Multiple Odontogenic Keratocyst In Non-Syndromic Patient– A Rare Case Report

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ABSTRACT: The main purpose of this study was to reveal the occurrence of OKC in non-syndromic patients, its aggressive behaviour and to discuss its treatment protocol. A 25 year old young patient reported to our institute diagnosed with OKC which was confirmed by clinical, radiographical and histopathological evidences. This article mainly distincts the use of Carnoy's solution and BIPP paste in treating the patient and managing the patient conservatively than focusing on aggressive approach.

Introduction

Odontogenic Keratocyst (OKC) is a unique and distinctive local lesion which behaves aggressively with a high recurrence rate. OKC is a cyst of dilemmatic origin due to its eccentric nature of its neoplastic behaviour.

The term OKC had been under controversy from its introduction in the 1956 by Philipsen.[1] In 1967, Toller suggested OKC as benign cystic neoplasm instead of cyst.[2] This was later described as a cyst with keratinized lining by World Health Organization (WHO) in 1992.[3] Because of its aggressive nature and high recurrence rate WHO (2005) reclassified it askeratocystic Odontogenic Tumor (KCOT) and defined as "a benign unicystic or multicystic, intraosseoustumor of odontogenic origin, with a characteristic lining of parakeratinised stratified squamous epithelium with a potential for aggressive, infiltrative behaviour".[4]In 2017, due to lack of evidence of tumor now it is recognised as Orthokeratinized Odontogenic cyst (OOC).[4]

OKC is most commonly found in mandible occurring predominantly in posterior body and ascending ramus. [5] It may be seen as a single or multiple cystic lesions.Occurrence of multiple OKCis rare. [6] Usually multiple OKCs are associated with Basal cell nevus syndromewhereas few cases of multiple OKCs are also reported in nonsyndromic patients. [7] In this paper, we are presenting a case report of bilateral OKC in a nonsyndromic young male patient.

A 25 year old male patient was referred to our department with a chief complaint of swelling on the right side of face since 3 months.**[Fig. 1a, 1b]**Patient had a complaint of mild pain in the right side of the jaw since 20 days. Patient visited a local dentist few days back for the same complain where a panoramic radiograph was advised and routine antibiotic therapy was prescribed for 5 days. There was no history of fever or pus discharge. Patient had no history of trauma. His past dental history revealed that he had undergone root canal treatment of the maxillary right 1st molar tooth 2 years back. Patient had no significant medical history.

Fig.1a, 1b showing mild swelling on right side of face.

raoral examination, present at right mandibular angle region**[Fig. 1a].** Right submandibular lymph nodes was palpable with a restricted mouth opening of 2 cmsapprox. **[Fig. 2].** There was obliteration of buccal vestibule with mild swelling extending from right canine to the right second molar region**[Fig. 3].** Swelling was soft, fluctuant and non tender on palpation. Submucous fibrous bands were palpable bilaterally. Buccal and

lingual cortical plate were expanded with no perforation.

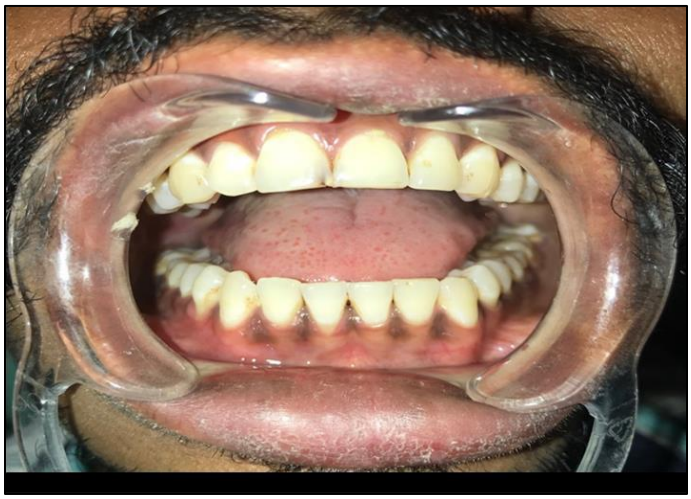


Fig. 2 – Reduced mouth opening.

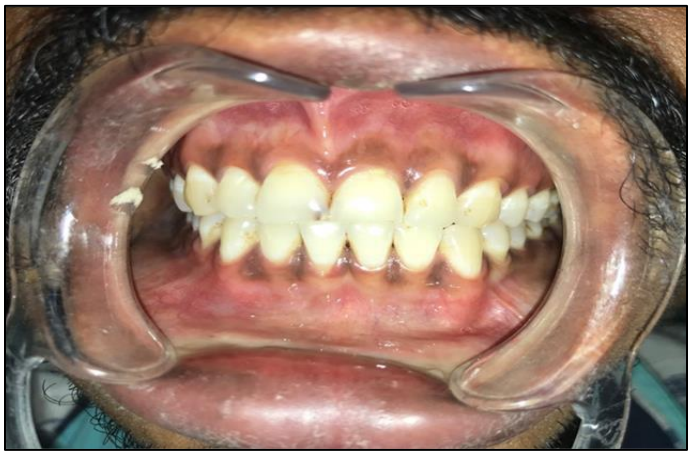


Fig. 3- obliteration of buccal vestibule

Investigations

On radiographic examination, OPG revealed a radiolucency on the right side extending from distal surface of second premolar (45) to entire mid ramus area or to the sigmoid notch extending to the lower border of mandible associated with right impacted mandibular third molar (48). The radiolucency was irregular in shape with well corticated scalloped borders. On left side, a well-defined unilocular radiolucency is seen involving distal root of 38 which is oval in shape and approximately 2 × 2 cm in size **[Fig. 4].**Chest radiograph showed absence of bifid ribs with normal lungs.

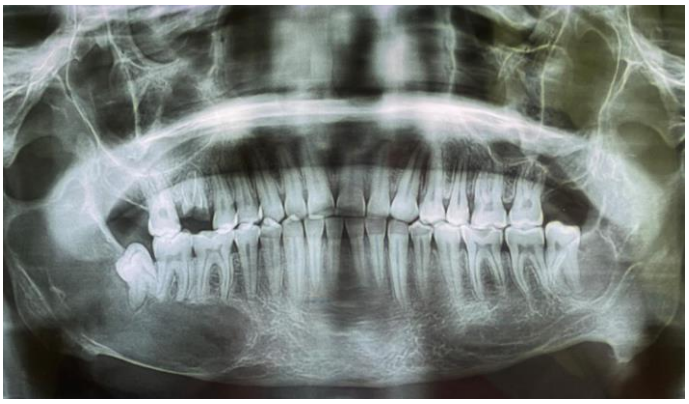
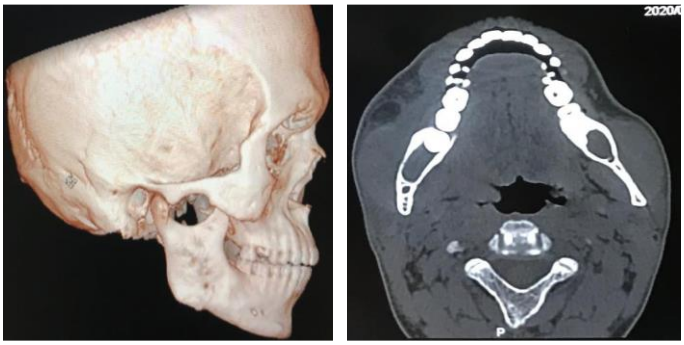


Figure 4 - OPG revealing radiolucency radiolucency on the right side extending from distal surface of second premolar (45) to entire mid ramus area or to the sigmoid notch associated with right impacted mandibular third molar (48). On left side, a well-defined unilocular radiolucency is seen involving distal root of 38.

Computed tomography (C.T.) was performed which demonstrated obvious buccolingual expansion of cortical bone of right side. Cortical plate were intact with no perforation**[Figure 5].** Deflection of inferior alveolar canal seen on right side of mandible. Fine-needle aspiration represented yellow thick white "cheesy" material.

Figure 5- 3D reconstruction and axial section showing buccolingual expansion (arrows) of the right side body and ramus of the mandible. The cortical plates are intact.



Based on the clinical and radiographic examination, a differential diagnosis of OKC, Dentigerous cyst, Ameloblastoma were outlined. After performing incisional biopsy, the histopathological features were suggestive of BILATERAL ODONTOGENIC KERATOCYST. Blood investigations revealed normal reports.

Treatment

Under all aseptic precaution, left nasotracheal intubation done and G.A. was induced. Local infiltration was done using 1:80000 adrenaline. On the right side, buccal vestibular incision was given extending from anterior border of ramus distally to the right 3rd molar carried anteriorly around the cervices of teeth anteriorly till distal to 2nd premolar**.** Full thickness mucoperiosteal flap was raisedfollowed by extraction of all involved tooth (46, 47, 48**).** Labial ostectomy was performed and cyst was enucleated. The cystic cavity was curetted. Carnoy's solution-soaked gauze was placed in the cystic cavity for 3 mins followed by irrigation**.** The resulting cavity was packed with povidone iodine gauze.**[Fig. 6a-e]**

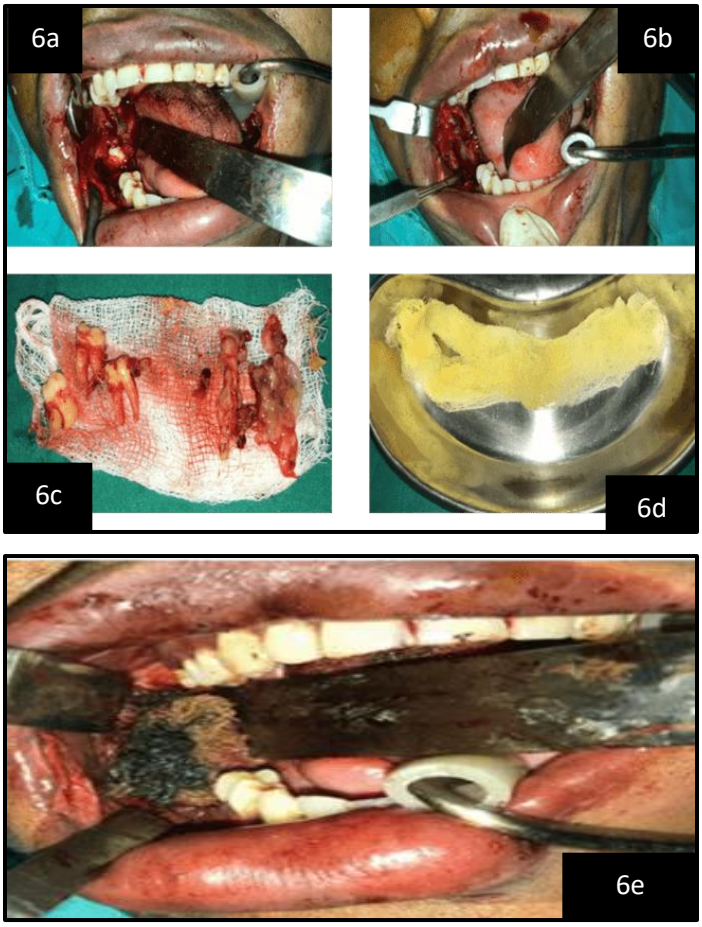


Figure 6. Intra operative Photographs of Right side.
Figure 6a: Buccal vestibular incision was given.
Figure 6b: Mucoperiosteal flap being raised.
Figure 6c: cystic lining with extracted involved teeth (46, 47, 48).
Figure 6d: Carnoy's solution-soaked gauze was placed in the cystic cavity for 3 mins.
Figure 6e: The cavity was packed with povidone iodine gauze.

On the left side of mandible, extraction of 38 was done and cavity has exposed. Cystic lining was removed and cavity has curetted thoroughly**.** Primary closure was done using 3.0 vicryl suture **[Fig. 7a-c].** Then specimen was sent to laboratory for histopathological examination**.**

Post-operative antibiotics and analgesics were prescribed. Follow up dressing was done with Bismuth iodoform paraffin paste three times in a week for 2 months. Acrylic prosthesis was given to the patient after 2 months**[Fig. 8].**

Haematoxylin & Eosin stained sections of both the lesions showed the parakeratinized stratified squamous epithelium with 5-8 call layer thickness. The lining epithelium showed palisaded tall columnar basal cells. The supportive connective tissue showed numerous thick bundles of collagen. Diffuse chronic inflammatory cells were also seen **[Fig.9].**

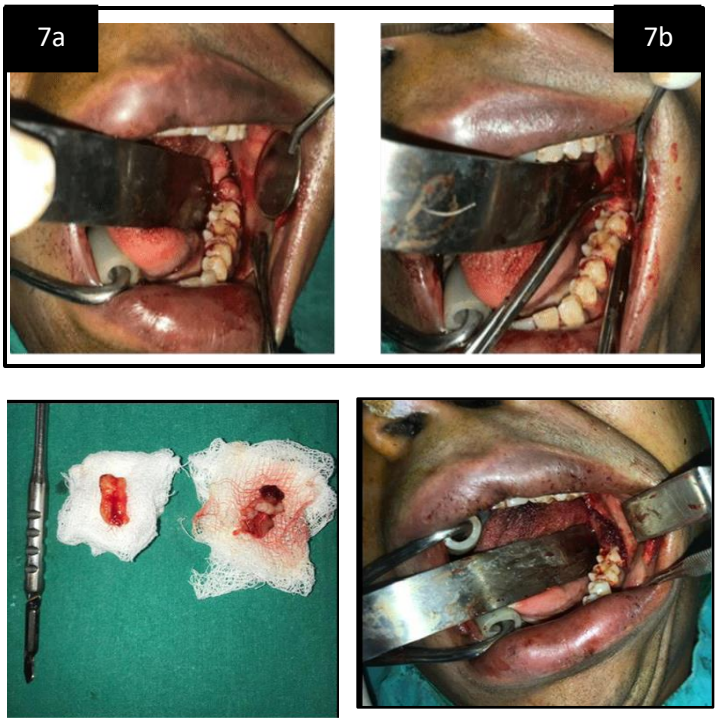


Figure 7 – Intra operative photographs of left side.
Fig. 7a: On the left side of mandible, extraction of 38 was done and cavity has exposed.
Fig. 7b: Cystic lining with extracted 3rd molar
Fig. 7c: Primary closure was done using 3.0 vicryl suture.

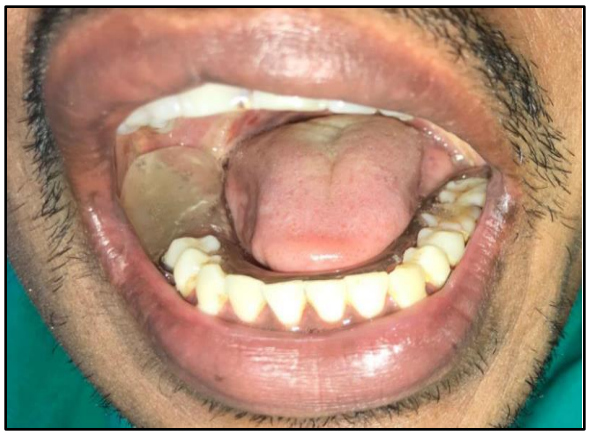
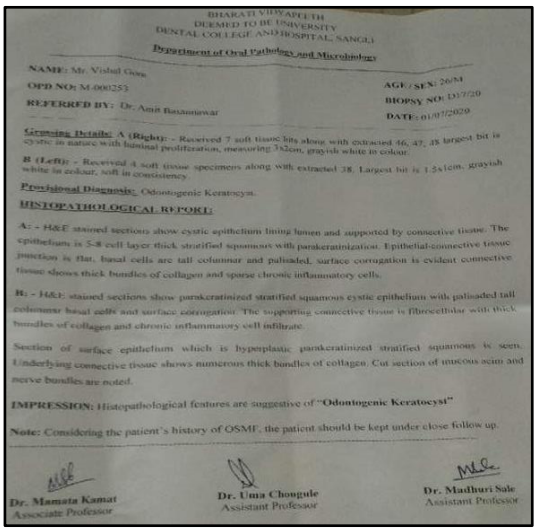


Fig. 8- Post -op obturator placed.



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Grossing Details: A (Right) - Received 7 soft tissue bits along with extracted 46, 47, 48 largest bit is cystic in nature with luminal proliferation, measuring 3x2cm, grayish white in colour.

B (Left) - Received 4 soft tissue specimens along with extracted 38. Largest bit is 1.5x1cm, grayish white in colour, soft in consistency.

Provisional Diagnosis: Odontogenic Keratocyst.

HISTOPATHOLOGICAL REPORT:

A: - H&E stained sections show cystic epithelium lining lumen and supported by connective tissue. The epithelium is 5-8 cell layer thick stratified squamous with parakeratinization. Epithelial-connective tissue junction is flat, basal cells are tall columnar and palisaded, surface corrugation is evident connective tissue shows thick bundles of collagen and sparse chronic inflammatory cells.

B: - H&E stained sections show parakeratinized stratified squamous cystic epithelium with palisaded tall columnar basal cells and surface corrugation. The supporting connective tissue is fibrocellular with thick bundles of collagen and chronic inflammatory cell infiltrate.

Section of surface epithelium which is hyperplastic parakeratinized stratified squamous is seen. Underlying connective tissue shows numerous thick bundles of collagen. Cut section of mucous acini and nerve bundles are noted.

IMPRESSION: Histopathological features are suggestive of "Odontogenic Keratocyst"

Note: Considering the patient's history of OSMF, the patient should be kept under close follow up.

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Fig. 9- Histopathological Report

Out come and follow ups -

Patient has been asymptomatic and exemplary healing is achieved since 1 year. Reduction in the size of the defect is seen upto 80 % clinically and radiographically as well. Due to high recurrence rate of the lesion the patient has been put under regular follow ups**[Fig. 10, 11, 12].**

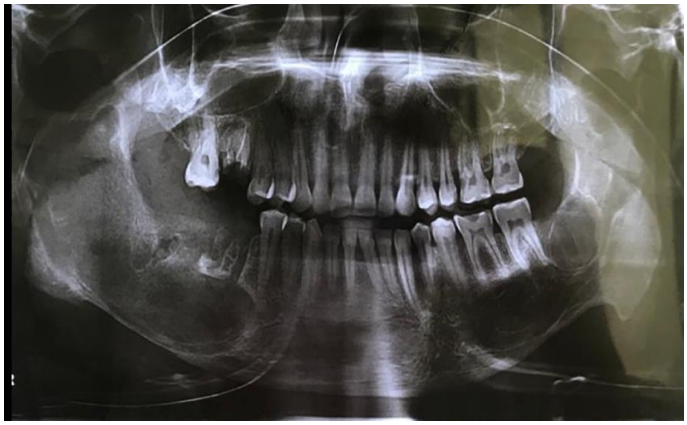

Fig. 10- 1ST WEEK FOLLOW-UP

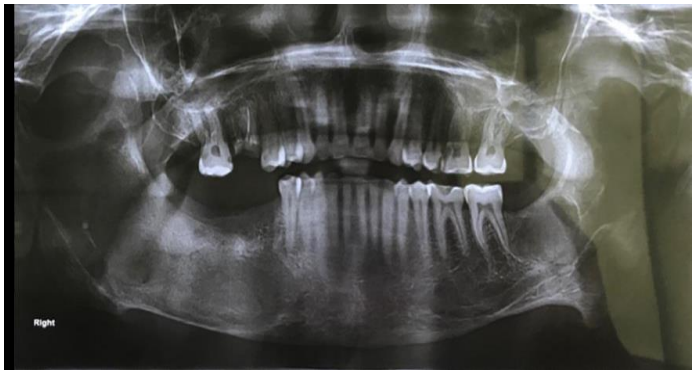

Fig. 11-3rd MONTH FOLLOW-UP

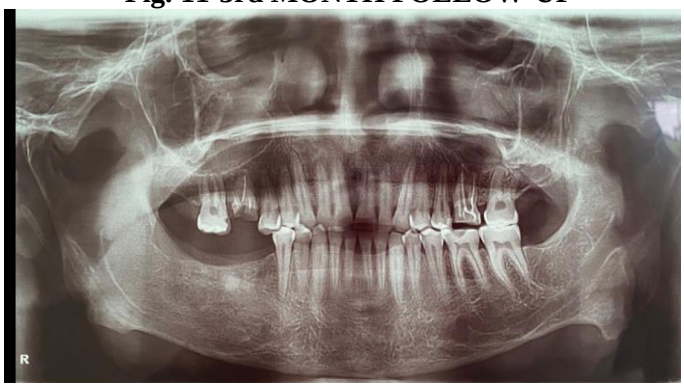

Fig. 12 – 1 YEAR FOLLOW-UP

Discussion

Odontogenic keratocyst is a most aggressive developmental cyst affecting the maxillofacial region arising from the cell rests of dental lamina or basal cell layer of oral epithelium usually are seen during the 2nd to 4th decade of life.[8] OKC involves approximately 10-15 % of all cystic lesion and to be more frequent in males (M/F 2:1).[9,10,11] Around 65-83 % of OKCs occur in mandible, more frequently found in posterior part of the jaw.[4,12,13]

OKC is the third most commonly occurring which accounts for 5.4% to 17.2% of all odontogenic cysts, periapical cyst (52.3-70.7%) and dentigerous cyst (16.6-21.03%) being 1st and 2nd most commonly occurring odontogenic cyst respectively.[14,15] OKC is mostly seen as enclosing the crown of the teeth thereby occurring similar to that of dentigerous cyst in 80 % of cases.[16] In about 25-40 % of cases OKC is associated with unerupted or impacted teeth.[11] Our case shows this characteristic involving mandibular right & left impacted 3rd molar. One of peculiar features of OKC is that is grows in antero-posterior direction in medullary spaces of bone

because of which cortical expansion is not seen in initial stages.[17,18]

Multiple OKCs are typical feature of nevoid basal cell carcinoma syndrome(NBCSS) also known as Gorlin-Goltz Syndrome however, rarely in some cases multiple OKCs are present in non-syndromic patients as well , our patient being one of the non-syndromic ones.[19] Characteristic features of NBCSS include basal cell carcinoma of the skin, bifid ribs, falxcerebri, stunted growth, bleeding diathesis, hyperextensible skin, hypermobile joints, cleft lip/palate, medulloblastomas, ocular malformations, frontal bossing, facial milia.[20,21] Recently other features such as low pitched voice, talon's cusp, supernumerary teeth, sloping shoulders, congenitally missing third molars are also considered for diagnosing NBCSS.[22]

Our patient was examined for these features by thorough clinical evaluation and CT scan which revealed absence of all these features therefore our patient being non-syndromic.In most of the cases, multiple OKCs may be the first and only sign in diagnosing NBCSS because of which most of the cases are first diagnosed by dentists, however other features of NBCSS may develop after a decade because of which our patient has been kept under follow up for other manifestations of NBCSS. [23]

Multiple OKCs are also seen in multiple syndromes such as orofacial digital syndrome, Ehler-Danlossyndrome, Noonan syndrome and Simpson – Golabi – Behmet syndrome. [24,25] Due to high proliferation rate of epithelial lining in syndromic cases, KCOTs related to NBCSS have more aggressive behaviour and high recurrence rate (82%) as compared to non-syndromic ones. [26]

Managing OKC has always been controversial as it depends upon multiple factors such as patient age, size & location of cyst, radiological features, and histological features as well as due to its high recurrence nature.[27,28] There are two approaches for managing OKC- conservative approach and aggressive approach. Conservative approach incorporates enucleation& marsupialization and aggressive approach includes peripheral ostectomy, chemical curettage with carnoy's solution, cryotherapy or electrocautery & resection.[29] Our patient being young was chosen for conservative management. Our patient was managed with enucleation and curettage with carnoy's solution which has shown to have decreased recurrence rate upto 2.5% as going for enucleation alone (13.5%).[30]

In this case, we have made use of Bismuth Iodine Paraffin Paste (BIPP) as it makes the gauze impermeable to body fluids and blood thereby providing minimum nutrition to the bacteria to survive.It does not initiate or promote wound healing but does provides an adequate environment for the healing take place. This BIPP was first introduced by Rutherford Morrison in 1916 which was routinely used to pack nasal cavities. BIPP is available either in the form of paste or in the form of sterile gauze packed with paste which includes one part of bismuth sub nitrate, two parts of iodoform and one part of sterile liquid paraffin by weight.[31]

Table – Role of the different constituents of BIPP.

Material	Quantity	Function	Adverse reaction
Bismuth Subnitrate	**250 mg/g**	**Antiseptic and Astringent Releases dilute nitric acid on hydrolysis**	**Encephalopathy caused by bismuth toxicity – causing headaches, nausea stomatitis and bismuth line in gingiva [31]**
Iodoform	**500 mg/g**	**Decomposes to release iodine Antiseptic and Antibacterial [31]**	**Hypersensitivity reaction causing an erythematous rash. Iodoform poisoning causing irritation of eyes and skin along with gastric and respiratory irritation[32]**
Paraffin Base	**250 mg/g**	**Acts as a lubricant aids in atraumatic placement and removal of BIPP [31]**	**Sensitivity reactions, Acne [33]**

Conclusion

Multiple OKCs when present in a patient should be thoroughly examined for other manifestations associated with syndromes. Non-syndromic patients should be kept under regular follow ups as the features related to syndrome tend to appears in later decades of life. Managing young patient conservatively rather than going aggressively can be better option in most of the cases. In our opinion, the lesion can be treated efficiently with enucleation and by using Carnoy's solution followed by use of BIPP in post-operative period instead of aggressive approach.

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