Multiple odontoma in Gardner syndrome: unusual report

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Abstract

Gardner’s syndrome (GS) is a genetic disorder characterised by intestinal polyps, multiple osteomas, and soft-tissue tumours. Dentists play an important role in the syndrome diagnosis considering that craniofacial osteomas are a major criterion for Gardner’s syndrome diagnosis. This case report is of a young individual with Gardner’s syndrome and presence of 28 odontomas in the mandibular posterior region which was surgically excised under general anesthesia.

Keywords: Gartners syndrome, odontoma, osteomas, radiopacities, mandible

Introduction

Gardner’s syndrome (GS) is an autosomal dominant disorder localized to a small region on the long arm of chromosome 5 (5q21-22) [1, 2, 3]. Menzel first described adenomatosis of the colon in 1721, and in 1863, Cripps discovered the heredity of colon polyposis and termed it familial adenomatosis [4]. Devic and Bussy in 1912 described a triad of intestinal polyps, soft tissue tumors, and multiple osteomas of the skull [5]. In 1943 Fitzgerald described a case of intraosseous osteomas, abdominal desmoids, and multiple colon polyposis and defined the oral and facial aspects of the disease [6, 7]. In the early 1950s, the syn-drome was defined by Gardner. In 1962, he also dis-covered the dental abnormalities and skeletal altera-tions of these patients [8]. Gardner syndrome is considered a phenotypic variant of familial adenomatous polyposis [9, 10]. Different mutations in the adenomatous polyposis coli gene have been shown to be associated with Gardner syndrome [11, 12]. Gardner syndrome includes intestinal polyposis, 1 multiple epidermoid cysts, [13] osteomas, [14] dental anomalies, [15] and congenital hypertrophy of the retinal pigment epithelium. 2 Chimenos- Küstner et al. [15] reported that approximately 50% of these patients presented with 3 or more osteomas in the maxillae as well as in other locations. Ida et al. [16] reported that Gardner syndrome should be considered if more than 3 osteomas are present. However, Herrmann et al. [9] reported that only the presence of intestinal polyposis was a marker of Gardner syndrome. The intestinal poly poses have a 100% potential for malignant change [17, 18], which usually occurs in the third to fifth decades of life [19, 20]. Oral and facial anomalies are commonly observed in patients affected with this disease complex, and intestinal polyps predominantly cause malignancy. Dentists should be aware that oral and facial anomalies may play an important role in the early diagnosis of GS. This article presents a case of GS and multiple odontomas in mandible posterior region.

Case Report

A 29-year old patient reported with missing teeth with the mandibular left posterior region of the jaw. History elicited that he never had any permanent tooth on the same region and was unknown about his deciduous scenario. On clinical examination there was missing premolar and molars on the defined region. The area was also confined with a mild vestibular swelling of the buccal side but there was no signs of any sinus discharge, infection or paresthesia to the involved site. On palpation the area was non-tender. Extra oral and dental structures have no abnormality due to supra eruption of maxillary posterior left teeth due to missing contra laterals. An orthopantomogram was advised which revealed multiple radiopaque structures on the parasympysis, body and angle region of mandible on right side.
Due to complexity of area higher diagnostic imaging was required. A three-dimensional computed tomography including axial, coronal and sagittal section were investigated which elicited a group of multiple radiopaque lesion with no perineural involvement suggesting odontomas. (Fig. 1) Excision was planned under local anesthesia. Vestibular incision was given extending from midline towards the left side extending into external oblique ridge of the left side a relieving incision on the midline. Lesion was exposed and with the help of micromotor handpiece and bur, the radiopaque structures were identified and excised. A total 28 odontomas were removed and area was thoroughly inspected and was cleared with betadine irrigation. Surgical closure was done. (Fig. 2) The patient was on regular follow up with no presenting complains and is planned for implants and prosthesis for oral rehabilitation after 6 months of follow up as per the healing scenario.

**Fig 1:** Investigations- A: OPG, B&C: IAN Canal, D: CT Axial Sections, E: CT Sagittal Sections, F: Coronal Sections, G: 3DCT Imaging

**Fig 2:** A: Excised Lesions, B: Exposure of Lesions, C: Area of Lesions, After Excision, D: Post-Operative Surgical Closure

**Conclusion**

Multiple oral osteomas can precede the clinical and radiographic evidence of colonic polyposis or Gardner’s syndrome, therefore, they can be considered sensitive markers for the disease. The oral manifestations may be used by both the dentist and the gastroenterologist to help clinically identify the syndrome at an early age. Patients with oral odontoma should be further examined for the possibility of Gardner’s syndrome. A dental patient with a suspected diagnosis of Gardner’s syndrome should be referred to a gastroenterologist and a colonoscopy should be performed. The patient should then schedule a follow up dental appointment, as described in this case.

**References**