Case report

Title: ~~**ODONTOGENIC KERATOCYST MIMICKING A DENTIGEROUS CYST: A CASE REPORT**~~

A REVIEW OF THE ODONTOGENIC KERATOCYST AND REPORT OF A CASE

Abstract: The Odontogenic Keratocyst (OKC) is a developmental, non-inflammatory chronic cystic lesion that, on radiograph may be either unilocular or multi locular. OKC is a cyst of odontogenic origin, usually asymptomatic, with aggressive clinical behavior, ~~including~~ a high recurrence rate and a tendency to invade bone and adjacent soft tissues. ~~the adjacent tissues including bone~~. ~~Clinically OKC is manifested by an asymptomatic growth. Radiographically, it appears as a well-defined unilocular or multilocular osteolytic lesion. The diagnostic approach is based on a combined analysis of~~ Diagnosis is based on the clinical history, clinical appearance, radiographs, and histology. A case of odontogenic keratocyst involving the ramus of the mandible is presented in this article emphasizing the characteristics and various features of OKC.

Key words: *odontogenic keratocyst, keratocyst odontogenic tumor.*

Introduction:

Odontogenic keratocyst is a distinctive form of developmental odontogenic cysts that deserve special consideration because of its specific clinical behavior and histopathologic features. The term odontogenic keratocyst was first given by Philipsen in 1956[1]. OKC's most commonly occur in the second and third decades of life and show a slight predilection for males (males to female ratio 1.3:1). The recent WHO classification categorizes OKC as a developmental non-inflammatory odontogenic cyst that arises from the cell rests of dental lamina[2]. Majority of the OKCS occur in the mandible, most commonly in the angle- ascending ramus region.

The clinical & radiographic features of OKC are indefinite; while some may be associated with pain, swelling or drainage, most of them are asymptomatic. OKC'S commonly occur in the tooth bearing areas(82%) and some of the cases show an association with at least one impacted tooth (27% usually

the mandibular third molar)[3]. Here we report a case of odontogenic keratocyst associated with an unerupted mandibular third molar.

CASE REPORT:

~~A 50 year old female patient presented to our department on account of swelling and pus discharge in the left lower back tooth region of 4 months duration. History revealed that the swelling was initially small in size but gradually increased to the present size with associated history of difficulty in swallowing and foul taste. Past medical history revealed that the patient had been blind since 15 years.~~

The patient was a 50-year-old female who presented to our department with a chief-complaint of a slowly enlarging swelling, purulent discharge, and bad taste of 4 months duration from the left posterior region of the mandible. She was also having difficulty ehrn swallowing. Her past medical history was negative for major systemic diseases. She has been blind for the past 15 years.

~~On extra oral examination slight facial asymmetry was present due to the presence of a solitary swelling over the right mandibular angle, approximately 4 x 3.5 cms in its anteroposterior dimension & 3.5 x 3 cms supero inferiorly (Fig 1) Swelling was non tender and firm in consistency. Skin over the swelling was normal.~~

Clinical examination revealed a slight facial asymmetry due to a swelling over the region of the left mandibular angle. Swelling dimensions were approximately 4 x 3.5 CMs antero-posteriorly and 3.5 x 3 CMs supero-inferiorly (Fig 1) The swelling was non-tender and firm in consistency. Skin over the swelling was normal.

On intra-oral examination there was an obliteration of the buccal vestibule ~~in relation to 36, 37 teeth~~ of the molar teeth (Fig 2). The overlying surface of the swelling was of same color as that of surrounding mucosa. On palpation it was firm in consistency, non-tender, and on application of pressure a white creamy exudate was noted. Aspiration of the swelling yielded a cream colored hazy fluid (Figure 3).

A panoramic radiograph revealed a well-defined radiolucency on the left ramus of the mandible which was approximately 4x2x1 cm in size, oval In shape, extending anteriorly from distal aspect of 38 to posteriorly to 0.5 cm below the condyle; radiopaque scalloping margins with uniform radiolucency, expansion of inferior border of the mandible at the left angle region, and the inferior alveolar nerve canal ~~is pushed inferiorly~~ was displaced inferiorly (Fig 4). **YOU SHOULD MENTION WHETHER OR NOT THERE WAS EVIDENCE OF BONE FRACRURE.** Computed tomography revealed a ~~cystic lesion~~ cyst-like radiolucency with scalloped and well-corticated borders (Fig 5a & 5b).

Based on our clinical and radiographic assessment, our differential diagnosis included dentigerous cyst, odontogenic keratocyst, ameloblastoma, and odontogenic myxoma

~~A provisional diagnosis of odontogenic keratocyst of the left mandible was made with a differential diagnosis of dentigerous cyst.~~

~~Panoramic radiograph showed a well defined radiolucency on the left ramus of the mandible which was approximately 4x2x1 cm in size, oval In shape extending anteriorly from distal aspect of 38 to posteriorly to 0.5 cm below the condyle; radiopaque scalloping margins with uniform radiolucency, expansion of inferior border of the mandible at the left angle region, inferior alveolar nerve canal is pushed inferiorly (Fig 4). Computed tomography revealed a cystic lesion with scalloped and well corticated borders (Fig 5a & 5b).~~ Radiographic findings were suggestive of odontogenic keratocyst with a differential diagnosis of unicystic ameloblastoma, and odontogenic myxoma.

An incisional biopsy was performed under local anesthesia. The histology revealed stratified squamous cell epithelium with parakeratosis and prominent basal layer without rete ridges. Sub-epithelium contained fibro-collagenous stroma with islands of squamous epithelium (daughter cysts) (Figs. 6a and 6b). A definitive diagnosis of odontogenic keratocyst involving the left angle-ramus area was made.

TREATMENT: YOU SHOULD EXPLAIN WHY YOU CHOSE TO RESECT THE MANDIBLE RATHER THAN TRY TO DECOMPRESS THE LESION. I AGREE WITH YOUR TREATMENT BUT THE READER SHOULD UNDERSTAND YOUR DECISION.

Discussion:

The history of odontogenic keratocyst dates back to 1826 when Mickuliz first described it as a part of familial condition affecting the jaws. In 1926 it was referred to as “cholesteatoma.”- meaning a cystic or “open” mass of keratin squames with a living “matrix”. Later in 1945 Robinson mentioned this cyst as primordial cyst as they arose from remnants of the dental lamina or the enamel organs before enamel formation has had taken place. However it was not until 1956 the cyst ~~has got the name~~ was named odontogenic keratocyst by Philipsen [1]. Since then, the terminology has become a matter of dispute due to the distinct clinical, radiological, and histopathologic features of OKC. ~~and gained a special attention since last two decades~~. Some investigators classify OKC as a benign tumor. The aggressive nature of OKC caused the dilemma as to whether it is a cyst or neoplasm. In 1967, Toller suggested that OKC is to be called a benign neoplasm [(4)].Shear used the term “keratocystoma” citing the aggressive nature of the odontogenic keratocyst and finally labeled it as a benign cystic neoplasm. In 2005, the OKC was reclassified and renamed as keratocystic odontogenic tumor (KCOT) in the WHO classification of head and neck tumors[(5)]. Re-designation of the OKC as the KCOT is based on the well-known aggressive behavior of the lesion, histology and new information regarding its genetic makeup.

The patched gene PCTH, a tumor suppressor gene involved in both nevoid basal cell carcinoma syndrome and sporadic KCOTS, commonly occur in chromosome 9q22.3-q31.36-40 [(6)]. PCTH forms a receptor complex with the oncogene SMO (smoothened) for the SHH (sonic hedge hog) ligand. The growth-signal-transduction is inhibited by PCTH binding to SMO. This inhibition is released by SHH binding to PTCH. When the normal functioning of PCTH is lost, the proliferation stimulating effects of SMO are permitted to predominate [(7)]. But there was substantial evidence that PTCH gene mutation can also occur in non-neoplastic lesions like dentigerous cysts[(8).] Moreover, many researchers challenged the neoplastic process of OKC as marsupialization causes resolution of the cyst [(9)]. Hence the 2017 WHO classification reverted back to the well-accepted terminology of odontogenic keratocyst-OKC[(10)].

Most of the OKC’S arise from the cell rests of dental lamina or from the basal cells of oral epithelium and are thus primordial-origin odontogenic keratocysts[(11).] The remaining 40% arise from the reduced enamel epithelium of the dental follicle and are dentigerous-odontogenic keratocysts as in our

case. The clinical identification is crucial in determining the treatment as the recurrences are more frequently seen after treatment of primordial type of OKC. OKCs may occur at any age but the highest incidence is generally in the second and third decades of life. There is a slight male predilection. Approximately 20-45% of OKC'S are associated with un-erupted tooth and about 70% of the cases involve the mandible, especially the molar, angle, and ramus regions. In our case, all three distinctive characteristics are present.

OKC'S tend to grow in antero posterior direction within the medullary cavity of the bone and may cause an obvious bone expansion [12] A hazy radiolucent lumen can be seen on a conventional radiograph which is suggestive of a dense proteinacious material such as keratin. Resorption of roots of the erupted teeth is rare with displacement of teeth adjacent to the cyst occuring more frequently than resorption.

The histopathologic features of OKC are specific. Diagnostic features include a uniform cyst lining, hyperchromatic and palisaded basal cells, wavy parakeratin production, and a flat interference between the epithelium and connective tissue wall. One of the most important characteristic features of OKC is the appearance of satellite cysts/islands of odontogenic epithelium [13] as was seen in our case. The high recurrence rate of OFC can be attributed to the satellite cysts that can be retained after enucleation.

Unicystic ameloblastoma and orthokeratinized odontogenic cyst (OOC) present with the same clinical and radiographic features. However, histologically a unicystic ameloblastoma has an ameloblastic epithelial lining which is pathognomic of this cyst. The suprabasilar areas often loosen, giving the appearanceof a stellate reticulum[14]. OKC shares similar characteristics with orthokeratinized odontogenic cyst with respect to age of occurrence and site, yet these two lesions differ in their biological activity[15]. OKC can be differentiated from orthokeratinized odontogenic cyst on features like an older age group, more antero-posterior extension without expansion, characteristic histopathological features different from orthokeratinized odontogenic cyst, a parakeratinized layer, a high recurrence rate, an an association with basal cell nervous syndrome.

The treatment options for OKC'S range from simple conservative enucleation, (with or without curettage), or marsupialization. Marsupialization is a technique relying on incomplete removal of the cyst lining. Opening a window into the cyst forms an invagination of the oral cavity or the maxillary antrum.It relies on the principle that decompression halts expansion of the cyst and appositional growth of bone occurs, and the former cyst lumen becomes smaller with time. Many modification of the procedure have been taken place over the time like usage of decompression tubes, marsupialization catheter. One such method has been demonstrated by Costa et al where he used a segment of polyethylene suction tube, prepared according to the radiographic size of the lesion. Using a disposable needle, a hole is drilled near the extremity, large enough to allow the passage of a 0.8-mm orthodontic stainless steel wire. With the aid of a needle holder, one end of the wire is shaped into a loop and the other end is inserted through the hole in the tube, pulled back, and twisted. The tooth crown is etched with acid and the loop is attached to the dental surface with composite resin. An advantage of this technique over other methods is that it provides greater stability and minimizes the need for additional surgical interventions. For example, when compared to traditional methods where decompression devices were attached to the surrounding structures with sutures often resulted in an insufficient stability if there was a surgical wound dehiscence.[16]

Though conservative treatment preserves the anatomic structures they have a high risk for recurrence. The aggressive treatment includes peripheral ostectomy, chemical curettage with Carnoys solution, or *enbloc* resection. Carnoy's solution is composed of 3 ml of chloroform, 6 ml of absolute ethanol, 1 ml of glacial acetic acid, and 1 g of ferric chloride, is often used as a complementary treatment of lesions with high recurrence rates such as odontogenic keratocyst. The action of this solution is given by chemical cauterization, promoting a superficial necrosis of about 1.5mm of depth after 5 minutes of bone cavities exposure. In a study conducted by Albuquerque et al on surgical treatment with or without carnoy's solution in aggressive tumors of odontogenic origin the authors found a beneficial effect of the Camoy's solution in reducing the recurrence rate in several cases of jaw aggressive odontogenic tumors. This emphasizes the importance of carnoy's solution when used in conjunction with conservative procedures like enucleation.[17] **YOU NEED TO WARN THEW READERS OF THE DANGER OF NERVE INJURY FEOM CARNOY'S SOLUTION.** Also, as seen in our case, surgical intervention also

may include the necessity of resection of the diseased mandible – hemimandibulectomy (figure 7, 8) followed by reconstruction with titanium condylar plates (figure 9.

Conclusion:

Odontogenic keratocyst is a unique entity among odontogenic cysts due to its varied clinical, radiological and histopathologic features. This case report re-iterates the importance of histology in differentiating a odontogenic keratocyst from other odontogenic cysts of the mandible. Hence the correlation of histopathologic findings with clinical and radiographic features is of paramount importance to achieve a correct definitive diagnosis. These lesions have a very different biologic course so an accurate diagnosis is required to plan the proper surgical treatment.

Consent Disclaimer:

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

Figure 1: Extra oral clinical photograph showing swelling in the left lower half of the face.

Figure 2: Intraoral clinical picture showing a diffuse swelling in relation to 36, 37

Figure3: showing dense creamy exudate on aspiration

Figure 4: Panoramic radiograph showed a well defined radiolucency in relation to left molar ramus area.

Figure 5a and 5b: Computed tomography revealed expansile corticated and scalloped cystic lesion.

Figure6a: Histopathologic section shows stratified squamous epithelium showing parakeratosis

With prominent basal layer without rete ridges.

Figure 6b: Histopathologic section shows sub-epithelium islands of squamous epithelium (daughter cysts)

Figure 7: showing the exposed cyst involving the angle- ramus up to the condyle of the mandible.

Figure 8: showing the resected mandible along with the condyle

Figure 9: showing reconstruction of the defect with titanium reconstruction plates.

References:

1. PHILIPSEN HP,Om keratocyst(kolesteatomer) I kaeberne. Tandlaegeabladet 1956;60:963-80.
2. Haring J.I, Van Dis M.L Odontogenic Keratocysts , A clinical , radiographic and histopathologic study, Oral med Oral surg Oral pathol 1988: 66 : 145-53.
3. Mortazavi H , Baharvand M, Jaw lesions associated with impacted tooth: A radiographic diagnostic guide, Journal of Imaging sci dent; 2016; 46(3): 147–157.
4. Toller P , Origin and growth of cysts of the jaws. Ann R Coll Surg Engl 1967; 40(5):306-36.
5. Philipsen HP , Keratocystic odontogenic tumour. In: Barnes L,Eveson JW , Reichart P, Sidransky D, editors. World Health Organization Classification of Tumours: Pathology and Genetics of Head and Neck Tumours. Lyon: IARC Press; 2005. P. 306
6. Hemavathy S , Roy S , Follicular odontogenic keratocyst mimicking dentigerous cyst-report of two cases. Arch Oral Sci Res 2011;1:100-3.

7. Chaudhary D , Bhargava M, Aggarwal S Keratocystic odontogenic tumour-a case report with review of literature. Indian j Stomatol 2012;3:66-9.
8. Pavelic B , Levant S , PTCH gene alteration in dentigerous cyst. Journal of Oral Pathol Med. 2001;30:569-76
9. Soluk Tekkeşin, Merva & M. Wright, John. (2013). The World Health Organization Classification of Odontogenic Lesions: A Summary of the Changes of the 2017 (4th) Edition. Turkish Journal of Pathology. 34. 10.5146/tjpath.2017.01410.
10. Passi, Deepak & Singhal, Deepika. (2017). Odontogenic Keratocyst (OKC) or Keratocystic Odontogenic Tumor (KCOT). journey of okc from cyst to tumor to cyst again : Comprehensive review with recent updates on WHO classification (2017). international Journal of Current Research. 9. 54080-54086.
11. Marx and Stern (2012) Oral and Maxillofacial Pathology: A Rational for diagnosis and Treatment.2nd edition, 616-631
12. Pogrel M.A , The Keratocystic Odontogenic Tumor Oral Maxillofacial Surg Clin N Am 25(1) (2013) 21–30.
13. Shear M , The aggressive nature of the odontogenic Keratocyst:Is it a benign cystic neoplasm? Part 1. Clinical and early experimental evidence of aggressive behavior. Oral Oncol 2002;38:219-26.
14. Bajpai M , Agarwal D, Bhalla A, Kumar M, Garg M, Kumar M, Multilocular unicystic ameloblastoma of mandible. Case Rep Dent. 2013;2013:835892.
15. Sarvaiya B , Vadera H ,Sharma V, Bhad K, Patel Z, Thakkar M , Orthokeratinised odontogenic cyst of the mandible: A rare case report with systemic review: J Int Soc Prev Community Dent. 2014;4(1): 71–76.
16. Costa F. W. G , Carvalho F. S. R , Chaves F. Nobre ; Soares E. C.S . A Suitable Device for Cystic Lesions Close to the Tooth-Bearing Areas of the Jaws. Journal of Oral and Maxillofacial Surgery , 2014; 72: 96-98.

17. Albuquerque A. F. M. . Silva P. G. B. , Bezerra T. M. M. , Aleves A. P. N. N. , Pereira K. M. A. , Ribeiro T. R. , Soares E. C.S , Costa F. W.G . Surgical Treatment with or without the Use of Carnoy Solution in Aggressive Tumors of Odontogenic Origin: A Systematized Critical Literature Review. International Journal of Clinical Dentistry.2015 v. 9, p-2.

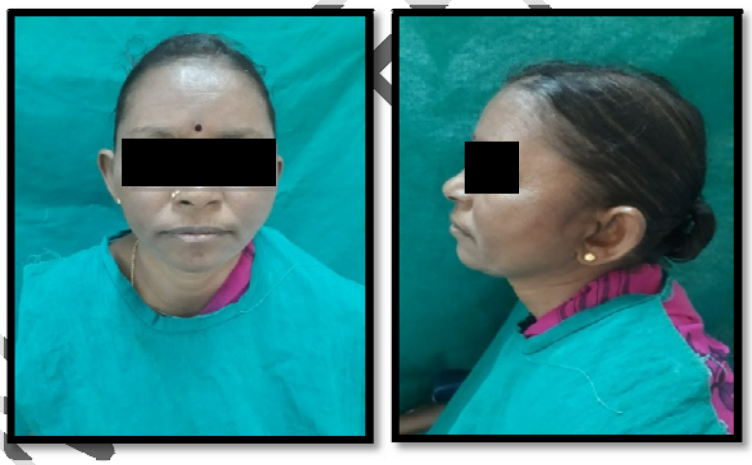


Figure 1:

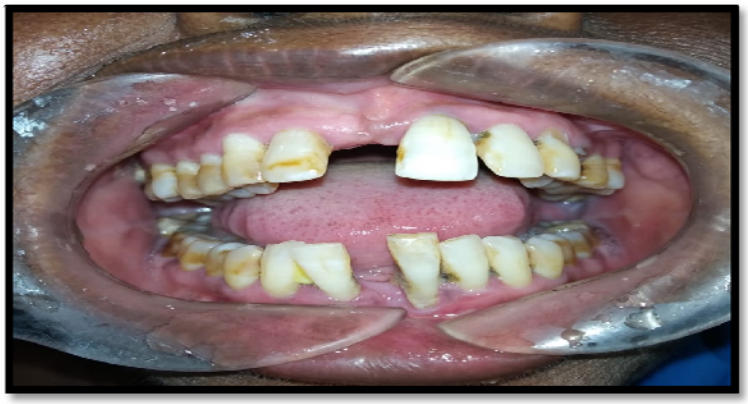
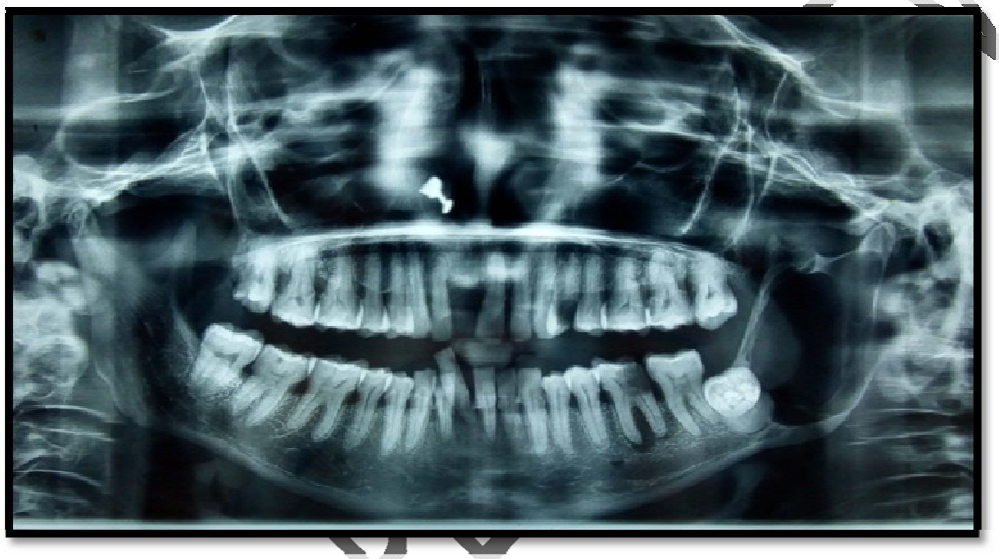


Figure 2:

Figure 3:



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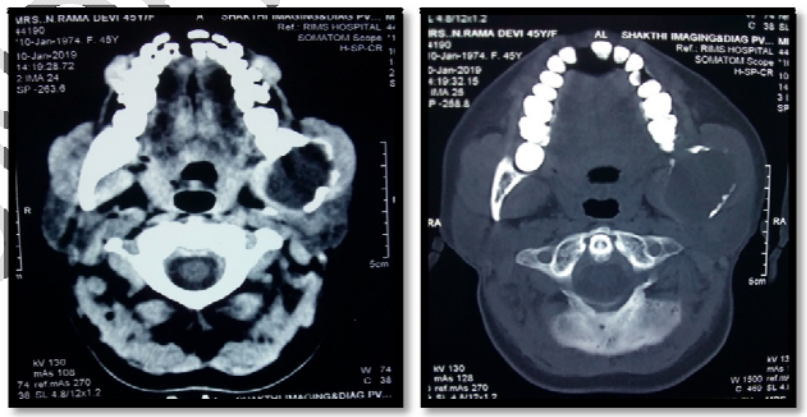

Figure 5a and 5b:

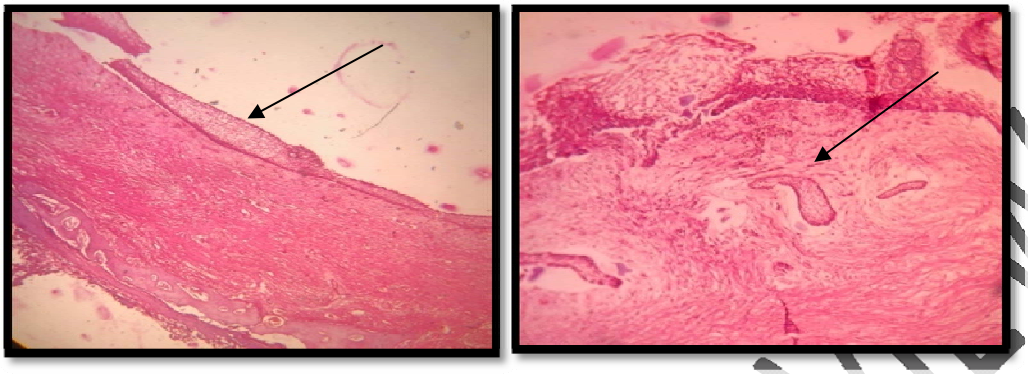

Figure 6a and Figure 6b.:

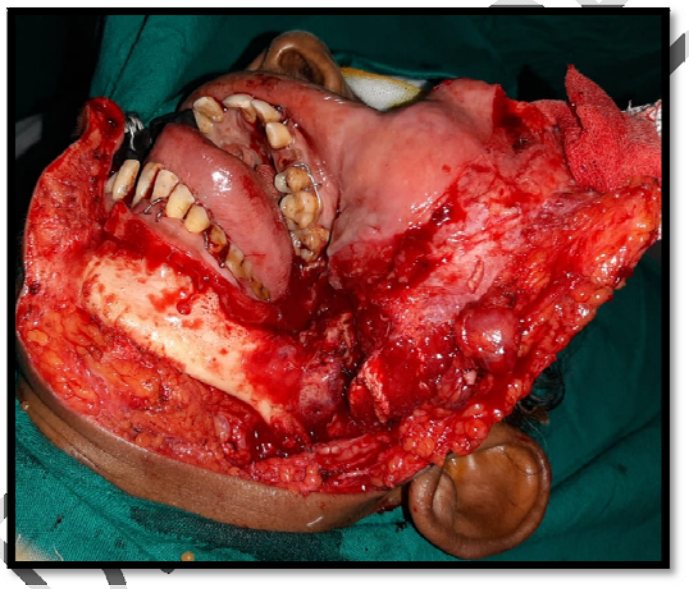

Figure 7

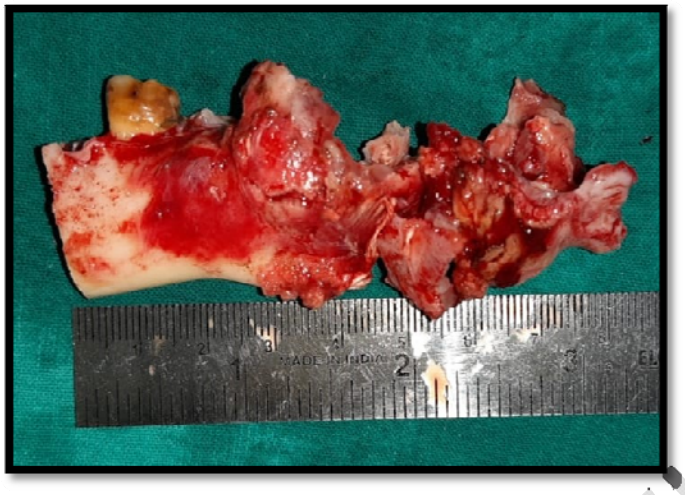

Figure 8

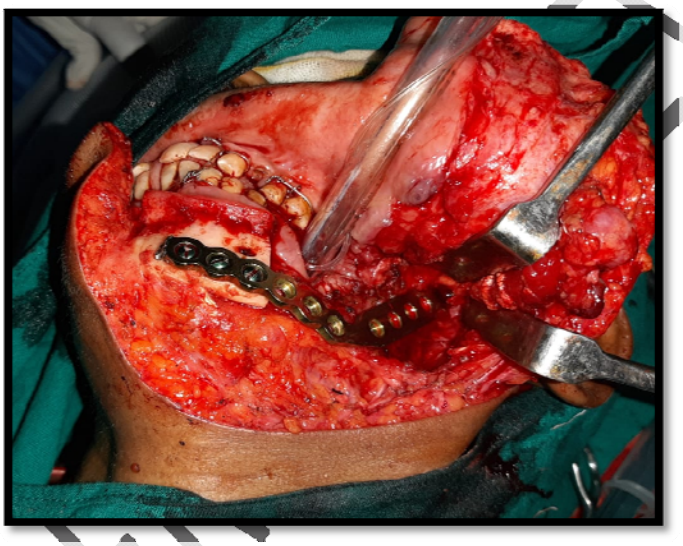

Figure 9