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The Clinical Features and Their Impact on the Prosthodontic Management In a Case of Gardner's Syndrome

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Abstract - *Gardner's syndrome is a variant of Familial Adenomatous Polyposis (FAP), a condition that manifests as hundreds of colorectal polyps likely to undergo malignant change by the fourth decade. Early diagnosis of this condition has the potential to be life saving for individuals and due to its inherited nature other family members can often also be affected. Additional features of Gardner's Syndrome include multiple jaw osteomas with missing teeth that can make prosthodontic treatment a challenge. This case report highlights the presenting features and the prosthodontic problems faced when treating a patient with Gardner's syndrome.*

KEY WORDS: Gardner's syndrome, jaw osteomas, prosthodontic management

INTRODUCTION

Gardner's syndrome, also known as familial colorectal polyposis, has a reported incidence of between 1 in 1,400 and 1 in 12,000 with equal gender distribution and is one of the rarest syndromes seen in dentistry[1]. The syndrome was first distinguished as its own variant in 1972 by Gardner who related the polyposis with extra colonic features and a hereditary pattern[2]. It is thought to affect 10% of Familial Adenomatous Polyposis (FAP) patients and is a genetically autosomal dominant condition with poor penetrance but can be considered as phenotypic variant of FAP [3,4]. A mutation of the tumour suppressor APC gene on band q21on chromosome 5 causes inactivation of the gene and allows proliferation of colonic polyposis[5].

Colorectal polyps first start to appear around adolescence and will progress to hundreds throughout the gastrointestinal tract. Without treatment there is an almost 100% progression to colorectal cancer. Extracolonic presentations include osteomas of the skull, osteomata, epidermal and sebaceous cysts, desmoid soft tissue tumours in approximately 15% of affected individuals (Figure 1) and dental anomalies. Thyroid, adrenal and biliary carcinomas may also develop[6]. Weight loss and abdominal pain can occur and misdiagnosis includes Irritable Bowel Syndrome, Coeliac and Crohn's disease. Treatment can include elective colectomy or proctocolectomy. Genetic screening is given to family members and those with the mutation undergo annual endoscopy of the colon[7].

Congenital hypertrophy of the retinal pigment epithelium (CHRPE) has been linked with Gardner's syndrome. It is usually diagnosed before the age of 30 years, is asymptomatic and does not tend to undergo malignant transformation[6].

Osteomas present as bony hard swellings which can be seen clinically (Figure 2) or discovered incidentally on panoramic radiographs. They consist of compact bone, cancellous bone or a combination of both. Usually they are painless, unless they enlarge to a size that predisposes to trauma. Commonly they present in the mandible, although can also be seen in the maxilla, frontal bone and other bones of the skull (Figure 3). Other dental anomalies include multiple unerupted supernumerary and permanent teeth, cemental lesions and odontomes[6]. Jaw osteomas lend a "cotton-wool" appearance to radiographs of the jaws and these may lack definition as a result [8].

This report describes the prosthodontic challenge arising from multiple jaw osteomas in a case of Gardner's syndrome. In this case the patient presented with failing crown and bridgework, multiple missing units and large osteomas in both the maxillary and mandibular alveoli.

CASE REPORT

Initial Presenting Features

A 79 year old male who had been previously diagnosed with Gardner's syndrome, presented at the restorative department at Liverpool University Dental Hospital. His main complaint included difficulty eating and concerns regarding his dental appearance, particularly spaces, although he was free from dental pain. He had never worn dentures previously.

The patient had been diagnosed with Gardner's syndrome from a young age and had many of the extra-colonic features including: multiple jaw osteomas in the mandible, maxilla and skull bones (Figures 1 & 2); desmoid tumours; and epidermoid cysts affecting the right eye and nose (Figure 3). In addition, he had impaired hearing in his left ear and suffered with angina. The medication list included Aspirin, GTN spray, Nicorandil and a statin.

Radiographic interpretation proved slightly difficult as the dense bone around the teeth prevented clear visualisation of the apices.

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Figure 1. *Desmoid tumour of the abdomen.*

Figure 2. *Osteoma of the occipital bone.*

Figure 3. *Epidermoid cysts*

Figure 4. *Osteoma and gingival hyperplasia around the LR7.*

Figure 5. *Mulitple osteomas of the mandible and maxilla.*

Figure 6. *Osteoma of the maxilla.*

Periodontal support was remarkably good with bone levels indicating a degree of periodontal resistance as no sextant had a BPE code greater than 3. The deeper probing depths were recorded around the lone standing 47 and these were false pockets rather than loss of clinical attachment. The bony exostoses and overgrowth of the right mandible posed difficulties in the maintenance of oral hygiene and he suffered from chronic gingivitis. The 47 appeared to be submerging. Gingival hyperplasia added to the difficulties particularly in the lower right quadrant where the one molar remained (Figure 4).

Occlusal examination revealed no stable or obvious intercuspal position (centric occlusion) and posterior open bites. The maxillary central incisors appeared to have overerupted within the boney exostosis, similar to dento-alveolar compensation resulting in premature contacts anteriorly and no occlusal contact posteriorly. The significantly deranged occlusion was a probable consequence of the alveolar bone overgrowth seen in Gardner's syndrome but this has not been previously described in the literature. (Figures 5 & 6).

The upper central incisors had labial porcelain veneers with recurrent caries on the palatal margins. A straight cantilever bridge from the 32 replaced the 31. Of the remaining lower teeth the 43 and 33, 34, 35 had porcelain jacket crowns, and the 47 was submerged in bone. The remaining upper

teeth 14, 11 and 21, 24 were heavily restored with amalgam, composite and glass ionomer and 14 failed to respond to pulp sensibility testing.

Management Considerations

A treatment plan was devised to improve plaque control, stabilise the caries and resolve the gingival inflammation. A targeted programme of non-surgical periodontal treatment with the hygienist was instigated. The false pockets around the 47 were debrided and the patient instructed in the home use of sub-gingival irrigation with Chlorhexidine gluconate. This was followed by gingivectomy and bone removal around the 47 to further aid cleaning.

On completion of non-surgical periodontal treatment and the stabilisation of caries together with endodontic treatment of the 14, future management options were considered. Long term care maintenance planning was discussed at length and the patient appreciated the limitation of restorative treatment with respect to a failing reduced dentition. Acrylic partial dentures were planned as training appliances to aid habituation, with a view to constructing cobalt chrome dentures at a later date should this become appropriate. In this situation where the dentition was failing the patient was made aware that future treatment needed to be simple and flexible.

The task of constructing upper and lower partial acrylic dentures proved challenging. Stock trays for primary impressions had to be greatly modified prior to making custom trays. Due to the complex anatomy that included large bony exostoses and significant undercuts, jaw registration was difficult and close liaison with the dental technicians was needed. The skeletal derangement prevented the establishment of posterior contact between upper and lower registration blocks resulting in a posterior open bite, unless the occlusal plane was significantly lowered. Moreover, the only remaining posterior teeth in the maxilla were the second premolars which were kept to prevent bone resorption but they dictated the position of the occlusal plane, see Figure 7. After discussion with the patient, it was decided to accept a posterior open bite and no posterior inter-digitation of teeth in ICP/CO. The prosthesis was successfully tolerated and the patient was extremely pleased with the improvement in function and appearance (Figure 7) despite the interocclusal gap. Further bone surgery prior to the construction of definitive cobalt chromium partial dentures was offered to improve aesthetics but this was declined. After a suitable period of habituation a lower cobalt chromium partial denture was provided (Figure 8). In order to maximise patient comfort and particularly to reduce pressure on the thin mucosa over the boney prominences a tooth supported prosthesis was provided. A feature was the wide and deep lingual connector which sat on the broad alveolus, which although providing extra support, may have interfered with the tongue. The extension of the lingual flange was deep into the sulcus and was comfortable. Clasping proved difficult as undercuts on the posterior teeth were shallow as a result of the combined gingival and bone overgrowth. Crowns with guide planes and rests were considered but rejected as the prognosis for several teeth was limited and gingivectomies would have been required to expose crowns of teeth.

Figure 7. *Upper and lower acrylic partial dentures*

Figure 8. *Lower Cobalt Chronium Partial Denture*

DISCUSSION

The diagnosis of Gardner's syndrome is usually made after genetic screening when other family members are affected. However new mutations may remain undiagnosed and these sufferers may be unaware that they have the condition. Dentists can help to make a diagnosis by recognising the dental implications and any patient presenting with multiple osteomas with unerupted teeth or odontomes and cementomas should be referred to Gastroenterology. Often osseous features can predate polyposis and therefore early diagnosis and treatment can be life saving[7].

Despite the difficulties in the dental management of patients with Gardner's syndrome it is important patients are regularly seen by their General Dental Practitioner. Malformation of the jaw bones may lead to difficulties in oral hygiene leading to periodontal disease and dental caries. As patients will often have congenitally absent teeth and as denture construction can be problematic, prevention of dental disease is imperative to ensure patients have a functional natural dentition for as long as possible. The boney prominences result in significant undercuts and the path of insertion has to be carefully determined. Thin overlying mucosa may be best left uncovered although in this case the gingival biotype was fairly thick and the keratinized gingiva was covered particulary lingually. Foil relief may be required should mucosa be thin and pressure result in pain. Surgery can be useful to eliminate undercuts from

osteomas, however, as demonstrated in this case not all patients opt for this treatment. Referral to secondary care may not be appropriate in all cases and often patients can be managed within primary care by keeping treatment simple and therefore reducing the difficulties posed in long term care and maintenance.

CONCLUSION

Features of Gardner's syndrome should be recognised clinically and appropriate referral should be made when needed. Dental treatment should be kept simple and thus referral for specialist care is not always indicated.

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Erratum notice

EJPRD June 2012; 20 (2): 86-91

The impact of fractured endodontic file removal on vertical root fracture resistance: three dimensional finite element analysis. Listed authors S A Romeed and S M Dunne.

Author omission, should read S A Romeed, S M Dunne and A A Madarati