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Gardner's Syndrome: A Report of Two Cases

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Abstract: *Gardner's syndrome is a rare autosomal dominant disease characterized by the presence of colonic polyposis, osteomas and a multitude of soft tissue tumors.The syndrome associated intestinal polyps have a 100% risk of malignant transformation. Early identification and surgical intervention when indicated are important to prolong the life of the patient. The presence of multiple osteomas, multiple unerupted teeth, can give an alarming sign to dentist to screen for other associated features in asymptomatic patients. We report two such cases to emphasize the importance of an early diagnosis of Gardner's syndrome through the detection of lesions appearing in the oral and maxillofacial area.*

Key words: *Gardner's syndrome, Intestinal polyposis, Osteomas.*

I. Introduction

Oral and maxillofacial anomalies are commonly observed in some systemic diseases and dentists should be aware that they may play an important role in the early diagnosis of these diseases even before other systemic features have appeared. In 1951, Gardner described a syndrome consisting of hereditary intestinal polyposis with osteomas and multiple cutaneous and subcutaneous lesions.[1]Its clinical presentation is variable and often imposes a diagnostic challenge to dental practitioners, despite the presence of clues for a significant amount of time. Osteomas usually precede the intestinal polyposis by often as much as 10 years so their detection may lead to appropriate further investigations for the early diagnosis of the syndrome and life-saving treatment of the condition.[2] We report two such cases with objective to highlight the importance of an early diagnosis of Gardner's syndrome(GS), since these settle in the oral and maxillofacial area and also, how important it is for general dental practitioners to be aware of the clinical and radiological characteristics of Gardner's syndrome as oral signs usually precede the other findings of Gardner's syndrome.
Here, we present the oral and maxillofacial considerations of this disease.

II. Case Report-1

A 45 year-old male presented to the department of Oral Medicine and Radiology, Sharad Pawar Dental College, Sawangi with dental deficiencies and chewing concern. History revealed painless swellings on back noted since adolescence and gradually increasing in size. There were no gastrointestinal symptoms. There was strong family history, his mother and brother had similar type of swellings on all over body, and both died at the age of 45-50 yrs due to some gastrointestinal related problems.

Examination revealed well-defined swellings of moderate size, non tender, soft in consistency and fluctuant with presence of punctum on it suggesting epidermoid cyst on back & spine region (Fig.1). Two other small swellings soft in consistency, fluctuant were noted on forehead region.

No gross deformity was observed on extraoral examination. On intraoral examination, a large number of permanent tooth eruptions had not been completed in the posterior region. Missing 15 25 35 45. The

periodontal status was poor. Pus discharge was present from 24 region. No carious tooth was present adjacent. No anomaly was observed in any intraoral soft tissue.

Due to clinical suspicion of gardener's syndrome additional investigations were done. OPG revealed multiple dense homogeneous radioopacities in whole maxilla & mandible giving a cotton-wool appearance to radiograph. The jaw bones also give diffused radiodensity. The pantomograph also revealed multiple unerupted teeth and compound odontomes (Fig. 2). The PNS & CT also revealed additional bony protruberences in frontal, parietal, and temporal region. (Fig 2,3)

Extraction of 24 along with surrounding tissue was done. The collected specimen on histopathological examination revealed osteoma. After extraction, fistula was created in the region for which obturator was placed. Colonoscopy revealed absence of intestinal polyps. Patient is currently on periodic clinical as well as radiological follow up.

III. Case Report-2

A 25 year old male reported with the chief complaint of multiple swellings over right side of forehead gradually increasing since 10 years. New swellings had then appeared over on medial aspect of right eye and chin region. There was no history of bowel disturbances. Family history of the patient was insignificant. On extraoral examination, face was bilaterally asymmetrical due to multiple swellings present on right side of his forehead over supraorbital rim, right infraorbital rim, medial aspect of right eye and right frontal region of size ranging from 1x1.5 to 4x3 cm, roughly oval in shape, smooth surface, diffuse margins . On palpation, they were non tender, consistency was bony hard, and fixed to underlying structure. (Fig. 4) Prominent antegonial notch was palpable on left side. Cafe-au-lait spots were present over right infraorbital region. Asymmetric right and left halves of the face was seen. There was no regional lymphadenopathy. TMJ movements were deviating the mandible towards left side.

Intraoral examination revealed similar swelling on lower anterior region of jaw of size 2x1 cm, well defined, nontender and bony hard in consistency. (Fig.5)

Orthopantomograph revealed asymmetric right and left halves of the mandible with prominent antegonial notch on left side. Increased length of condylar process on right side and decreased height of ramus of mandible on left side. A well defined radioopacity was seen in the symphysis region of 2x1.5cm, oval in shape.(Fig. 6) Lateral skull view showed bony protrusion on frontal bone. Thickening of anterior wall of frontal sinus. (Fig. 7) Bony expansion of supraorbital region extending from root of nose upto lower 1/3rd of forehead. Increase in width between inner and outer table of frontal bone. PA mandible view showed asymmetric right and left halves causing deviation of mandible towards left side. (Fig.8) Waters view revealed increased superoinferior diameter of orbital cavity on right side. Haziness seen on right frontal sinus and right orbit. CT scan and 3D multiplanar reformatted image revealed bony protruberances on medial rim of right eye, thickening of right infraorabital rim, bony protruberance on symphysis region and frontal bone and prominent antegonial notch on left side. (Fig 9,10) Endoscopy and colonoscopy did not detect any polyps.

Surgical recontouring of multiple bony masses was done on maxillofacial region, Lefort I osteotomy with superior repositioning on right side was done. Condylectomy (1.7mm) on right side and advancement and sliding genioplasty with shaving of lower border of mandible under GA was performed. The postoperative course was uneventful. Histopathological examination confirmed a diagnosis of Osteoma.

Thus, considering all the above findings, the present case was provisionally diagnosed as Multiple osteomas with Hemifacial atrophy on left side. Although intestinal polyps are not detected at present, he is on close periodic follow up.

IV. Discussion

Gardner's syndrome is an autosomal dominant disorder which emerges in relation to the X gene. It is caused by a dominantly inherited mutation in the adenomatous polyposis coli gene localized on chromosome 5. It has an incidence ranging between 1 in 4,000 and 1 in 12,000.[1]The syndrome may present at any age from 2 months to 70 years.[3] In our case, the observed age of the cases was 45 years and 25 years.The syndrome is autosomal dominant inherited, so, diagnosis of this condition also has implications for other family members who need to be screened.

Solitary osteoma of the jaw is a common incidental finding on dental panoramic tomography, if more than three such lesions are found this is highly suggestive of Gardner's syndrome[1]and has been proposed as a screening method for Gardner's syndrome.[3]They are the hallmark of Gardner's syndrome and appear in about half of patients and manifest earlier than polyposis. As in the present cases, multiple osteomas were found in the craniofacial region. Although mandible is the most common location, they may occur in the skull, long bones or paranasal sinus cavities[3,2]. The osteomas may be either exostoses, often referred to as peripheral osteomas, or endostoses, which are detectable only radiographically. The radiographic appearance of either type is a localized radiopaque lesion with a sharp border. In the second case, similar finding of osteoma with radioopaque lesion

and sharp borders was found. Another type of lesion has been described, which appears as a large and diffuse radiopaque cotton wool-like area in either jaw, and is referred to as a widespread radioopaque lesion.[4,5]which was found in the first case in both maxilla as well as mandible.

About seventy percent of GS patients exhibit dental anomalies such as impacted or unerupted teeth, as seen in the first case, congenitally missing teeth, supernumerary teeth, hypercementosis, dentigerous cysts, fused molar roots, long and tapered molar roots, hypodontia, compound odontomes, and multiple caries.[6,5]

Cutaneous findings may include desmoid tumors, epidermoid cysts, and other benign tumors. Multiple Epidermoid cyst was one of the finding in the first case. Desmoid tumors may occur in the skin of the anterior abdominal wall or intra-abdominally and are present in about 3.5-5.7% patients.[3] Other cutaneous lesions that have been described in Gardner syndrome include fibroma, lipoma, leiomyoma, neurofibroma, basal cell carcinoma and pigmented skin lesions[7].

Intestinal polyps that develop in the colon as well as in the stomach and upper intestine, along with multiple osteomas is another feature of Gardner syndrome. The intestinal polyps of the disease typically occur prior to puberty and become generalized in the 20–40 year age group[6], which however was not a finding in either of our cases. The significance of this dominantly inherited condition to the dentist is that the polyps have a 100% risk of undergoing malignant transformation change by the fourth decade.[7]Although these are often subclinical, oral manifestations could be diagnostic.

A dental panoramic tomography is a useful initial investigation as suggested by Payne et al in 2002. In young patients with no palpable exostoses, an orthopantomogram may reveal multiple radiopacities (enostoses) throughout the mandible and the maxilla (Antoniades et al., 1987).

Computer tomography is also an important tool for the diagnosis of this disease, especially as it is able to show wavy cortical thickening on the buccal side of the mandible and gives the location details of osteomas.[5]

Management of Gardner syndrome patient is multidisciplinary. Considering the rate of malignant transformation of intestinal polyps the surgical treatment is opted.[7,8] Treatment options for the cutaneous cysts of Gardner's syndrome are the same as for ordinary cysts. Treatment is indicated for symptomatic cases or for cosmetic reasons.[7] Osteomas must be resected if they interfere with function or for cosmetic reasons.[7,4] Histopathological evaluation of resected tissues following surgical intervention are recommended. Recurrence is rare after surgical intervention.

Impacted tooth should be surgically extracted. After extraction of all impacted teeth conventional partial or total prosthetic rehabilitation is advisable.The patients must be aggressively followed-up, since there is a constant threat to their lives at any age.

V. Conclusion

A general dental practitioner may be the first health care professional to review the dental panoramic tomogram of a patient with Gardner's syndrome. His or her knowledge on the clinical and radiological stigmas of Gardner's syndrome may lead to appropriate further investigation and treatment, which might be life saving not only for the patient involved but also for their family members. Patients with GS present a wide variety of manifestations, and must be diagnosed and treated as early as possible.

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None

Conflicts Of Interest:
None declared

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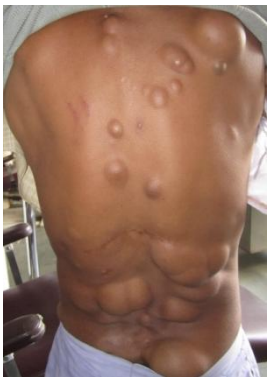


Fig. 1: Multiple epidermoid cyst on back & spine region

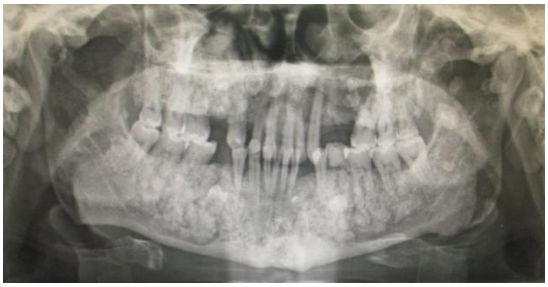
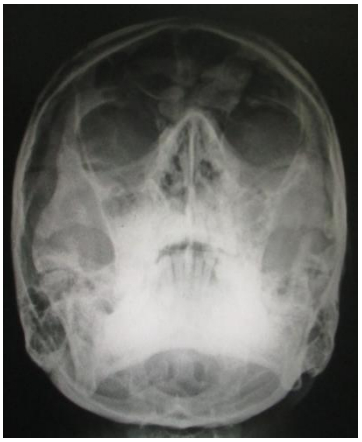


Fig. 2: OPG and PNS showing multiple dense homogeneous radioopacities in whole maxilla & mandible giving a cotton-wool appearance and multiple unerupted teeth



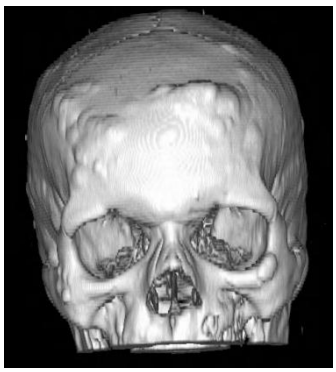


Fig. 3: 3D CT reformatted view showing Bony protruberences in frontal, parietal, and temporal region

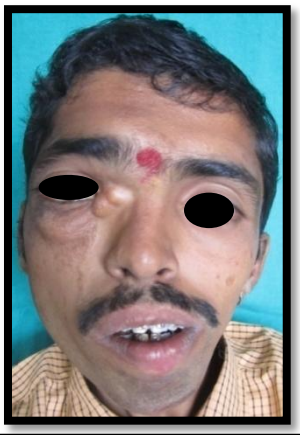


Fig. 4: Bony protruberences in frontal, parietal, and temporal region

Fig 5: Bony hard swelling on lower anterior region of jaw

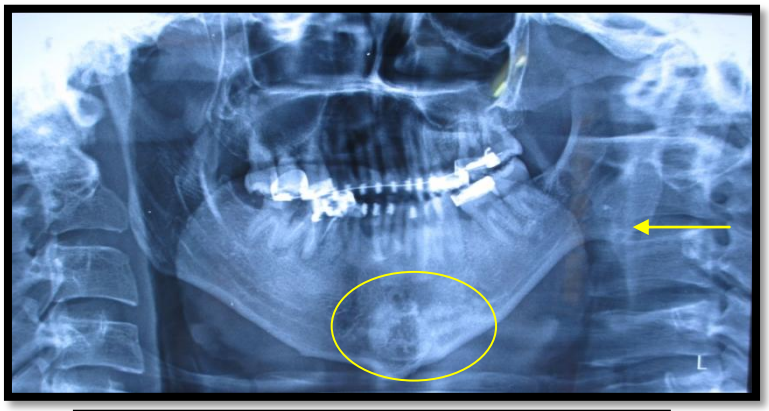


Fig 6: Well defined radioopacity was seen in the symphysis region, Prominent antegional notch on left side

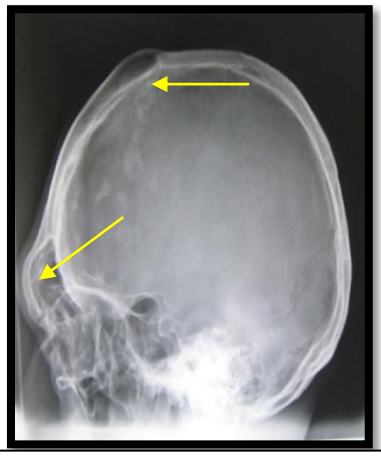


Fig 7: Bony protrusion on frontal bone. Thickening of anterior wall of frontal sinus.

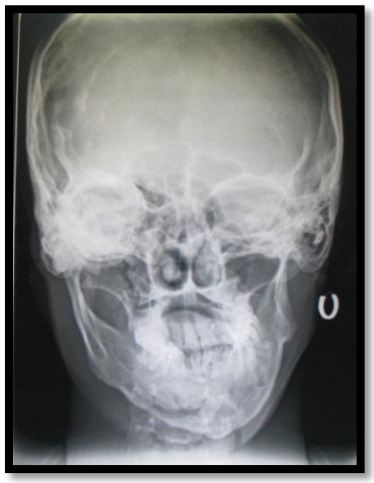


Fig 8: PA mandible showing Deviation of mandible towards left side.

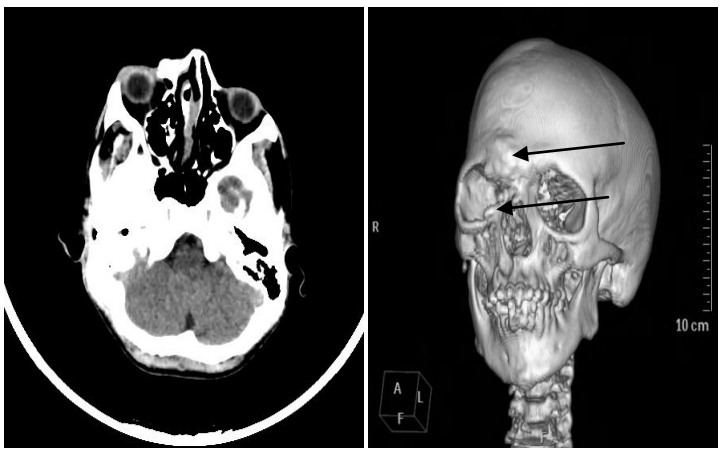


Fig 9,10: Axial CT scan and 3D CT reformatted view shows bony protruberances on medial rim of right eye, and on frontal and temporal bone.