Odontogenic Keratocyst: A Case Series of five Patients

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Received: 13 December 2011 / Accepted: 8 March 2012 / Published online: 21 March 2012
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Abstract During the time period of August 2009 to August 2010, five cases of odontogenic keratocyst were admitted and treated under the care of Department of Otorhinolaryngology, MGMC and RI, Puducherry. Patients came to the ENT OPD with history of swelling in the cheek region, nasal obstruction, numbness in the upper alveolar region. On examination diffuse swelling of size 7×3 cm in one patient and size of 5×3 cm in two patients, and other two patients size of 6×3 cm present in the maxillary region with ill defined borders, the swelling was firm in consistency, no warmth, non tender. Anterior rhinoscopy reveals mass pushing the lateral wall medially, septum pushed to opposite side, mucopus present in nasal cavity, airway reduced on the side of swelling. On examination of oral cavity, a small granulation of size 1.0×0.5 cm present in two patient and swelling of size 1.5×1.0 cm seen in two patients in vestibule, no swelling in one patient and swelling of size 3×2 cm seen in hard palate of two patients and no swelling in three patients, no loosening of tooth seen in all patients. X-ray PNS reveals maxillary haziness, diagnostic nasal endoscopy reveals lateral wall of nose pushed medially and septum pushed to opposite side. FNAC reveals resolving inflammatory aspirate in one patient, few macrophages seen in two of patients, few keratinocytes seen in two of the patients. CT nose and PNS revealed a large cystic lesion with erosion of anterior and medial wall and floor of maxilla in relation to the root of the last molar tooth in two patients and there is erosion of anterior and medial wall in other three patients. A combined endonasal and external sublabial (Caldwell-luc) approach was performed in four

patients and the cystic lesion was removed and in other one patient only endonasal approach was done and cystic lesion was removed and sent for biopsy. Biopsy sent for HPE revealed odontogenic keratocyst.

Keywords Odontogenic cysts · Keratocyst · Tumours of mandible and maxilla

Introduction

Odontogenic keratocyst was first described in 1876 and named by Philipsen in 1956. It is one of the most aggressive odontogenic cysts of the oral cavity [1]. It generally thought to be derived from either the epithelial remnants of the tooth germ or the basal cell layer of surface epithelium [2]. The incidence of male to female ratio is about 1.6:1 [3]. Odontogenic keratocysts may occur in any part of the upper and lower jaw with the majority occurring in the mandible, most commonly in the angle of the mandible and ramus [4]. The maxillary sinus is a frequent site for pathologies of odontogenic origin due to its close anatomical relationship with teeth and periodontal tissues, inflammatory disease as well as neoplastic and tumourous lesions can occur in this area. Maxillary sinus involvement must be carefully assessed because orbital damage and the spreading of associated infections could lead to local and systemic compromise to the patient, present with pain, swelling and paresthesia of lower lip or teeth. Displacement of tooth and destruction of the floor of the orbit, proptosis of eyeball when it involves maxilla are the most common features. Maxillary cysts cause buccal expansion but palatal expansion are rare. Investigation include X-ray PNS, diagnostic nasal endoscopy, computed tomography of PNS and FNAC. Treatment includes marsupialization,

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enucleation and curettage, enucleation and peripheral ostectomy, osseous resection, cryotherapy which include the use of liquid nitrogen.

Case History

Case 1

18 year old male presented to ENT OPD, MGMC and RI, Puducherry. Complaints of swelling in the left cheek, nasal obstruction and numbness in the upper alveolar region. O/E: diffuse swelling of size 7×3 cm in the left maxillary region with ill defined borders, swelling was firm in consistency extending inferiorly to hard palate, no warmth, non tender. Oral cavity—a small granulation of size 1.0×0.5 cm present in the vestibule and swelling of size 3×2 cm in hard palate; no loosening of tooth. Anterior rhinoscopy reveals pushing of lateral wall medially and pushing septum to opposite side and mucopus present in left nasal cavity, airway reduced on left side. X-ray PNS reveals left maxillary haziness and DNE-lateral wall of nose pushed medially, and septum pushed to opposite side. FNAC—reveals resolving inflammatory aspirate. CT nose and PNS reveals a large cystic lesion with erosion of anterior, medial wall and floor of left maxilla in relation to root of last molar tooth. A combined endonasal and Caldwell-luc approach was done and cystic lesion removed. Biopsy sent for HPE and it reveals odontogenic keratocyst.

Case 2

20 year old female presented to ENT OPD, MGMC and RI, Puducherry. Complaints of swelling in the right cheek and nasal obstruction. O/E: diffuse swelling of size 5×3 cm in the right maxillary region with ill defined borders, swelling was firm in consistency, no warmth, non tender. Oral cavity—normal, no loosening of tooth. Anterior rhinoscopy reveals pushing of lateral wall medially and mucopus present in right nasal cavity, airway reduced on right side. X-ray PNS reveals right maxillary haziness and DNE-lateral wall of nose pushed medially. FNAC—few keratinocytes seen. CT nose and PNS reveals a large cystic lesion with erosion of anterior and medial wall of right maxilla in relation to root of last molar tooth. A endonasal approach was done and cystic lesion removed. Biopsy sent for HPE and it reveals odontogenic keratocyst.

Case 3

16 year old female presented to ENT OPD, MGMC and RI, Puducherry. Complaints of swelling in the left cheek, nasal obstruction. O/E: diffuse swelling of size 6×3 cm in the

left maxillary region with ill defined borders, swelling was firm in consistency, no warmth, non tender. Oral cavity—a small granulation of size 1.0×0.5 cm present in the vestibule and no loosening of tooth. Anterior rhinoscopy reveals pushing of lateral wall medially and mucopus present in left nasal cavity, airway reduced on left side. X-ray PNS reveals left maxillary haziness and DNE-lateral wall of nose pushed medially. FNAC—few macrophages seen. CT nose and PNS reveals a large cystic lesion with erosion of anterior and medial wall of left maxilla in relation to root of last molar tooth. A combined endonasal and Caldwell-luc approach was done and cystic lesion removed. Biopsy sent for HPE and it reveals odontogenic keratocyst.

Case 4

20 year old male presented to ENT OPD, MGMC and RI, Puducherry. Complaints of swelling in the left cheek, nasal obstruction. O/E: diffuse swelling of size 5×3 cm in the left maxillary region with ill defined borders, swelling was firm in consistency, no warmth and non tender. Oral cavity—a small swelling of size 1.5×1.0 cm present in the vestibule, no loosening of tooth. Anterior rhinoscopy reveals pushing of lateral wall medially and mucopus present in left nasal cavity, airway reduced on left side. X-ray PNS reveals left maxillary haziness and DNE-lateral wall of nose pushed medially. FNAC—few keratinocytes seen. CT nose and PNS reveals a large cystic lesion with erosion of anterior and medial wall of left maxilla in relation to root of last molar tooth. A combined endonasal and Caldwell-luc approach was done and cystic lesion removed. Biopsy sent for HPE and it reveals odontogenic keratocyst.

Case 5

16 year old male presented to ENT OPD, MGMC and RI, Puducherry. Complaints of swelling in the right cheek, nasal obstruction and numbness in the upper alveolar region. O/E: diffuse swelling of size 6×3 cm in the right maxillary region with ill defined borders, swelling was firm in consistency extending inferiorly to hard palate, no warmth, non tender. Oral cavity—a small swelling of size 1.5×1.0 cm present in the vestibule and swelling of size 3×2 cm in hard palate; no loosening of tooth. Anterior rhinoscopy reveals pushing of lateral wall medially and pushing septum to opposite side and mucopus present in right nasal cavity, airway reduced on right side. X-ray PNS reveals right maxillary haziness and DNE-lateral wall of nose pushed medially and septum pushed to opposite side. FNAC—few macrophages seen. CT nose and PNS reveals a large cystic lesion with erosion of anterior and medial wall and floor of right maxilla in relation to root of last molar tooth. A combined endonasal and Caldwell-luc

approach was done and cystic lesion removed. Biopsy sent for HPE and it reveals odontogenic keratocyst.

Materials and Methods

During the time period of August 2009 to August 2010, five cases of odontogenic keratocyst were admitted and treated under the care of Department of Otorhinolaryngology, MGMC and RI, Puducherry. Patient underwent thorough history taking and complete ear, nose and throat examination.

Routine blood and urine investigation, nasal swab were done for all patients. Diagnostic nasal endoscopy, X-ray para nasal sinuses, CT nose and PNS done to confirm the diagnosis.

The diagnosis of odontogenic keratocyst was established by direct examination of swelling in the maxillary region and pushing of lateral wall medially by the swelling and deviation of septum to opposite side, mucopus in the nasal cavity by anterior rhinoscopy. CT nose and PNS revealed a large cystic lesion with maxillary erosion of anterior and medial wall and floor in two patients, anterior and medial wall in other three patients in relation to the root of the last molar tooth. HPE-basal layer is showing prominent palisading pattern. Some cells are showing clear cell changes.

Results and Analysis

Out of 5 patients; 3 are male and 2 are female, around the age group of 15–20 years. The patient presented with swelling in the maxillary region as their major complaint. Their presenting symptoms are nasal obstruction and numbness over the upper alveolar region.

In all 5 patients, on examination, a diffuse swelling of varying size 7×3 cm in one patient and 5×3 cm in two patients and other two patients size of 6×3 cm present in maxillary region with ill defined borders, the swelling was firm in consistency, no warmth, non tender and the swelling was extending inferiorly to the hard palate in two of our patients. Anterior rhinoscopy reveals pushing of lateral wall medially, septum pushed to opposite side, mucopus present in nasal cavity, airway reduced on the side of swelling.

On examination of oral cavity, a small granulation of size 1.0×0.5 cm present in two patient and swelling of size 1.5×1.0 cm seen in two patients in vestibule and no swelling in one patient in vestibule, swelling of size 3×2 cm seen in two patients and no swelling in other three patients in hard palate, no loosening of tooth seen in all patients (Figs. 1, 2).

X-ray PNS reveals maxillary haziness on all patients. Diagnostic nasal endoscopy reveals septum pushed to

opposite side in two patients and lateral wall of nose pushed medially in all patients. FNAC reveals resolving inflammatory aspirate in one patient and few macrophages in two patients and few keratinocytes seen in two patients. CT nose and PNS revealed a large cystic lesion with erosion of maxilla (Anterior and medial wall and floor in two patients, anterior and medial wall in other three patients.) in relation to the root of the last molar tooth for all patients (Fig. 3). HPE-basal layer is showing prominent palisading pattern. Some cells are showing clear cell changes (Fig. 4).

Routine blood investigations and urine investigations reveals normal for the patients. Pre-anesthetic assessment obtained for all patients for surgery.

Fig. 1 Granulation in vestibule and swelling in hard palate

Fig. 2 Swelling in hard palate

Fig. 3 CT nose and PNS

Fig. 4 HPE-basal layer shows prominent palisading pattern, some cells shows clear cell changes

A combined endonasal and external sublabial (Caldwell-luc) approach was performed for four patients and only endonasal approach was performed in one patient and the cystic lesion was removed (Fig. 5). Part of anterior and medial wall, floor of maxilla found to be eroded in two patient and anterior and medial wall erosion in other three patients. Biopsy sent for HPE revealed odontogenic keratocyst for all the patient. The average hospital stay of the patients was 7–10 days. The patients had no complication and recovered completely (Fig. 6).

Fig. 5 Post operative maxillary antrum

Fig. 6 Post operative hard palate

Discussion

Odontogenic keratocyst is known for its rapid growth and its tendency to invade the adjacent tissues including bone. It has a high recurrence rate and is associated with the basal cell nevus syndrome. Although the term only indicates the formation of keratin and is non specific, as keratin formation may be seen even in radicular cysts and dentigerous cysts as a result of metaplasia, it is so well established in literature that it continues to be in use. Subsequently a lot of work on the molecular biology and genetics have been carried out by many researchers and based on which it has been suggested to be considered as a benign tumour and hence be renamed keratocystic odontogenic tumour (KCOT).

The majority of patients are in the age ranges of 20–29 and 40–59, but cases ranging from 5 to 80 years have also been reported. The distribution between sexes varies from equality to a male to female ratio of 1.6:1, except in children. Odontogenic keratocysts may occur in any part of the upper and lower jaw with the majority occurring in the mandible, most commonly in the angle of the mandible and ramus. Odontogenic keratocysts of the maxilla are smaller in size compared to the mandible. When they are large, they tend to expand bone. It is important to note that OKCs can be confused with inflammatory lesions since the patients with this type of cyst usually have inflammatory symptoms such as pain, swelling and drainage [5].

Though odontogenic keratocyst are rare in maxilla especially in the second decade and in our patients it is present in maxilla and in second decade. In, our study the incidence is same as the reference mentioned above i.e., male to female ratio is 1.6:1. A combined endonasal and Caldwell-luc approach and only endonasal approach was performed in our patients and cyst removed in toto.

The histologic features of odontogenic keratocysts were defined by Pindborg and Hansen [6], as the cyst is lined by keratinizing stratified squamous epithelium that is usually thin and without rete pegs. The basal layer is well defined and consists of either columnar or cuboidal cells. The stratum spinosum is inconspicuous, may have vacuolated cells or may be absent. The layer of superficial cells is often wavy and parakeratotic. There usually is no significant inflammation. However, when inflamed, the cyst epithelium may exhibit rete pegs, and parakeratosis may disappear [7]. Satellite cysts are commonly described and are important in the biologic activity of keratocysts [8]. They are lined by thin keratinizing stratified squamous epithelium and may show continuity with the main cyst in serial sections.

Gorlin syndrome has been studied as an autosomal dominant disorder arising from defects on chromosome 9q23.1-q31 [9]. Some authors suggested that about half of odontogenic keratocyst cases are associated with this

syndrome [10]. When so associated, the cysts are usually multiple and the patient is younger. All cases associated with Gorlin syndrome had parakeratinized epithelial lining and showed a 6:1 male to female preponderance in the age range 11–20 years.

Panoramic and periapical images usually show well-defined borders of a similar density to calcified dental tissue, having a ground-glass appearance, and a radiopaque mass occupying the affected maxillary sinus, surrounded by a thin radiolucent halo [11]. CT is the gold standard, especially for ruling out any suspicion of associated orbital involvement, and revealing the spread and intracranial extension of infectious process. In cases when obstruction of sinus drainage is evident, one should be completely aware of serious complications such as orbital infections, epidural and subdural empyema, meningitis, cerebritis, cavernous sinus thrombosis, brain abscess and death [12].

Treatments of OKCs have ranged from marsupialization and enucleation to en bloc resection. According to Schmidt and Pogrel, the ideal treatment for the odontogenic keratocyst would be enucleation or curettage followed by treatment of the cavity with an agent such as liquid nitrogen or Carnoy's solution to kill the epithelial remnants or satellite cysts.

The most important feature of the OKC is its unusually high recurrence rate that ranges from 5 to 62.5 %. Brannon stated that the recurrence rate of keratocyst, which was treated with enucleation alone, was 12 %. Browne evaluated three different treatment methods, which were marsupialization, enucleation and primary closure and enucleation and packing open. Browne concluded that there was no correlation between treatment method and the rate of recurrence [13].

Recurrence of an OKC was due to the nature of the lesion itself, namely the presence of additional remnants of dental lamina, from which a cyst might develop. Schmidt and Pogrel [14] treated 26 keratocysts, with enucleation and cryotherapy, and three of the 26 patients (11.5 %) developed a recurrence after treatment. The authors evaluated that two of the three recurrences might have been secondary to technical errors and the recurrence rate for enucleation and cryotherapy might in fact be lower than 11.5 %.

Voorsmit et al. [15] compared two treatment methods: 52 keratocysts were treated with enucleation alone, and 40 keratocysts were removed along with excision of the overlying mucosa and treatment of the cyst cavity with Carnoy's solution. In the first group, 13.5 % of the cysts recurred, whereas 2.5 % of the cysts in the second group recurred. In Voorsmit's series of cases, the use of Carnoy's solution and soft tissue excision gave a very low incidence of relapse [15].

Conclusion

In conclusion, any cystic lesion should be evaluated cautiously. It generally thought to be derived from either the epithelial remnants of the tooth germ or the basal cell layer of surface epithelium. The incidence of male to female ratio is about 1.6:1. Odontogenic keratocysts may occur in any part of the upper and lower jaw with the majority occurring in the mandible, most commonly in the angle of the mandible and ramus. The maxillary sinus is a frequent site for pathologies of odontogenic origin due to its close anatomical relationship with teeth and periodontal tissues. Treatment includes marsupialization, enucleation and curettage, enucleation and peripheral ostectomy, osseous resection, cryotherapy which include the use of liquid nitrogen.

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