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ORIGINAL ARTICLE

Odontogenic Tumours of Jaw: A Prospective Study on Clinico-Pathological Profile and Their Management

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Abstract Odontogenic tumours are a group of heterogeneous diseases that range from hamartomatous or non-neoplastic tissue proliferations to benign neoplasms to malignant tumours with metastatic potential. They are rare, comprising about <2–3% of all oral and maxillofacial biopsy specimens. The aim of the present study was to determine the clinico-pathological presentation of this heterogeneous group of lesions and review of literature. The present study was conducted in the ENT department of a Government Medical College and Hospital, West Bengal, India, over the period of 5 years from January 2011 to December 2015. It included a total of 15 patients who were clinico-radiologically diagnosed as odontogenic tumours, and were given appropriate treatment. Their diagnostic and management approaches are discussed. Among 15 odontogenic tumours, 13 were benign and two were malignant. Male to female ratio was 2:3. Mandible to maxilla ratio was 1.8:1. The patients were in between 4 and 56 years of age with highest incidence in 3rd decade of life. All patients are doing well till date with a minimum follow-up of 1 year. Incisional biopsy is considered as gold standard for preoperative diagnosis but FNAC can offer clinicians a less invasive alternative. CT is the choice of investigation for study of lesion, analysis of its extension and surgical planning. The challenge to proper management lies in balancing between conservative and radical approach to reduce morbidity and recurrence both. Final diagnosis is made by post-operative histopathological examination.

Keywords Odontogenic tumours · Jaw · CT scan · Biopsy · Follow-up

Introduction

Odontogenic tumours are a group of heterogeneous diseases that range from hamartomatous or non-neoplastic proliferations to benign neoplasms to malignant tumours with metastatic potential [1, 2]. They are derived from epithelial, ectomesenchymal and/or mesenchymal elements of tooth-forming apparatus. Odontogenic tumours are rare, comprising about <2–3% of all oral and maxillofacial biopsy specimens sent to pathology department [3]. They pose a significant diagnostic and therapeutic challenge. Knowledge of basic clinical features such as age, sex, and location can be extremely valuable in developing differential diagnoses of odontogenic tumours. Available literature of on odontogenic tumors are mostly among Africans and Americans. Very few studies are reported among Asians, especially from the Indian subcontinent [4]. The aim of the present study was to evaluate the clinico-pathological presentation of this heterogeneous group of lesions and review of literature.

Materials and Methods

The present prospective study was conducted in the ENT department of a Government Medical College and Hospital, West Bengal, India, over the period of 5 years from January 2011 to December 2015. It included a total of 15

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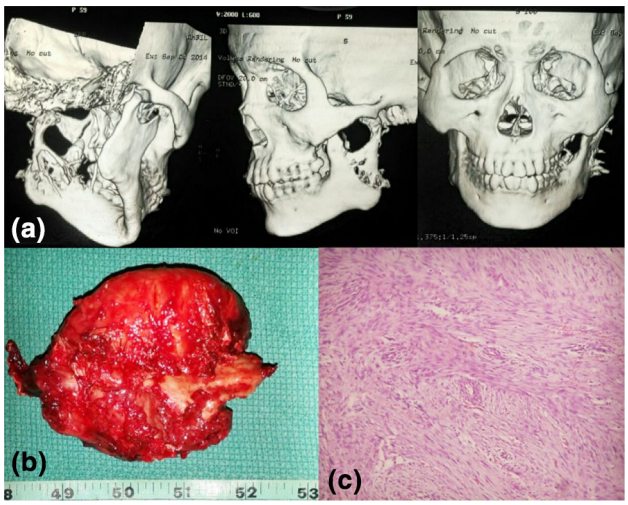

Fig. 1 **a** 3D CT reconstruction of Case no. 4. **b** Specimen of segmental mandibulectomy. **c** Post-operative histopathological picture suggesting odontogenic fibromyxoma

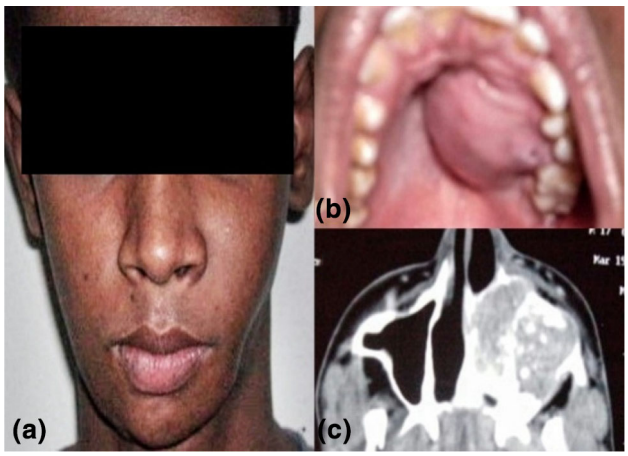

Fig. 2 **a** Profile picture of Case no. 5 with swelling of *left* cheek. **b** Intraoral picture of Case no. 5 showing *left* sided palatal bulge. **c** CECT of nose and PNS axial section showing expansible mass with calcified matrix arising from *left* maxillary sinus with extension into *left* nasal cavity

patients who were clinico-radiologically diagnosed as odontogenic tumours, and were given appropriate treatment. Odontogenic cysts and non-odontogenic tumours like squamous cell carcinomas were excluded from the study. Tumours were grouped under three main headings as per WHO guidelines (2005). A detailed history including age, sex and location of the tumour was taken followed by a thorough clinical examination of the patient. Radiological examination included orthopantomogram (OPG), CT scan or MRI whichever was appropriate for the specific case. Pre-operative cytological or histopathological examination was performed in suitable cases. The treatment modalities were based on clinical, radiological and wherever possible cytological or histopathological diagnosis. All patients with benign tumours were managed by appropriate surgery. Patients with malignant tumours were managed by surgery along with radiotherapy and/or chemotherapy. All patients were followed up regularly for a minimum of 1 year (Figs. 1, 2, 3, 4).

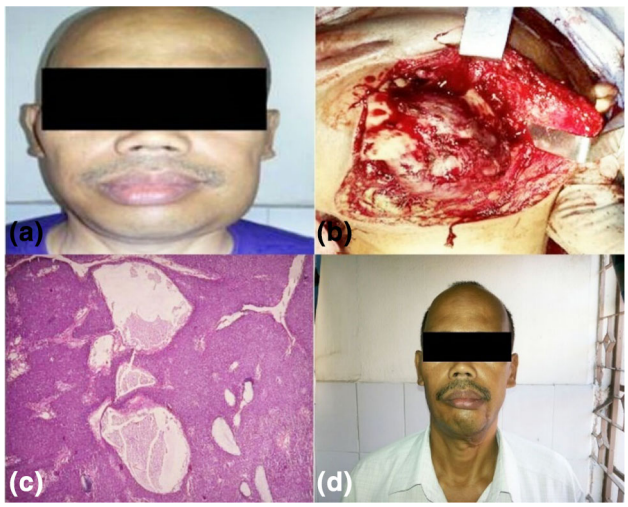

Fig. 3 **a** Profile picture of case no. 11 with swelling over *left* angle of mandible. **b** Intra-operative picture of *left* segmental mandibulectomy. **c** Post-operative histopathological picture suggesting ameloblastic carcinoma. **d** Follow-up at 1 month post-op

Results

A total of 15 cases with odontogenic tumours were diagnosed and treated during the 5 years period from January 2011 to December 2015. Of the 15 odontogenic tumours, 13 were benign and two were malignant. Among these, six were in males and nine were females (M:F = 2:3). Nine tumors were encountered in mandible, five in maxilla and one in ethmoid, with an overall mandible to maxilla ratio of 1.8:1. The patients were in between 4 and 56 years of age with highest incidence in 3rd decade of life (Table 1). Clinical, radiological and pre-operative cytological or histological findings are described in Table 2. Treatment and final histopathological diagnosis are discussed in Table 3. All patients are doing well till date with a minimum follow-up of 1 year.

Discussion

Odontogenic tumours are derived from tooth-forming elements. Odontogenesis occurs through a complex process involving enamel organ, dental follicle, and dental papilla. The enamel organ is an epithelial structure, derived from oral ectoderm. The dental follicle and dental papilla are

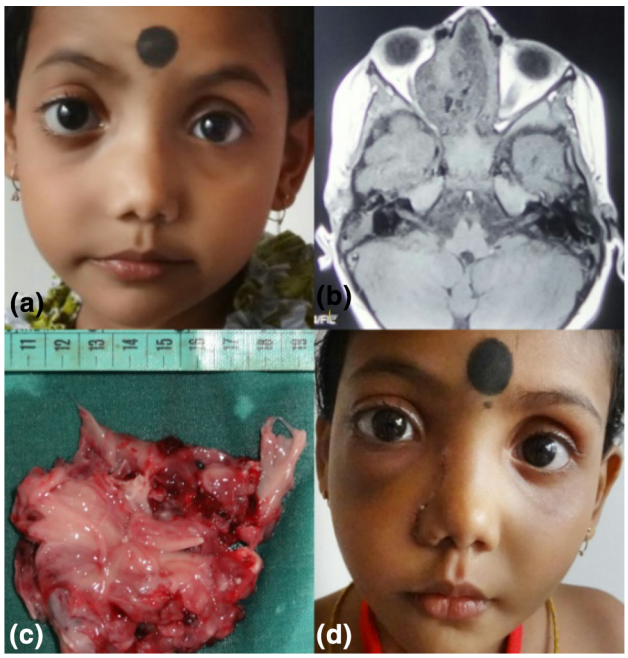

Fig. 4 **a** Profile picture of Case no. 14 with *right* sided proptosis. **b** CECT of nose and PNS axial section showing sinonasal mass pushing *right* eyeball. **c** Specimen of *right* medial maxillectomy with removal of the tumour. **d** Follow-up at 1 month post-op

Table 1 Distribution of patients according to age, sex and site of tumour

Case number	Age (years)	Sex	Site
1	23	Female	Right angle of mandible
2	27	Female	Right ramus of mandible
3	28	Female	Right maxilla
4	17	Male	Left angle and ramus of mandible
5	18	Male	Left maxilla
6	46	Female	Left angle of mandible
7	32	Female	Left angle of mandible
8	56	Female	Right side of body of mandible
9	26	Male	Left maxilla
10	37	Male	Right posterior ethmoid
11	55	Male	Left angle of mandible
12	34	Male	Body of mandible
13	16	Female	Left maxilla
14	4	Female	Right maxilla
15	21	Female	Body of mandible

derived from neural crest cells and are therefore ectomesenchymal in nature. Odontogenic tumours demonstrate varying interactions between the odontogenic epithelium and odontogenic ectomesenchyme, and are sub-classified by their tissue of origin [3] These tumours are grouped under three main headings as per WHO guidelines (2005): malignant (odontogenic carcinoma and odontogenic sarcoma), benign (Odontogenic epithelium without odontogenic ectomesenchyme; Odontogenic epithelium with odontogenic ectomesenchyme; Mesenchyme and/or odontogenic ectomesenchyme with or without odontogenic epithelium; Bone-related lesions), and other tumours [5].

Ameloblastoma arising from odontogenic epithelium without odontogenic ectomesenchyme, is the most common tumour of odontogenic origin [6]. Odontogenic fibromyxoma and cementoblastoma, arising from odontogenic ectomesenchyme with or without odontogenic epithelium, are uncommon tumours comprising 3–6% [7] and 1–6.2% [8] of all odontogenic tumours respectively. Cemento-ossifying fibroma, fibrous dysplasia, brown tumour of hyperparathyroidism and giant cell tumour are within the subgroup of bone-related lesions [5]. PNET is a tumour of neuroectodermal origin [5]. Ameloblastic carcinoma is grouped under malignant odontogenic tumour [5].

Odontogenic tumours occur over a wide range of ages with almost equal frequency in men and women [4]. Ameloblastoma occurs within mean age of 20 s and 30 s [9]. Odontogenic fibromyxoma occur in 2nd–4th decades with slight female predilection [7]. The peak incidence of cementoblastoma is between 2nd and 3rd decades of life with caucasian race and male predominance (M:F = 2.1:1) [10]. Greatest number of cases cemento-ossifying fibroma are encountered during 3rd and 4th decades of life with definite female predilection (M:F = 1:5) [11]. Fibrous dysplasia manifests more frequently in childhood [12]. The mean age of presentation of giant cell tumour is 8.2 years, more frequently affecting girls [13]. Brown tumour of hyperparathyroidism is more frequent in women aged over 50 years [14]. Most of the reported cases of PNET of jaw region are less than 20 years of age [15]. The mean age of occurrence of ameloblastic carcinoma is 30 years [16].

Most of the odontogenic tumours occur in the posterior aspect of mandible [17]. In the present series, 40% tumours (6 out of 15) were located at angle of mandible. Among five cases of ameloblastoma, two were at angle of mandible, two were at body of mandible and one at the ramus. Odontogenic fibromyxoma occurred in maxilla and ramus of mandible in one case each. One case each of cemento-ossifying fibroma, giant cell tumour and PNET were found in maxilla. Fibrous dysplasia occurred in maxilla and posterior ethmoid in one case each.

Many a times, odontogenic tumours are asymptomatic and are incidentally diagnosed by imaging studies [17]. The commonest presenting symptoms are swelling over jaw, dental malocclusion and mal-alignment. Larger and malignant tumours may present with pain, paresthesia or loosening of tooth. On clinical examination, the swelling is usually firm to hard and at times cystic due to perforation

Table 2 Distribution of patients according to clinical and radiological features

Case number	Clinical features	Radiological features	Pre-operative FNAC/HPE
1	~5 × 3 cm^2swelling over right angle of mandible for 4 years	Well defined multi-loculated lytic lesion over right angle of mandible with soap bubble appearance	Not done
2	~7.5 × 5 cm^2 swelling over right ramus of mandible for 9 years	Well defined multi-loculated lytic lesion over right ramus of mandible with soap bubble appearance	HPE—ameloblastoma
3	Swelling over right cheek and right half of palate for 4 years, right nasal obstruction with intermittent epistaxis for 2 years	~8 × 6 cm^2 expansile bony mass with ground glass appearance involving right maxillary sinus andnasal cavity	Not done
4	~8 × 6.5 cm^2 swelling over left angle and ramus of mandible for 2 years, pain for 3 months	Well capsulated mass over left ramus of mandible with bony septations	FNAC—inconclusive
5	~5 × 4 cm^2 swelling over left cheek for 3 years	Expansile mass with calcified matrix involving left maxillary sinus and nasal cavity	Not done
6	~4 × 3 cm^2 swelling over left angle of mandible for $3^1/2$ years	Well defined multi-loculated lytic lesion over left angle of mandible with soap bubble appearance	Not done
7	~6 × 4 cm^2 swelling over left angle of mandible for 5 years	Well-defined circular radio-opacity at left body and angle of mandible	Not done
8	~9 × 6 cm^2 swelling in right lower jaw for last $2^1/$ 2 years and pain for last 6 months	Expansile osteolytic lesion in the right side of body of mandible	Not done
9	~3 × 3 cm^2 swelling over left cheek for $1^1/$ 2 years	Well capsulated mass in left maxilla with bony septations	Not done
10	Headache for 2 years	Bony mass with ground glass appearance involving right posterior ethmoids	Not done
11	~5×4 cm^2 swelling over left angle of mandible for 7 years	Multi-loculated lytic lesion over left angle of mandible with ill-defined borders	FNAC—ameloblastoma
12	~3 × 2 cm^2 swelling over chin for $4^1/2$ years	Well defined multi-loculated lytic lesion over body of mandible with soap bubble appearance	Not done
13	~2.5 × 2 cm^2 firm swelling over left cheek for 2 years, left sided proptosis for 1 year	Heterogeneously enhancing growth involving left maxillary sinus and left nasal cavity with destruction of medial wall and floor of left maxillary sinus	HPE—malignant round cell tumour IHC—PNET
14	Right sided nasal obstruction and Epistaxis for 1 year, right cheek swelling and proptosis for 6 months	~7 × 5 cm^2osteolyticmultiloculated lesion in right maxilla	HPE—haemangio-pericytoma
15	~4 × 4 cm^2 swelling over chin for 6 years	Well defined multi-loculated lytic lesion over body of mandible with soap bubble appearance	Not done

of cortex. Signs of nasal obstruction, epistaxis, hyposmia, headache, change in visual acuity, proptosis, impairment of eye movements and trismus are to be searched. Cervical lymphadenopathies are to be looked for while suspecting malignant tumour [17].

The radiological investigations i.e. OPG, CT scan and MRI are adjunct to the armory. For a localized lesion, OPG is good enough to evaluate the extension. For extensive lesion, CT is the choice of investigation for study of the lesions, analysis of their extension and surgical preparation. Moreover, MRI and CT predict whether the tumour is resectable or not, detect distant metastasis and assess the tumour response to treatment. Some of the odontogenic tumours has a characteristic but not diagnostic radiological appearance. Ameloblastoma usually appears as unilocular or multilocular radiolucent area with a honeycomb appearance [9]. Ameloblastic carcinoma mostly presents as ill-defined or irregular margin and osteolytic radiolucency [16]. Odontogenic fibromyxoma may present as bone destroying lesion with ill-defined borders [18]. Cementoblastoma appears as well-defined circular radio-opacity with a radiolucent halo, fused to partly resorbed root(s) of the associated tooth [19]. Cemento-ossifying fibromas are well circumscribed, solitary radiolucencies with scattered radiopaque foci [20]. There are three radiographic patterns of fibrous dysplasia: pategoid, that alternates the radio-dense and radio-transparent areas and give characteristic "ground glass" appearance; sclerotic, homogeneously dense; cystic standard, with spherical or ovoid radiolucent area surrounded by dense limits [21]. GCT has classical osteolytic and multi-loculated appearance [13]. Brown tumour of hyperparathyroidism appears as mono-locular or multi-locular osteolytic lesion with root resorption and loss of lamina dura occasionally [22]. On CT, PNETs usually

Table 3 Distribution of patients according to treatment and post-operative HPE report

Case number	Treatment	Post-operative HPE
1	Right segmental mandibulectomy	Ameloblastoma
2	Right segmental mandibulectomy	Ameloblastoma
3	Right medial maxillectomy	Fibrous dysplasia
4	Left segmental mandibulectomy	Odontogenic fibromyxoma
5	Left partial maxillectomy	Cemento-ossifying fibroma
6	Left segmental mandibulectomy	Ameloblastoma
7	Left segmental mandibulectomy	Cementoblastoma
8	Right segmental mandibulectomy	Brown tumour of hyperparathyroidism
9	Left partial maxillectomy	Odontogenic fibromyxoma
10	Right ethmoidectomy	Fibrous dysplasia
11	Left segmental mandibulectomy followed by radiotherapy	Ameloblastic carcinoma
12	Segmental mandibulectomy	Ameloblastoma
13	Chemotherapy (ifosphamide, vincristine, etoposide, actinomycin D, doxorubicin) and radiotherapy followed by salvage left total maxillectomy, reconstruction with temporalis muscle rotational flap	PNET
14	Right partial maxillectomy	Giant cell tumour
15	Segmental mandibulectomy	Ameloblastoma

appear iso-dense or slightly hypo-dense compared to normal muscle and tumour calcifications are uncommon. On MRI, the majority of PNETs are iso-intense or slightly hyper-intense on T1WI and hyper-intense on T2WI. Furthermore, the tumour is often heterogeneously marked following intravenous administration of gadolinium [23].

FNAC can offer the clinician a conservative alternative to the more invasive procedures such as open biopsy. Thinning or destruction of cortical bone permits the use of thin needles for aspiration [24]. Free flowing fresh blood on aspiration alerts the pathologist and precludes biopsy in view of risk of severe hemorrhage. But, the role of FNAC for diagnosis of intra-osseous jaw pathology is limited. FNAC of intra-osseous jaw lesions is often problematic because of their proximity to tooth apices and neurovascular bundles [24]. Obtaining cellular aspirates may be quite difficult in heavily calcified and fibro-osseous lesions, such as cement-ossifying fibroma and fibrous dysplasia. FNAC can broadly diagnose odontogenic tumors, fibro-osseous lesions, giant cell lesions and cystic lesions; however, definitive categorization may not be always possible due to lack of specific cyto-morphological features [24]. Ultrasonography-guided percutaneous core needle biopsy is less invasive compared to open surgical biopsy [25]. It allows precise needle position and avoids vascular damage. Incisional biopsy has been considered as the main diagnostic modality for preoperative presumptive diagnosis of intra-osseous jaw tumours due to difficulty in accessing and lack of well-established cytological features [24].

Irrespective of the histological type, surgical resection is the treatment of choice. In case of benign tumours, there are two options: conservative surgery and radical surgery [26]. Conservative approach includes enucleation or curettage. In the radical surgery, the tumour is resected with 1–2 cm of macroscopically healthy bony margin. But the recurrence rate is much higher in cases of conservative surgery (up to 90%) than radical surgery (up to 25%). In case of malignant tumours, if the tumour is resectable a more radical approach is employed. Lymph node dissection should be considered when there is obvious lymphadenopathy. If margins are close or histopathologically positive then post-operative adjuvant radiotherapy should be given. In case of PNET, surgery along with adjuvant multi agent chemotherapy is used. In locally advanced tumours not amendable to surgical resection or with distant metastasis, radiotherapy and/or chemotherapy is used.

Overall prognosis is good in cases of benign odontogenic tumours. All are prone to recurrence even with proper management. So long term follow up is essential. Distant metastasis may occur in ameloblastic carcinoma or PNET, even in the absence of recurrence at primary site.

Conclusion

Odontogenic tumours occur over a wide range of ages with almost equal frequency in men and women. Most of them occur in posterior aspect of mandible. Incisional biopsy has

been considered as a prime diagnostic modality for pre-operative diagnosis but FNAC can offer clinicians a less invasive alternative. CT is the investigation of choice for study of lesion, analysis of its extension and surgical planning. Irrespective of the histological type, surgical resection is the treatment of choice. The challenge to proper management lies in balancing between conservative and radical approach to reduce morbidity and recurrence both. Final diagnosis is made by post-operative histopathological examination. Overall prognosis is good in cases of benign odontogenic tumours.

Compliance with Ethical Standards

Conflict of interest None.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was obtained from all individual participants included in the study.

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