Non Malignant Maxillary Lesions: Our Experience

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Abstract A wide variety of lesions occur in maxilla. Non specificity of clinical and radiological features makes diagnosis of these lesions a difficult task. We report six interesting cases of maxillary swelling among a total number of 37 such lesions of maxilla. These six cases are as follows two cases of central giant cell granuloma, two cases of fibrous dysplasia, one case of pigmented melanotic neuroectodermal tumor and one case of solitary myofibroma.

Keywords Benign · Maxilla · Myofibroma

Introduction

A variety of benign lesions involve maxilla. The presenting features, symptomatology and advanced imaging technique help to reach a presumptive diagnosis but histopathological examination remains the mainstay of definitive diagnosis. Careful histological workup is essential for a correct diagnosis and timely intervention of many of these lesions. Among 37 cases of non malignant lesions of maxilla received at our institute during last 2 years, we report two cases of giant cell reparative granuloma, two cases of

fibrous dysplasia, one case of pigmented melanotic neuroectodermal tumor and one case of solitary myofibroma, for their rarity and overlapping features of these lesions clinically and radiologically.

Materials and Methods

Of 37 cases (Table 1) of benign lesions of maxilla received in 2 years between 1st February 2010 and 31st January 2012 in our institute, we report six uncommon and interesting lesions of maxilla.

Case-1: (Fig. 1a) A 28 year woman presented with a mass in the right side of the face since the last 2 years. The swelling was diffuse, non-tender and extending into right nasal wall. CT scan of paranasal sinuses showed large well defined soft tissue density lesion with central necrosis in the right maxilla with erosion of anterior and medial walls of maxillary sinus and alveolar process. No calcification or tooth noted in the lesion. Ground glass opacities involving right zygomatic bone, right mandible, occipital, frontal and right parietal bones were also noted. Right maxillectomy along with lesion in the right zygomatic bone and frontal bone was done.

Case-2: (Fig. 1b, c) A 26 year lady presented with recurrent swelling in right hemi-face since last 8 months causing proptosis. CT scan showed a hyperdense heterogeneous expansive mass of uneven contour involving ethmoid, right maxillary bone and right orbit. Biopsy was taken from the maxillary bone.

Case-3: (Fig. 2a) A 40 year woman came with recurrent painful swelling of left face since 4 months. CT scan showed a soft tissue density lesion involving left maxillary antrum with erosion of lateral wall of the maxillary sinus and the floor of left orbit. Tumor showed extension into the

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soft tissue of the cheek, intra-orbital and extraconal fat. Biopsy was sent from maxillary sinus.

Case-4: (Fig. 2b) A 26 year man presented with a recurrent lesion in left face since 1 year. CT scan revealed an irregular soft tissue shadow involving left maxillary antrum with marked expansion of maxilla anteriorly. Inferiorly, expansion of alveolar margins involving hard palate were seen, with thinned out cortex and bony trabeculations traversing the mass.

Table 1

Sl. no	Diagnosis	No. of cases
1	Dentigerous cyst	12
2	Radicular cyst	08
3	Odontogenic kerato cyst	02
4	Odontogenic myxofibroma	01
5	Cementifying fibroma	03
6	Giant cell reparative granuloma	02
7	Osteochondroma	02
8	Fibrous dysplasia	02
9	Ossifying fibroma	03
10	Melanotic pigmented neuroectodermal tumor	01
11	Solitary myofibroblastoma	01

Case-5: (Fig. 3a, b) A boy of 12 years presented with infraorbital swelling in the left maxillary region. The swelling was noticed as 1 cm mass at the age of 8 months and was slow growing. Since last 1 year, the swelling was rapidly growing, reached present size of 8 × 7 cm. Boy complaints of difficulty in eating and phonation. CT scan of paranasal sinuses showed expansile lesion of left maxillary bone with altered attenuated and thickened left nasal plate. Un-erupted teeth were present within the lesion. Hemi-maxillectomy was done and entire lesion was removed and reconstruction surgery was done.

Case-6: (Fig. 4a) A 13 yr girl presented with swelling involving left maxillary region since 8 months gradually increasing in size. CT scan revealed an expansile lytic lesion involving alveolar process of maxilla with partial erosion of upper medial canine tooth. The lesion was expanding into antrum with remodeling of floor and medial wall of left maxillary sinus. Internal areas of calcification noted. No evidence of internal septations.

Results

Case-1 and -2: (Fig. 1d) Biopsy from maxilla showed disorganized bone surrounded by immature fibrous tissue.

Fig. 1

Fig. 2

The bony trabeculae lack osteoblastic rimming. These irregular, misshapen trabeculae formed odd geometric patterns. Case-1 is diagnosed as polyostotic fibrous dysplasia in co-ordination with clinical data, and case-2 was diagnosed as monostotic fibrous dysplasia.

Case-3 and -4: (Fig. 2c) Biopsy from maxilla showed fibrovascular connective tissue stroma composed of proliferating plump fibroblasts along with osteoclastic giant

Fig. 3

cells, hemosiderin laden macrophages, hemorrhage and foci of osteoid. Both case-3 and -4 were diagnosed as giant cell reparative granuloma.

Case-5: (Fig. 3c) Sections from the soft tissue and decalcified sections showed large cells containing deep brown pigment and small blue round cells forming rosettes and small clusters. Melanin bleach was done to

demonstrate that the brown pigment in larger cells is melanin. HMG45 was positive in melanin containing cells. Synaptophysin and GFAP was focally positive in small cells, neuron specific enolase was positive in both pigmented and small cells. These features are consistent with melanotic neuroectodermal tumor of infancy.

Case-6: (Fig. 4b) Pathologic analysis of the excised specimen revealed primitive fibroblastic cell proliferation with infiltration into the skeletal muscle and adipose tissue. There was no evidence of cellular atypia and no identifiable mitoses. Immunohistochemical staining was positive for vimentin, smooth muscle actin and HHF-35, thus confirming myofibroblasts. The lesion was diagnosed as solitary myofibroma.

Discussion

Fibrous dysplasia

It is a painless proliferation of osteo-progenitor tissue [1]. It occurs in teenagers and young adults of both sexes. When the jaws are affected, the monostotic type (Lichtenstein-Jaffe syndrome) is more common than the

polyostotic variety (McCune-Albright syndrome). Fibrous dysplasia is located in the maxilla more often than in the mandible. It is usually unilateral with an intact lamina dura. Mandibular lesions are more likely to be mottled and multicystic. Mature maxillary lesions have a homogeneous, ground-glass, and peau d'orange appearance [2] and blend with adjacent trabecular bone without margination or borders. The maxillary sinus may be obliterated. The primary differential diagnosis for fibrous dysplasia of the jaw is ossifying fibroma [3]. Our both cases did not show histological appearances of ossifying fibroma.

Giant cell reparative granuloma (GCRG)

It was introduced by Jaffe in 1953. It is not a true neoplasm but rather a reactive process; it could be triggered by trauma or inflammation [4, 5]. GCRG often arises in the mandible or maxilla and affects children and young adults, predominantly females, in the 2nd and 3rd decades of life [6]. GCRGs are classified, according to location, as central or peripheral, occurring respectively, in bone or gingival soft tissues [7]. Fast-growing lesions have rarely been reported. In these cases, GCRGs are characterized by an aggressive behaviour against an innocent histological appearance, pain and rapid facial swelling and high recurrence rate (as early as 3, and as late as, 22 years) [8].

Several morphological parameters, such as mitotic index, stromal characteristics, and values reflecting the average dimension of giant cells and the percent of tumour mass they occupied, have been tested in an effort to reliably divide giant cell lesions of the jaw into indolent and aggressive cases [7]. However, none of these have shown a significant association with clinical behaviour. Further studies, on large series of giant cell lesions of the jaw, could possibly produce some useful data on this point.

Giant cell reparative granuloma might be treated by non-surgical methods such as radiotherapy, daily systemic doses of calcitonin, and intra-lesional injections of corticosteroids. A non-surgical approach is probably a good treatment option for small, slow-growing lesions.

As far as concerns surgery, Becelli et al. [8] described a case of excision of the lesion and reconstruction via an autologous iliac crest bone graft, osseo-integrated implants, and “overdenture” prosthesis. Other authors have treated aggressive varieties by combined curettage and cryosurgery [8], or by curettage and peripheral ostectomy [7]. Bataineh et al. [8], instead, advocated resection without continuity and peripheral ostectomy and, finally, Choung et al. [6] suggested that aggressive cases should be treated with en bloc resection. However, the traditional treatment of GCRG is represented by surgical removal via an intra-oral approach and the extent of tissue removal ranges from a simple curettage to an en bloc resection. Curettage alone, or in

Fig. 4

combination with a periosteal bone resection is the treatment modality most often used [8]. Curettage has also been supplemented with cryosurgery and peripheral osteotomy. The surgical defect usually heals by secondary intention.

We have successfully treated both cases presented in this group by excision of the lesion and reconstruction of the jaw with an autologous iliac crest bone graft, allograft bone and membranes fixed on the lateral and medial walls of the mandible.

Melanotic neuroectodermal tumor of infancy

It is a rare oral melanotic neuroectodermal tumor of infancy (pigmented epulis of infancy) is a congenital neoplasm of uncertain histogenesis which was first described by Krompecher in 1918 [9]. Thus far, ~200 cases have been reported. Referred to additionally as retinal anlage tumor, melanotic progonoma, pigmented ameloblastoma, melanotic adamantinoma, and congenital melanocarcinoma, it is considered to arise from neural crest cells, the cells which are embryologically responsible for much of the development of the maxillofacial region [10]. Extraoral examples have been reported from the mediastinum, brain, anterior fontanelle, epididymis, and soft tissues of the arm (Table 2) [11]. Malignant variants

Table 2 Location of melanotic neuroectodermal tumors, as reported in the literature, 1990–1997. Modified from Yukiko et al. 2000 [11]

Location	Number of lesions reported
Maxilla	19
Skull	8
Epididymis	7
Brain	3
Mandible	2
Retina	1
Meninx	1
Transverse sinus	1
Total	42

Table 3 [16]

Authors	Year	Cases	SMA	Vimentin	S-100p	Desmin	HHF-35
Jones et al. [12]	1994	12	+	+	–	–	ND
Lingen et al. [4]	1995	04	+	+	–	–	ND
Sugatani et al. [9]	1999	01	+	+	–	–	ND
Montgomery et al. [13]	2000	09	+	ND	–	–	ND
Jordan and Regezi [5]	2003	08	+	ND	–	–	ND
Scheper et al. [7]	2005	01	+	ND	–	ND	ND
Azevedo et al. [11]	2008	02	+	+	–	–	+
Present study	2012	01	+	+	–	–	+

represent less than 5% of reported cases [12]. Our case-5 has all the features of benign melanotic neuroectodermal tumor of infancy.

Solitary myofibroma

Solitary myofibroma of adults is an uncommon neoplasm that typically arises in soft tissue and subcutaneous sites in the head and neck, but rarely within bone. According Jordan and Regezi [13], the prevalence of solitary myofibroma in the oral cavity is very low. Surgical excision is the treatment of choice for myofibroma. Electron microscopic examination shows myofibroblast-like cells, fibroblastic-like cells and intermediate cells. A majority of the tumor cells are composed by intermediate cells type and myofibroblastic-like cells have cytoplasmatic filaments with dense bodies, basal lamina, pinocytotic vesicles and nuclear invaginations [14]. However, electron microscopic evaluation of tumor specimens is not routinely used in most pathology services.

In the current case, definitive diagnosis was made on the basis of the histopathological and immunohistochemical findings (Table 3) [16]. Microscopically, a typical biphasic pattern can be observed. Peripheral zone of myofibroma is composed of spindle-shaped cells with long oval nuclei and abundant eosinophilic cytoplasm that resembled smooth muscle. Central portion of this tumor comprise a population of more primitive spindle cells associated with a hemangiopericytoma-like vascular pattern [15]. The prognosis of this rare tumor is excellent after conservative surgical curettage or resection [16]. According Daimaru et al. [16], the recurrence rate is only 7% after excision. In agreement with previous reports, there was no evidence of recurrence after over the 2 years of follow-up.

Conclusion

We present six rare lesions of maxilla and reviewed the available literature regarding these lesions. These are being presented for their rarity and they mimic malignant lesions.

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