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A Case Report of Paget's Disease of Bone Involving Maxilla and Mandible: A Diagnostic Dilemma

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Abstract:- A chronic, progressive bone illness with an unknown aetiology, Paget disease of bone (PDB) causes excessive, disorganised bone growth that results in pain, fractures, and deformities. It starts out by increasing bone resorption. Both monostotic and polyostotic illness manifestations are possible. The Anglo-Saxon population has a high prevalence of PDB, yet In India, they are not common. The condition is frequently asymptomatic and is prevalent in elderly populations. The majority of the time, radiographic analysis and biochemical indicators of bone turnover are used to determine the disease's diagnosis. While calcium and phosphate levels are normally within normal ranges, markedly increased serum alkaline phosphatase (SAP) is a frequent feature. We disclose a case of a 65-year-old male patient with maxillary-mandibular involvement. Although PD is frequently seen in western nations, it is uncommon in Asian populations.

Keywords:- *Cotton wool appearance, leontiasis ossea, Paget's disease of bone, serum alkaline phosphatase, Bone disease, bone turnover, osteitis deformans.*

I. INTRODUCTION

Sir James Paget, a British surgeon, first identified Paget disease of bone (PDB) in 1877. He called the disorder "osteitis deformans" because he thought it was caused by persistent inflammation This osseous dysplasia affects middle-aged and older people. PDB can damage the axial skeleton in either monostotic or polyostototic forms, with the former being the more prevalent condition.[1] However, different bones in the craniofacial complex may be impacted to differing degrees. It is distinguished by a localised change in bone remodelling, which results in a bone with an abnormal structure and changed mechanical characteristics, linked to pain and pathological fractures. There could potentially be complications including heart failure and neurological impairments.[1,2]

The radiograph displays the typical look of cotton wool, and other radiographic findings include well-circumscribed radiolucency, lamina dura loss, root resorption, hypercementosis, and radioopacity. Although the cause of PD is yet unknown, genetic and environmental factors might be involved. It has also been suggested that viruses such paramyxovirus, canine distember virus, and respiratory syncytial virus are to blame.[3]

Among the often employed therapeutics include mithramycin, bisphosphonates, and calcitonin. According to a recent clinical trial, second- and third-generation bisphosphonates, compared to calcitonin and editronate, the first generation bisphosphonate, bisphosphonates like pamidronate and alendronate were more efficient.[4]

Here, we describe a remarkably uncommon instance of PD with maxillilary-mandibular involvement.

II. CASE REPORT

A 65 years old male patient reported to the dept of OMDR and his chief complaint was pain and swelling in the lower right front tooth region since 1 week. Patient was apparently asymptomatic 1 week back when while eating he experienced pain in the lower right front tooth region. Pain was sudden in onset, mild, dull, continous, non-radiating and relieved on taking medication. It was followed by swelling and pus discharge 2 days later. The swelling was initially localized in the front tooth region but gradually increased over a period of 5 days to involve the right lower half of the

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face. The pus discharge is present in the affected area on application of pressure.

Past medical history was not significant but past dental history revealed, extraction at the same site 6 months back. He got the extraction done due to pain and swelling in that region. The pain and swelling subsided following extraction and medication.

On extra oral examination revealed facial asymmetry due to presence of 3 swellings on the right side of the face. The swelling was apparent on the right forehead (since childhood), right side of nasal bridge and lower one third of the right side of face adjacent to midline.

Diffuse swelling in the upper one third of the right side of forehead adjacent to midline, measuring approximately 4 x 3 cms. (Fig.1)

The second swelling was present on the right side of the nasal bridge. It is a diffuse swelling measuring approximately 3 x 2 cms, obliterating the nasolabial fold and extending below the right lower palpebral margin. (Fig.1)

Third swelling was seen in the lower one third of the right side of the face in the chin region adjacent to the midline, measuring approximately 3 x 5cms, extending medially from the symphyseal region to the right corner of the mouth, superiorly from the vermillion border of lower lip to 2 cms below the lower border of mandible. (Fig.1)

Margins of all the three swellings were diffuse. Overlying skin was stretched. No secondary changes and visible pulsations were seen.

On palpation, the inspectory findings of site, size, shape and extent were confirmed. The temperature overlying the swelling was not raised. The swellings was non tender, firm to hard in consistency. Margins were diffuse, overlying surface is smooth. The swellings was not movable and attached to the underlying bone. The swellings was non-compressible, non-fluctuant and non-pulsatile. Single right submandibular lymph node was palpable. It was firm in consistency and non-tender.

Intra oral examination revealed the size and shape of the mouth was normal. The mouth opening and jaw movements were normal in all the directions. No deviation of jaw on opening was present.

Total number of missing teeth were 11 12 42 43, medially displaced teeth were 13, 21. Discharging sinus present in the vestibular region of 11 12 42 43. Grossly decayed tooth was 36. Root stumps present were 11 28 46 48. Diffuse swelling was present in the right anterior region of hard palate irt 14 to 22. In the floor of the oral cavity single, irregular, well-defined swelling was present irt 32 to 45. (Fig.2 & 3)

There was a single irregular swelling in the lower right front teeth region, measuring approximately 4 x 4 cms, extending from the distal aspect of 32 to the distal aspect of 45 with buccolingual expansion. Buccally the swelling extended from the alveolar mucosa to the buccal vestibule and lingually from the alveolar mucosa to the lingual vestibule. Margins of the swelling were distinct. The overlying surface was lobulated. The mucosa on the labial aspect was not intact, with sinus irt 42 and 43 which was associated with pus discharge. (Fig.2 & 3)

Diffuse swelling in the upper right anterior region of hard palate, measuring approximately 4 x 2 cms, extending from the distal aspect of 14 to the distal aspect of 22 with buccopalatal expansion. Buccally the swelling extended from the alveolar mucosa to the buccal vestibule and palatally from the alveolar mucosa to 1 cm in the medial direction towards the midline of the palate. Margins of the swelling were diffuse. The mucosa on the labial aspect was not intact, with sinus irt 11 and 12 which is associated with pus discharge. (Fig.2 & 3)

On intraoral palpation, the inspectory findings of site, size, shape and extent were confirmed. The swelling in the lower right front teeth region was non tender. On the labial aspect it was firm in consistency and hard on the lingual aspect. Margins of the swelling were distinct and overlying surface was lobulated. The swelling was not movable and attached to the underlying bone. It was non-compressible, non-fluctuant and non-pulsatile. On the labial aspect there was a sinus irt 42 and 43 associated with pus discharge. The associated teeth were not mobile.

The swelling in the upper right anterior region of hard palate was non-tender, hard in consistency with diffuse margins. The palatal mucosa is smooth while on the labial aspect there is sinus irt 11 and 12 associated with pus discharge. The swelling was not movable and attached to the underlying bone. It was non compressible, non-fluctuant and non-pulsatile. The associated teeth were not mobile.

Hence, on the basis of history and clinical examination, a provisional diagnosis of ‘Chronic Suppurative Osteomyelitis’ was given. Differential diagnosis were given as, Polystotic fibrous dysplasia, Paget’s Disease, Florid cement osseous dysplasia, Generalized osteopetrosis, Multiple osteomas.

On radiological investigation, intra oral periapical radiograph of maxillary anterior region revealed missing 11 and 12. There was mesial tipping of 13 and 21 with distally displaced roots. Multiple radiopaque masses were seen with few radiolucent areas.

Intra oral periapical radiograph of right mandibular anterior region showed 31, 41 and 44 .42 and 43 were missing. There were multiple radiopaque masses having a cotton wool appearance. The radiopacities obscured the root of 31, 41 and 44. (Fig. 4 & 5)

In the mandibular occlusal topographic view 42 and 43 were missing. There was an increase in the density of bone in the anterior part of the right side of mandible with expansion of buccal and lingual cortical plates. (Fig.6)

In the maxillary occlusal topographic view, teeth 11 and 12 were missing. Radiopaque masses interspersed with few radiolucent areas were seen in the anterior part of the right

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side of maxilla. 13 and 21 exhibit mesial tipping with distally displaced roots. (Fig.7)

Panoramic radiograph showed normal left and right condyles. In the mandible, 42 and 43 were missing and there root stumps irt 28, 36 and 46. There was a radiopaque mass in the right body of the mandible measuring approximately 5 x 2 cms extending from the distal aspect of 32 to the distal aspect of 45. Superiorly it obscures the roots of 32, 31, 41, 44 and 45, which exhibit distal displacement and inferiorly it extends 1 cm above the lower border of the mandible. In the maxilla, 11 and 12 were missing. There were multiple radiopaque masses intermixed with few radiolucent areas in the anterior part of the right side of maxilla involving the alveolus i.r.t. 11 and 12, measuring approx. 2 x 2 cm., extending superior-inferiorly, from the right inferior nasal concha to the maxillary alveolus. (Fig.8)

Paranasal sinus view revealed, multiple radiopaque masses intermixed with few radiolucent areas measuring approx. 2 x 2 cm. extending superior-inferiorly, from the right inferior nasal conncha to the maxillary alveolus. Anterior-posteriorly, it involved the alveolus i.r.t. 11 and 12.

In the coronal section of CT, multiple radiopaque masses intermixed with few radiolucent areas in the anterior part of the right side of maxilla involving the alveolus i.r.t. 11 and 12 were seen (Fig.9).

On aspiration, no fluid is drawn from the swelling. On biopsy, the decalcified hard tissue section revealed multiple criss crossing resting and reversal lines giving an appearance of mosaic pattern or jigsaw puzzle pattern of bone (Fig 10).

After corelating the clinical, radiological and histopathological findings a final diagnosis of Paget's disease was given.

III. DISCUSSION

Although Sir James Paget first referred to it as "osteitis deformans" in 1877, this is a misnomer. PDB is a persistent, non-inflammatory condition that causes regional bone remodelling. The word "osteodystrophia deformans" would be more appropriate because it affects numerous, non-contiguous sections of the skeleton. Following osteoporosis, PDB is the second most prevalent bone disorder.[2]

Although PD is a condition that affects a fair number of people in western nations including England, the United States, New Zealand, Canada, South Africa, and France, incidence in the Asian population is higher. That is also extremely uncommon among Indians. The highest recorded prevalence is in Britain. It is not equally distributed throughout all the states in Britain or America. In North America, Yorkshire and Lancashire are specific regions with high incidence. Despite the disease being uncommon in Africa, New York has a far greater prevalence than Atlanta. Black and white Americans each experience the same incidence.[6]

With a little male gender predisposition, this disease affects roughly 34% of the population over 50 and is rather common among elderly persons. Juvenile Idiopathic hyperphosphatasia, often known as Paget disease, is a rare metabolic bone dysplasia that is inherited as an autosomal recessive condition. It is unrelated to the conventional PDB that affects the elderly and is characterised by increased bone turnover as a result of increased osteoclastic activity.[7,8]

PDB's pathophysiology has been linked to both genetic and environmental variables, which are the focus of ongoing research into the disease's cause. Genes are undoubtedly a factor. Considering that 15–40% of affected patients have a first-degree relative who has PDB, this is a significant factor in the genesis of PDB. Canine distemper virus was located in the pagetic osteoclasts, and inclusion bodies resembling paramyxovirus nucleocapsid particles were seen.[9,10,11]

The maxilla-to-mandible ratio of 2:1 indicates jaw involvement. As seen in the cases, the maxilla progressively enlarges, the alveolar ridge widens, and the palate flattens. The current instance, teeth may move and become movable, creating some space. Patients with edentulous conditions who wear dentures frequently lament how tightening caused by the growth of the jaws make them harder to wear.[8]

When the maxilla increases in size it may encroach to orbit, the nasal cavity and the antra. PDB sufferers may experience "Leontiasis Ossea," or a lion-like face, which is characterised by an overgrowth of the facial and cranial bones. However, people with severe lepromatous leprosy may also experience it. The mandible and maxilla affected by PDB are extremely prone to infection, especially by organisms prevalent in the normal oral flora. Dental extractions could be more challenging, because the roots could be ankylosed or hypercementosed. Localized osteitis is frequent, and the healing of the extraction sites is frequently sluggish. Osteomyelitis is a common side effect of localised extraction sites infection in individuals.[12,13]

In addition to bone pain and gait issues brought on by a shortened, bowed leg, other symptoms of pagetic bone deformities may include an enlarged skull, kyphosis, and bowing of the affected femur or tibia, lumbar spinal stenosis or other disorders of neural compression with accompanying sensory or motor impairments, headaches or hearing loss involving a significant portion of the skull. The enhanced vascularity of pagetic bone greatly increases the risk of heart failure. Osteosarcoma that develops in the damaged bone is an uncommon and terrifying consequence that affects 0.3% of patients.[14,15,16]

Jaw PD is typically diagnosed based on clinical and radiological evidence. The primary method for diagnosing PDB is radiological evaluation. Early on in the disease's progression, localised osteolytic lesions are caused mostly by lytic activity. Therefore, regions of the development of sclerosis, which results in the recognisable symptoms of mixed lytic and sclerotic regions, thickened trabecula, bone expansion, cortical thickening, and deformity. Although the radiographic appearances are typically recognisable, it is occasionally necessary to consider the differential diagnosis of sclerotic or lytic metastases.[5]

All patients may be advised to undergo a radioisotope bone scan as part of the first diagnostic evaluation to ascertain the disease's spread, particularly the involvement of areas that could lead to difficulties include the spine, long bones, and the base of the skull. The radiotracer may occasionally be taken up by the mandible when it is heavily involved, resulting in the "Lincoln sign" or "Black beard" sign. Particularly for patients with deafness, spinal stenosis, or other neurological problems, computed tomography scanning is useful for assessing skull-base involvement.[4,12,17]

Large patches of radiolucency are shown on skull radiographs as a result of progressive osteolysis, which is also known as "osteoporosis circumscripta" and typically affects the frontal and occipital bones. later noted the diploic space, often known as a "Tam O'Shanter" skull, is noticeably enlarged when the inner calvarial table thickens. As exhibited in the current example, uneven regions of localised osteosclerosis produce the traditional "cotton wool" appearance.[8]

Loss of lamina dura, hypercementosis, resorption, and replacement of roots may all be visible on radiographs of the jaw bones and teeth. The trabecula can appear like cotton wool in severe cases[6], like present case.

Other well-known signs are the "picture frame," "double contour," or "windowed" vertebral body, the "blade of grass" or "flame sign" in the long bones, and the enlarged pelvic brim known as the "Brim sign.". The most frequent side effect of PDB is pathologic fracture, which can be disabling due to the high rate of non-union in affected bone.[10,14]

Different biochemical indicators of bone remodelling that are elevated in PDB are helpful in the disease's diagnosis. The increasing indicators of accelerated bone resorption are serum deoxypyridinoline crosslinks of type I collagen, serum N telopeptide of type I collagen, serum C telopeptide of type I collagen, and urinary hydroxyproline. Elevated markers of bone production include osteocalcin, serum N-terminal propeptide of type I collagen, serum bone-specific alkaline phosphatase, and SAP.[12,17]

A benign fibro-osseous pattern is often seen under the microscope. Large multinucleated osteoclasts are seen in disorganised areas of resorption during the early stages of osteolysis. Osteoblastic cells phase, the creation of braided bone and randomly constructed new bone matrix are visible. The stroma is dotted with trabecula of braided bone with irregular shapes. Osteoblasts and osteoclasts are frequently found lining the bone, indicating simultaneous bone production and resorption. Small, irregularly shaped bone pieces with deeply stained hematoxyphilic (basophilic) reversal lines appear to be connected together in a mosaic or jigsaw pattern as a result of several events of bone removal and creation.[14,15]

It has been attempted to treat people with symptomatic PDB using a variety of drugs. Calcitonin has been utilised in the past because it prevents bone resorption and offers quick pain relief. Unpleasant Flushing, nausea, and vomiting were frequent side effects of calcitonin therapy. The second-generation nitrogen-containing bisphosphonates (disodium pamidronate, alendronate, risedronate), which are strong inhibitors of bone resorption and offer prolonged remission of the disease in addition to a more drastic reduction in the parameters of bone turnover than calcitonin, are currently the mainstay of treatment for PDB.[17] Following such therapy, the new bone formation has a more typical, lamellar pattern, and anomalies in mineralization are infrequent to nonexistent, with the newer chemicals inducing a more aggressive management concept in which asymptomatic disease in areas with a risk of development and subsequent consequences is considered as a clear indication for treatment. The cytotoxic antibiotic mithramycin, which has been used to treat Paget disease when other therapeutic options have failed, is another treatment option.[1,6,13]

IV. CONCLUSION

Diagnosis of Paget disease, a bone disease that can initially be asymptomatic and is detectable by its characteristic radiological findings, can help avert possible complications. consequences such pathological fractures, arthritis, hearing loss, further neurological issues, heart failure, and, very infrequently, osteosarcoma. Numerous strong bisphosphonate compounds are now available as a result of therapy advancements, and the majority of patients experience normal or almost normal bone turnover indices.

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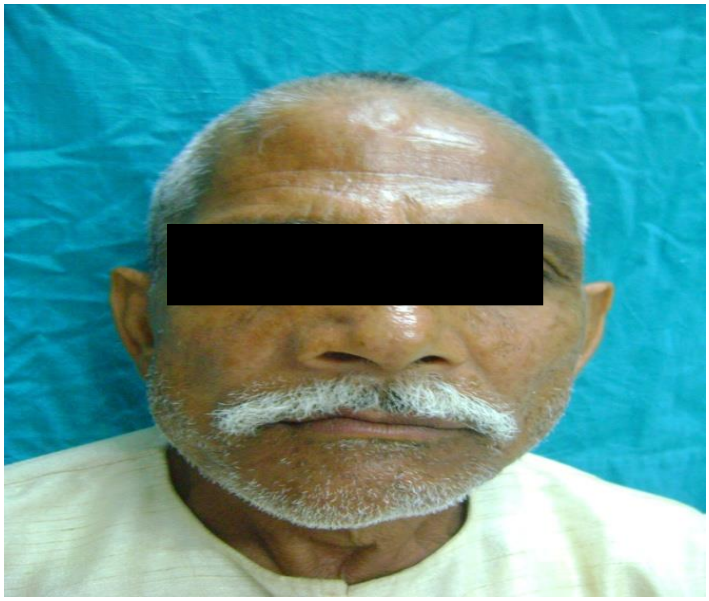

Fig. 1: Extraoral profile of patient

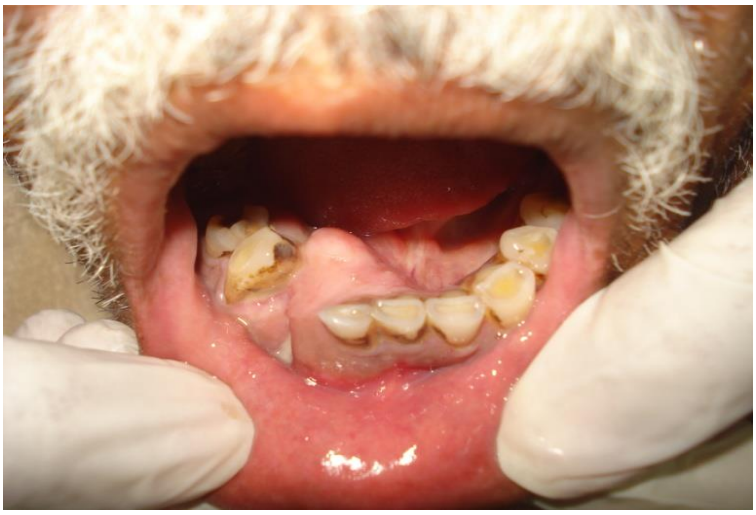

Fig. 2: Intra oral view showing swelling in right mandibular front region

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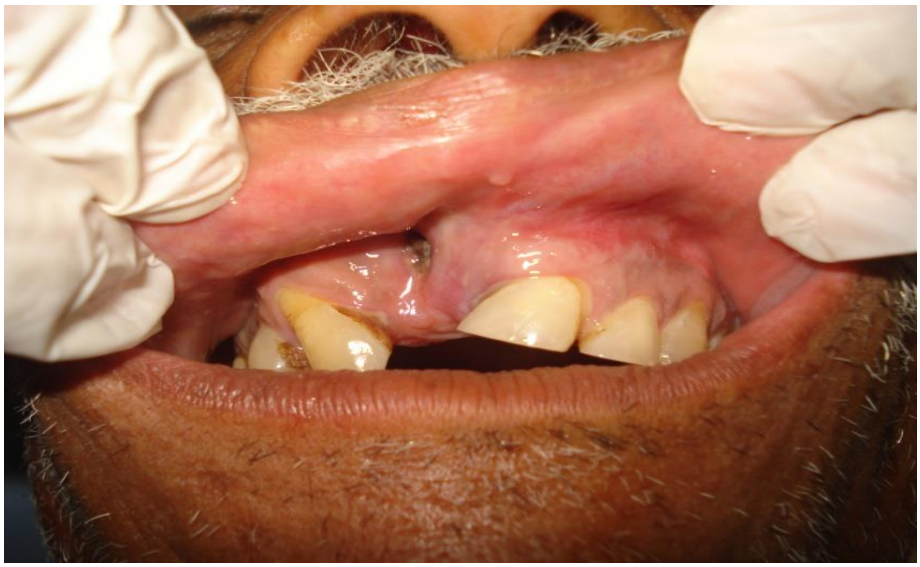

Fig. 3: Intra oral view showing swelling in right maxillary front region

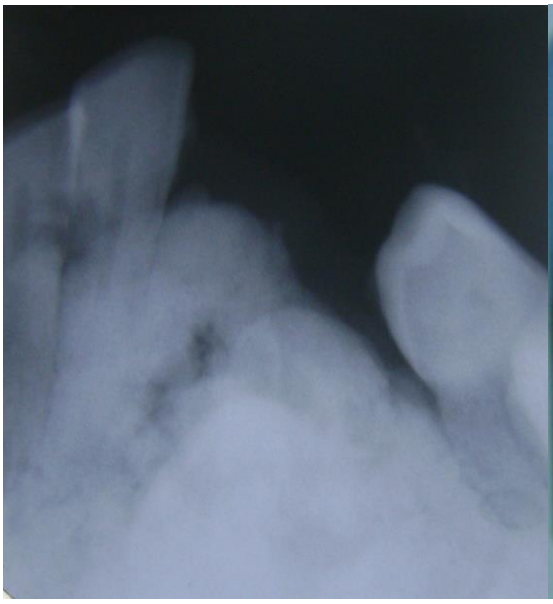

Fig. 4: IOPA showing missing 11 12 with radiopaque masses

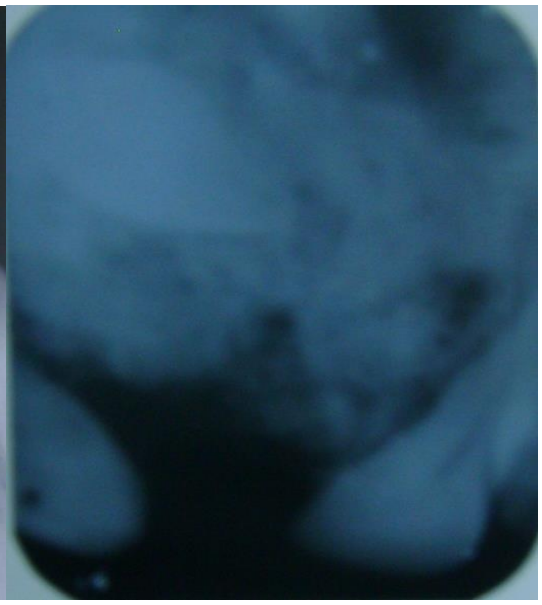

Fig. 5: IOPA showing missing 42 43 with radiopaque masses

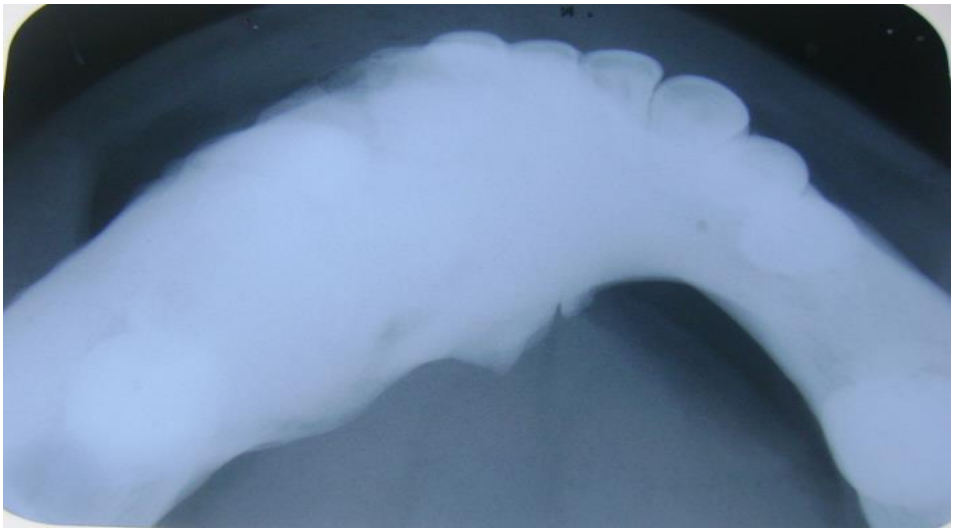

Fig. 6: Mandibular occlusal topographic view with missing 42 43 showing buccolingual cortical plate expansion

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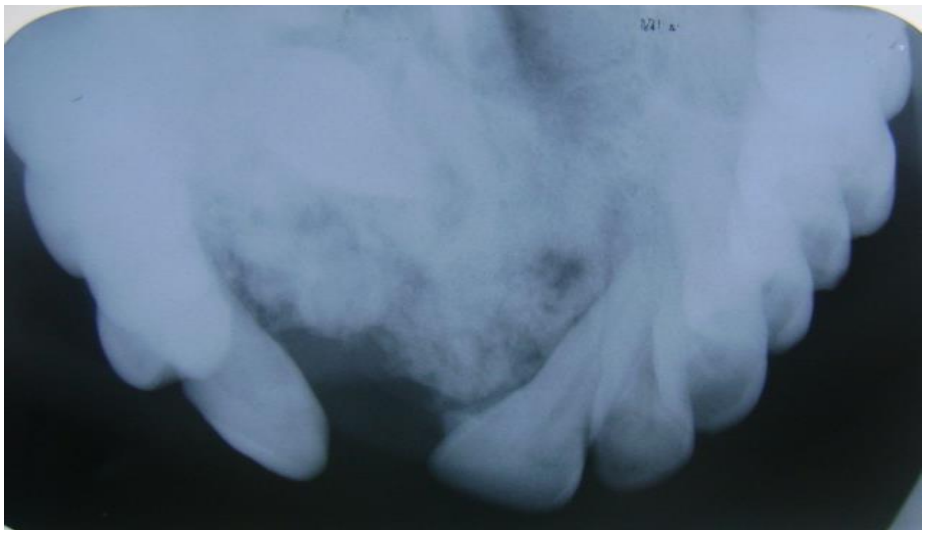

Fig. 7 Maxillary Occlusal topographic view with 11 12 missing showing radiopacities

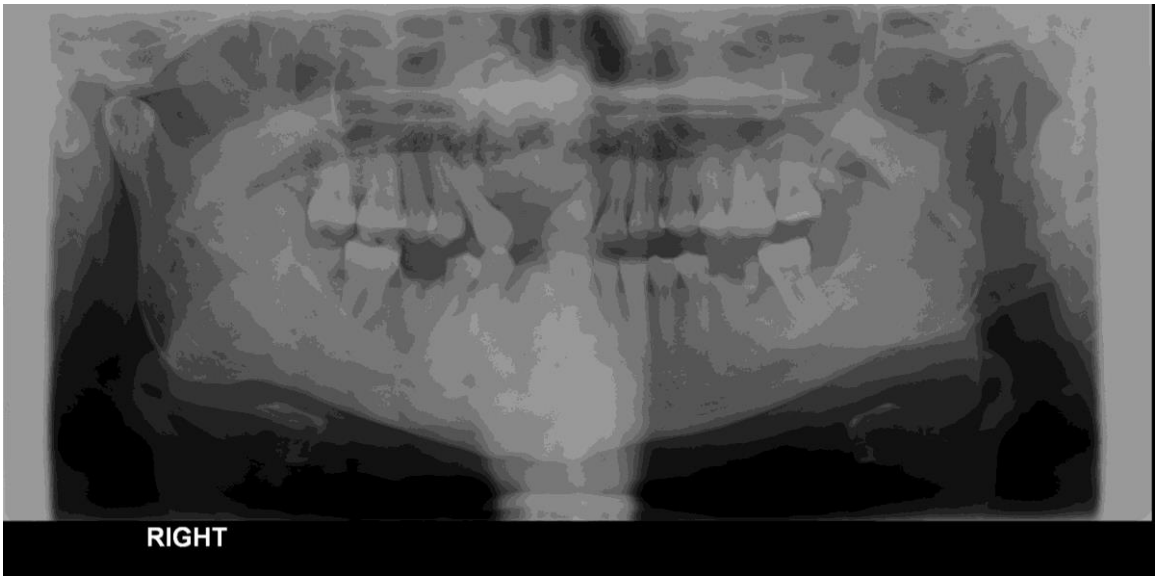

Fig. 8: OPG showing radiopaque masses in anterior right region of mandible and maxilla

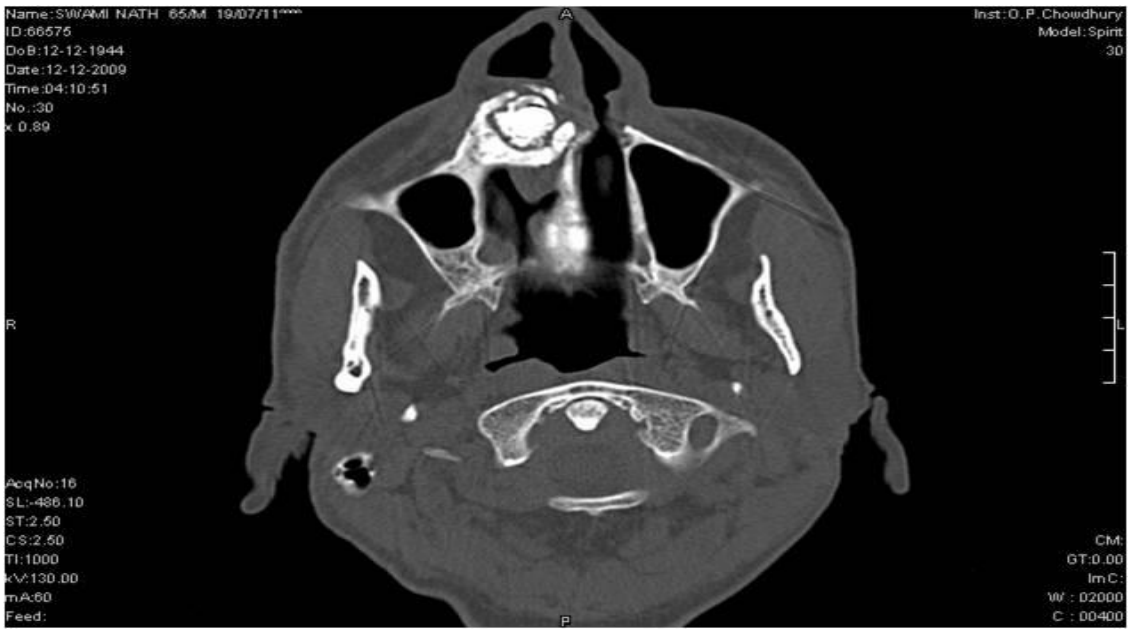

Fig. 9: CT shows hyperdense masses in right anterior region of maxilla

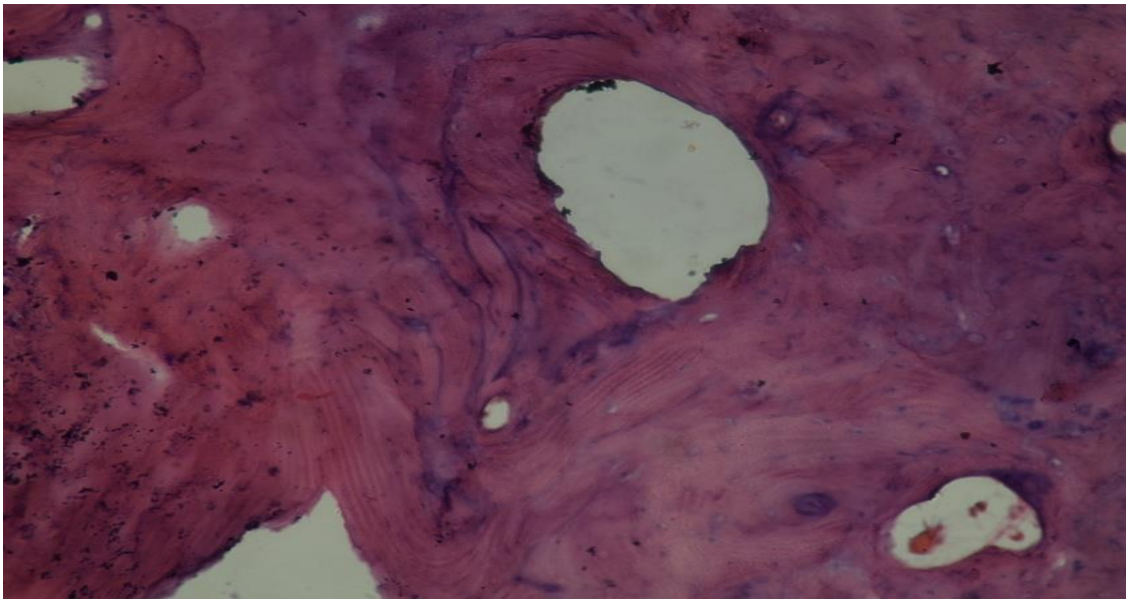

Fig. 10: Photomicrograph shows multiple criss crossing resting and reversal lines