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CASE REPORT

Gardner syndrome—review and report of a case

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KEYWORDS

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Summary Gardner syndrome represents one end of the spectrum of the disorder known as familial adenomatous polyposis (FAP). The syndrome consists of intestinal polyps which are predominantly adenomas, together with extracolonic features including osteomas, epidermoid cysts, desmoid tumours and dental abnormalities. Desmoid tumours, although histologically benign, may behave aggressively and can cause death in some patients. Several other neoplasms are also associated with Gardner syndrome.

The inheritance pattern is autosomal dominant, with a locus on chromosome 5. However, 20% of cases may represent new mutations.

The intestinal polyps carry a 100% risk of malignant change, and early diagnosis is therefore essential. As the extracolonic manifestations of the disorder often pre-cede the colonic polyps, they may facilitate early identification of affected patients and allow timely intervention. Several of these manifestations occur in the oral and maxillofacial region and may be discovered during routine dental examination.

We report a case of a 37-year-old patient with Gardner syndrome, who has previously undergone a pancolectomy with ileo-rectal anastomosis. Extracolonic features include a recurrent abdominal desmoid tumour, multiple epidermoid cysts, osteomas of the mandible and skull, and dental abnormalities.

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Introduction

In 1953, Gardner described a syndrome consisting of hereditary intestinal polyposis with osteomas and multiple cutaneous and subcutaneous lesions.¹

This syndrome has since been modified by the addition of other features such as dental abnormalities, abdominal desmoid tumours and a number of malignant tumours.² At one time, Gardner syndrome was thought to be a separate entity from familial adenomatous polyposis (FAP). However, it is now accepted that it represents one end of the spectrum of that disorder, and around 50% of those with FAP have extracolonic manifestations, although these are often subclinical. The frequency

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How rare is gardner's syndrome. How is gardner's syndrome diagnosed. What age is gardner's syndrome diagnosed. What causes gardner's syndrome. Gardner syndrome case report.

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Flores MJ, Cisneros IJC, Vides IJA, Hernández PMS. Osteoma periférico de la mandíbula y dentición decidual. ¿Un preludio del síndrome de Gardner? An Orl Mex. 2014; 59 (1): 79-83. John E. Khachemoune A. Gardner syndrome. Skin manifestations, differential diagnosis and management. Am J Clin Dermatol. 2010; 11 (2): 117-122. Sriharibabu M, Sarma Y, Bhaskararao G, Nayak S, Satyaprakash T. Gardner's syndrome presenting a duodenal carcinoma in a young male. J Clin Sci Res. 2015; 4 (4): 296-300. Sapp JP. Patología oral y maxilofacial contemporánea. 2 ed. Barcelona, España: Elsevier; 2004. p. 326. View PDFVolume 41, Issue 5, May 2005, Pages 89-92 rights and contentFamilial adenomatous polyposis Gardner's syndrome: A case report General dental practitioners' awareness of the clinical and radiological characteristics of Gardner's syndrome is crucial for early detection. Early detection of polyps associated to Gardner's syndrome could be life saving. Gardner's syndrome is characterised by polyposis coli associated with multiple hard and soft tissue tumours including osteoma and odontoma.1 The major significance of the condition lies in the gastrointestinal polyposis which usually undergoes malignant change by the fourth decade. Extra-intestinal components may be apparent before those in the bowel, and their early detection may lead to appropriate evaluation and life saving treatment. A case is reported to demonstrate how important it is for general dental practitioners to be aware of the clinical and radiological characteristics of Gardner's syndrome. A 19-year-old army recruit was admitted to the hospital for his first periodical examination. On examination, a painless bony hard swelling was palpable at the right mandibular corpus, extending posteriorly to the right mandibular angle. When asked, the patient confirmed the presence of that swelling for a period of more than five years. There were no obvious soft tissue lesions and the physical examination failed to reveal any other pathological findings. A panoramic dental tomogram revealed a sharply defined, large radiopaque lesion consisting of several clumped toothlets, exhibiting a thin radiolucent rim on the periphery (Fig. 1). Computed tomogram, acquired on axial and coronal planes, showed a dens lobulated bony lesion on the right mandibular corpus extending both into the intra and extraoral soft tissue planes, consistent with compound odontoma. When reviewing the coronal CT images, a round shaped osteoma, arising from the medial wall of the left bony orbit and protruding into the frontal sinus, was discovered (Fig. 2). With a provisional diagnosis of Gardner's syndrome in mind, a CT colonoscopy was performed. Two polyps were detected in the colon measuring 2 cm in diameter, which were associated with a high risk of malignant transformation (Fig. 3). Colonoscopy and the following histological examination confirmed the presence of colonic adenomatous polyps with no malignant change. The patient was registered for colectomy and his family members advised to undergo colonoscopic examination.Figure 1: Dental panoramic tomogram shows a sharply defined, large radiopaque lesion consisting of several clumped toothlets on the right mandibular corpus.Figure 2CT image in the coronal plane shows a round shaped osteoma, arising from the medial wall of the left bony orbit and protruding into the frontal sinusFigure 3CT image in the axial plane acquired following rectal air insufflation and intravenous contrast injection shows a polyp (white arrow) measuring 2 cm in the sigmoid colon.Familial adenomatous polyposis coli has a prevalence of about 1 in 8,000 and is the most common form of the hereditary polyposis syndromes.2 It is inherited in an autosomal dominant pattern and linked genetically to chromosome band 5q21.3 Gardner's syndrome is believed to be a variant of familial adenomatous polyposis, but has a wider spectrum of abnormalities including gastrointestinal polyps that develop in the colon as well as in the stomach and upper intestine, sebaceous cyst, and multiple hard and soft tissue tumours including osteomas and odontomas.1Osteomas are the hallmark of Gardner's syndrome. Although the mandible is the most common location, they may occur in the skull, long bones or paranasal sinus cavities as in the above presented case. Radiographic features of an osteoma are generally characterised by a round or oval radiopaque mass attached by a broad base.4 Computed tomography might be used to precisely delineate the size of the tumour as well as its relation to any important adjacent structure.Odontoma is an odontogenic tumour which is also associated to Gardner's syndrome. Odontomas are divided into two basic types: compound or complex.5 Complex odontomas are seen as a solitary radiodens lesion, accompanied by a radiolucent halo, and can easily be misinterpreted as a mature fibro-osseous lesion.6 Therefore complex odontomas cannot be diagnosed by radiographic appearance alone and require histological sampling. On the other hand, the diagnosis of compound odontomas is usually determined by radiographic appearance alone and is seldom confused with any other lesion.6 As illustrated in the above presented case, they appear as a radiopaque lesion containing small, multiple, immature or rudimentary teeth surrounded by a thin radiolucent layer. In contrast to compound odontomas, complex odontomas appear radiographically as more or less amorphous, solitary conglomerates of indistinct radiopacities rather than toothlike structures. Treatment of bone lesions encountered in Gardner's syndrome, including osteomas and odontomas, depends on the symptomatic or cosmetic nature of the findings. For a case like the one presented above, an elective cosmetic surgical approach should be considered for the mandibular odontoma, whereas no treatment is found to be necessary for the osteoma.In Gardner's syndrome polyps may be found in any part of the gastrointestinal tract, including the stomach and the duodenum. The mean age of symptomatic presentation is 39.2 and they have a 100% risk of undergoing malignant transformation.3 In fact polyps begin to form in puberty, but the average age of diagnosis is 22 years. Formation of osseous lesions precede that of polyposis.4 Therefore radiographic recognition of osseous lesions in Gardner's syndrome can lead to early detection of polyps before they undergo any malignant differentiation. The incidental odontoma encountered in the dental panoramic tomogram, followed by an osteoma in the above presented case, raised suspicion for a presumptive diagnosis of Gardner's syndrome. Colonoscopic biopsy, performed from polyps found on a subsequent CT colonoscopy, confirmed the diagnosis and the patient underwent a life saving surgical treatment before any malignant transformation had occurred.7 Knowing that Gardner's syndrome is inherited in an autosomal dominant fashion, the patient's family members should also be screened for polyps. But it must also be kept in mind that as many as 30% of individuals with Gardner's syndrome are reported to have a new dominant mutation.8 Therefore a prior genetic mutation test can be useful to identify affected members of the family to be screened.In 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. Gardner EJ, Richards RC . Multiple cutaneous and subcutaneous lesions occurring simultaneously with hereditary polyposis and osteomatosis. Am J Hum Genet 1953; 5: 139-147.PubMed PubMed Central Google Scholar Payne M, Anderson JA, Cook J . Gardner's syndrome — a case report. Br Dent J 2002; 193: 383-384.Article Google Scholar Luk GD . 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