Tetsuo Shimoyama	2004	32/F	Palate	2 years	R	Hard nonulcerated mobile polypoid mass	Not done	Immunopositive for vimentin. negative for S-100 protein , smooth muscle actin , desmin , CD68 .
Kishino[5]	2005	49/F	Post mandible	NR	L	Swelling and pain	Well-demarcated soap-bubble	Positivity for vimentin, CD68, α-1-

							appearance without sclerotic rim	antitrypsin, α-1-antichymotrypsin Negativity for SMA, S-100 protein, desmin, cytokeratin, CD34
Skoulakis et al. [4]	2007	19 /M	Cheek					
Katagiri[3]	2008	48/M	Mandible-condyle	Unknown	R	Asymptomatic	Ill-defined unilocular radiolucency Without sclerotic rim	Positivity for vimentin, CD68 Negativity for cytokeratin, SMA, S-100 protein, CD34
Bage and Bylappa [15]	2010	51 /F	Buccal mucosa					
Paraskevi	2010	36 /M	Buccal mucosa					
Wagner[6]	2011	41/M	Post mandible	NR	R	Swelling	Well-demarcated multilocular radiolucent lesion with a reactive hyperostotic border	Positivity for vimentin, CD68 Negativity for cytokeratin, SMA, S-100 protein, desmin
Gupta[2]	2011	24/F	Post mandible	NR	L	Swelling	Well-defined unilocular radiolucency	Not done
Pia et al.	2011	8 /F	Tongue					

[17]								
Saluja, et al.	2012	23/F	Post maxilla	8 months	L	Pain and swelling	Well-demarcated multilocular radiolucent lesion without a reactive hyperostotic border	Positivity for CD68, α-1-antichymotrypsin Negativity for SMA
Himanshu et al. [19]	2012	62/F	Buccal mucosa					
Narendra et al. [20]	2013	26 /M	Tongue					
Pradipta et al. [21]	2013	45 /M	Submandibular space					

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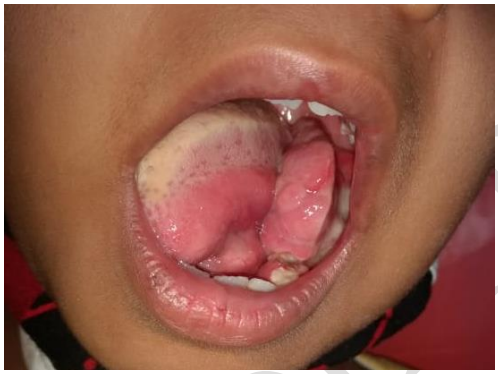

Fig.1: Intraorally , well-defined solitary swelling in the left posterior mandible .

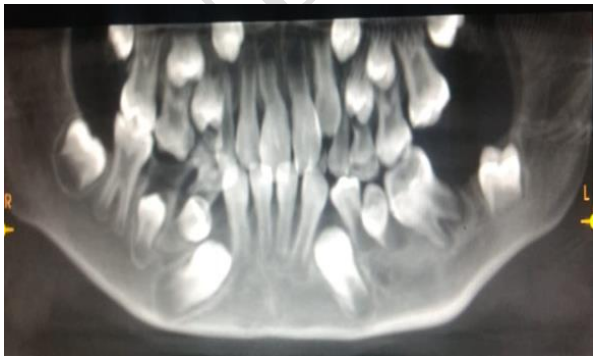

Fig.2: Radiographic examination in cone beam computed tomography (CBCT) radiograph -showed bone loss in 34,35,36 region . Displaced and incomplete root with 34,35,36 . Erupting and incomplete and erupting root formation with 33

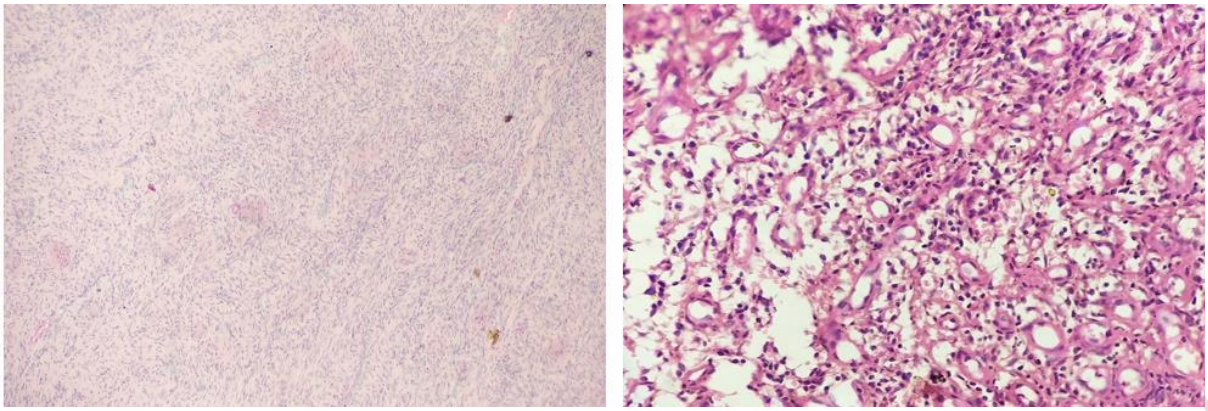

Fig.3 : Histological images showing fascicular & storiform arrangement of spindle cells, scattered histiocytes & touton type giant cells respectively .

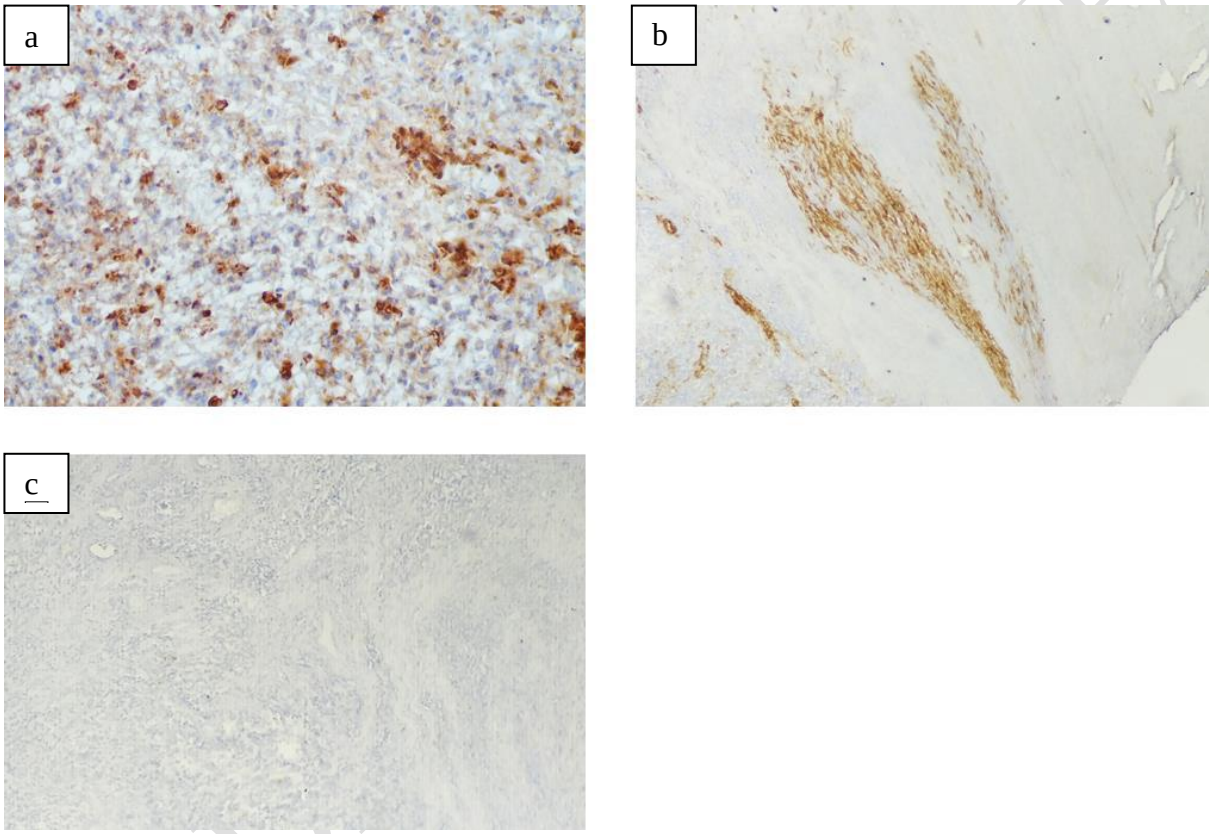

Fig.4 : Immunohistochemical images showing positivity a) , b) CD 68, SMA & c)negativity for S 100