Int. J. Oral Maxillofac. Surg. 2024; 53: 219–222
https://doi.org/10.1016/j.ijom.2023.10.032, available online at https://www.sciencedirect.com

International Journal of Oral & Maxillofacial Surgery

Case Report
TMJ Disorders

A role for total alloplastic temporomandibular joint replacement in Gardner syndrome

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D. F. Ângelo, M. Nunes, F. Monje, B. Mota, F. Salvado: A role for total alloplastic temporomandibular joint replacement in Gardner syndrome. Int. J. Oral Maxillofac. Surg. 2024; 53: 219–222. © 2023 Published by Elsevier Inc. on behalf of International Association of Oral and Maxillofacial Surgeons.

Abstract. Gardner syndrome (GS) is a rare autosomal dominant disorder that can present with craniomaxillofacial abnormalities. The identification of osteomas or craniomaxillofacial abnormalities can therefore serve as a marker of this condition, facilitating early referral and diagnosis. A 17-year-old female with GS was referred for the management of severe limited mouth opening, causing a major problem for routine endoscopy to monitor the gastrointestinal alterations of GS. Clinical and radiological evaluations showed multiple osteomas in the mandibular angle, condylar and coronoid regions bilaterally and maximum mouth opening of 8 mm. The patient underwent surgery for osteoma removal and bilateral customized alloplastic total temporomandibular joint replacement (TMJ-TJR). At the 2-year follow-up, the patient showed improvements in quality of life, with a maximum mouth opening of 34 mm, allowing routine upper endoscopy to be performed. This is the first report of GS, a rare and challenging craniomaxillofacial abnormality, treated with TMJ-TJR. A comprehensive overview of the patient's clinical presentation, diagnostic assessment, treatment planning, and outcomes is provided.

Keywords: Gardner syndrome; Temporomandibular joint; Osteoma; Joint prosthesis; Total joint replacement.

Accepted for publication 27 October 2023
Available online 18 November 2023

Gardner syndrome (GS) is a variant of familial adenomatous polyposis (FAP), with classic features including the triad of colonic polyps, osteomas and cutaneous and subcutaneous soft tissue tumours (desmoid tumours, Gardner fibroma), as well as dental abnormalities and non-neoplastic lesions (epidermoid cysts)[1,2]. FAP is a rare genetic disorder with autosomal dominant inheritance, characterized by multiple adenomatous polyps of the colon and rectum, increasing in number and size with increasing age. Studies suggest that individuals with FAP will inevitably show progression to colorectal carcinoma unless the condition is detected and managed early.

Osteomas, odontomas, and abnormal dental findings frequently occur in patients with FAP. Osteomas, usually located in the jaws, are the most common skeletal abnormality associated with GS and are of significant

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relevance from a dental perspective. Frequent dental abnormalities include impacted teeth, supernumerary teeth, odontomas, and congenitally missing teeth. The simultaneous presence of osteoma(s) with dental anomalies is highly suggestive of underlying GS[3,4].

Nearly half of the patients with FAP are reported to have three or more osteomas of the jaws. Osteomas typically increase in size with increasing age. Although the majority are asymptomatic, they can result in aesthetic complaints (facial disfigurement and asymmetry) and restricted mandibular movements, depending on their location within the craniofacial complex. Radiographically, they appear as a well-defined radiopacity. Despite the presence of multiple osteomas in the maxillofacial complex being suggestive of GS and perhaps contributing to early diagnosis, their detection has no prognostic value regarding the development of colonic cancer[2].

The orofacial features of GS often precede intestinal polyposis[2]. Typically, cutaneous and bony lesions appear 10 years prior to intestinal polyposis, although the lesions can present in a different chronological order. As craniofacial osteomas and dental anomalies are major criteria for GS diagnosis, stomatologists and oral and maxillofacial surgeons play an important role in the diagnosis and treatment of the syndrome[3,5]. The identification of osteomas or craniomaxillofacial abnormalities can therefore serve as an early marker of this condition, facilitating early referral, diagnosis, and initiation of life-saving treatment.

Case report

A 17-year-old female was referred to the Department of Oral and Maxillofacial Surgery at Instituto Português da Face in Portugal, for the management of craniomaxillofacial manifestations of GS. Her main complaint was limited mouth opening, causing a major problem by limiting the performance of routine endoscopy to monitor the gastrointestinal alterations of GS.

A detailed history revealed multiple cutaneous and subcutaneous benign tumours and lesions since the age of 3 years, with a diagnosis of FAP when she was 10 years old. The patient had been followed up by her paediatrician, an otolaryngologist, a gastroenterologist, and a dentist every year since the diagnosis. There was no family history of FAP.

The initial clinical examination showed multiple nodular formations that were palpable along the mandibular angle bilaterally. On palpation, these lesions were hard, well limited, immobile, and non-adherent to skin. The oral mucosa was normal, and the regional lymph nodules were not palpable. Physical examination revealed limited mouth opening, with a maximum mouth opening (MMO) of 8 mm.

Neither crepitus nor clicking on mouth opening was noticed in the temporomandibular joint (TMJ) region bilaterally. No deviation of the mandible was observed, and no pain in mouth opening was evident.

A panoramic radiograph showed the presence of multiple round radiopaque lesions in both the maxilla and mandible. Diffuse sclerosis could also be noted throughout the mandibular body.

A computed tomography (CT) scan showed multiple osteomas in the mandibular angle, condylar and coronoid regions bilaterally, limiting function and MMO (Fig. 1).

No sensory or motor dysfunction in the trigeminal or facial nerve distribution was noted.

Due to the functional impairment and resulting difficulty performing routine upper endoscopy, as well as oral hygiene and dental treatment, a treatment plan for the removal of the major osteomas along with TMJ reconstruction was proposed. Because of the severe anatomical changes, the surgical team decided to perform a bilateral customized total alloplastic TMJ replacement (TMJ-TJR) (Stryker/TMJ Concepts, Ventura, CA, USA) with

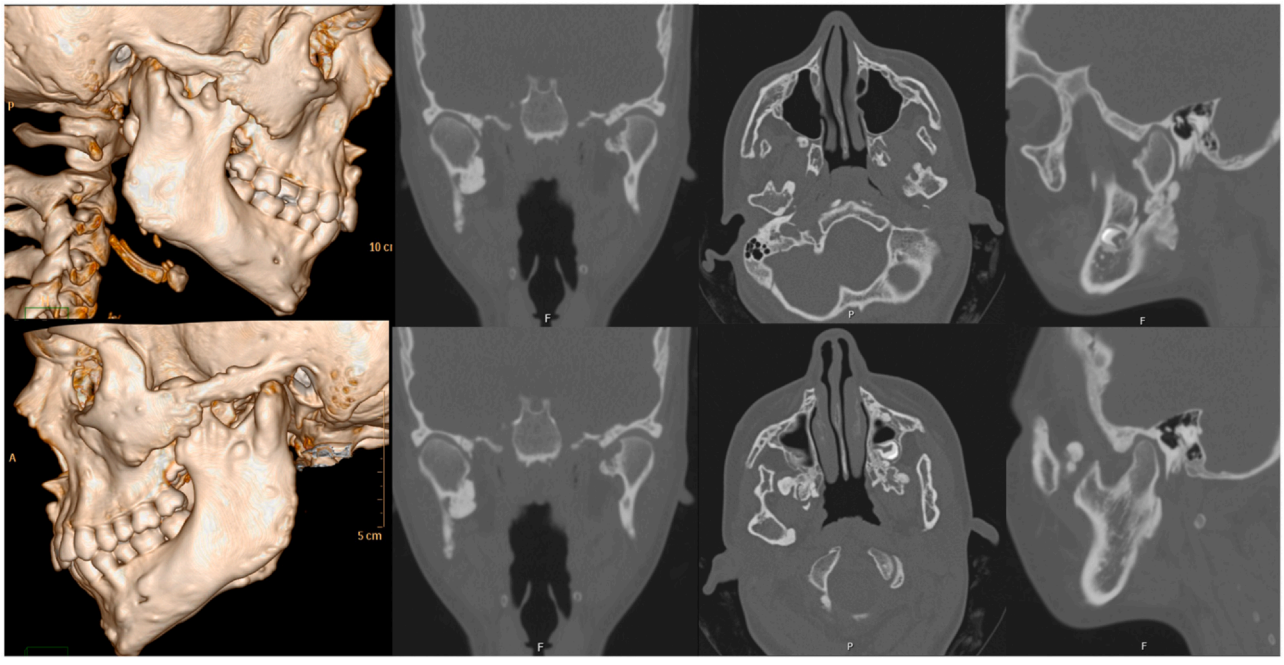

Fig. 1. Preoperative CT scan showing multiple osteomas in the maxillofacial region. The mandibular angle, condylar and coronoid regions bilaterally are the most important landmarks for surgery planning.

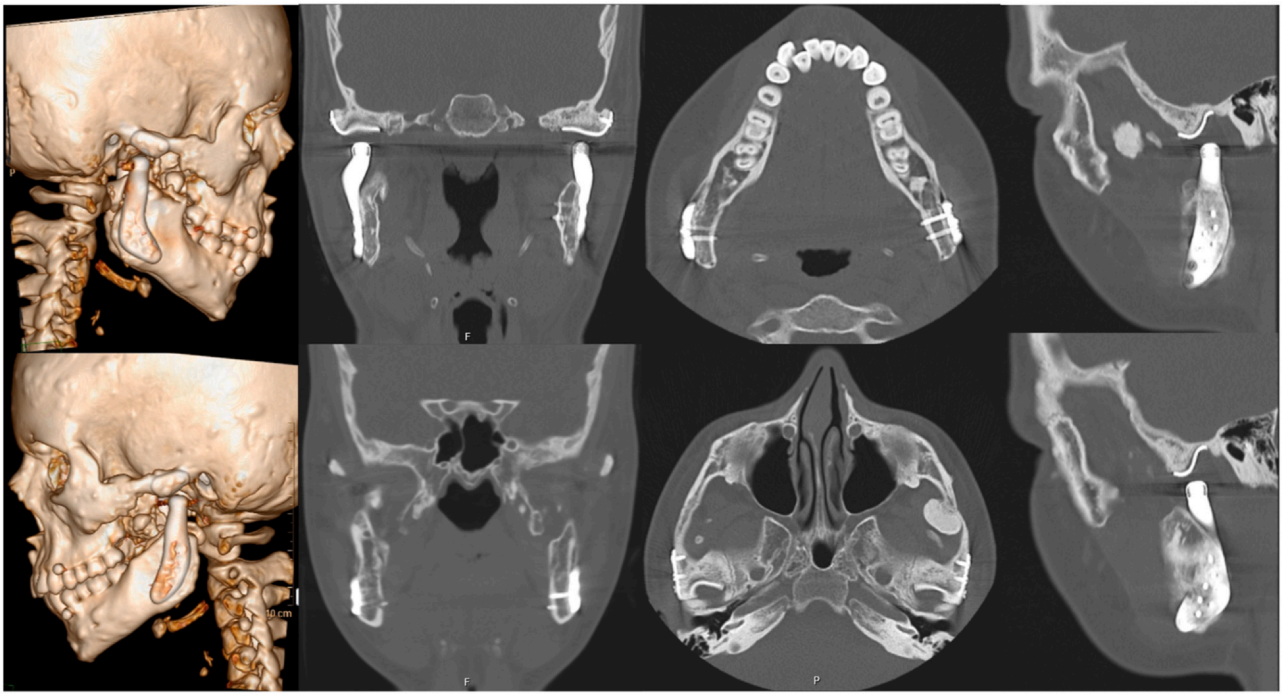

Fig. 2. Postoperative CT scan showing a regular and symmetrical contour of the mandible, and well-adapted bilateral joint prostheses.

coronoidectomy. The smaller osteomas were not removed as they were insignificant to the objective of improving MMO and quality of life. There were no surgical complications and the patient was advised to undergo physiotherapy every week for 2 months and daily TheraBite exercises. The postoperative CT scan showed a regular and symmetrical contour of the mandible and well-adapted bilateral TMJ prostheses (Fig. 2).

Two years after surgery, the patient was satisfied with the improvement in her ability to perform oral hygiene and had no diet restrictions; her MMO was 34 mm. Subsequently, routine endoscopy was performed easily by her gastroenterologist, with the identification of multiple gastric polyps. A routine colonoscopy was also performed, revealing multiple intestinal polyps.

The patient was recommended to undergo an annual radiographic examination of the jaws, as well as upper endoscopy and colonoscopy to monitor the intestinal polyposis closely. No signs of relapse in the jaws were observed at the 2-year follow-up.

Discussion

GS is a rare genetic disorder of significant oral and maxillofacial relevance. It is an inherited autosomal dominant disorder characterized by the development of multiple osteomas, along with other clinical manifestations such as gastrointestinal polyps, cutaneous lesions, and dental abnormalities. As highlighted in this case report, the identification of oral manifestations of GS is essential, leading to confirmation of the diagnosis and the instigation of appropriate treatment. Initial management involves confirmation of the GS diagnosis, and in the majority of cases a strong family history is reported. However, almost one third of cases result from a new mutation within a family member, as highlighted in this case[2].

The clinical features of GS can be divided into colonic and extra-colonic. Intestinal adenomatous polyps can occur anywhere in the gastrointestinal tract except the oesophagus, and have the potential for malignant transformation. Gastric polyps are less common, and they do not show a risk of malignant transformation. Peptic ulcers can also be observed. Consequently, routine upper endoscopy and colonoscopy are crucial for early diagnosis and treatment. Annual upper endoscopy and colonoscopy are advised until age 35 years and every 2–3 years thereafter[3,6]. In the case presented here, the difficulty in proceeding with routine upper endoscopy because of limited mouth opening was critical, underlining the need for prompt management and treatment.

Multiple soft tissue lesions on the face, scalp, and extremities are typically observed in GS patients[6]. Epidermoid cysts are present in approximately 50–65% of patients, with no malignant potential[3]. Dental abnormalities such as supernumerary teeth, congenital missing teeth, impacted teeth, and odontomas are present in 30–75% of GS patients[3,6]. In the case presented here, the 17-year-old female had a history of multiple cutaneous and subcutaneous benign tumours and lesions since she was 3 years old, and no dental abnormalities were identified on routine radiographs.

One of the significant challenges faced by patients with GS is the presence of osteomas in the craniofacial region, leading to limited mouth opening and functional impairment. Over 70% of patients suffering from GS have osteomas. The identification of multiple osteomas is pivotal in the diagnosis of GS. Clinically, most osteomas are asymptomatic, however, depending on their location within the craniofacial complex, they can cause disfigurement, asymmetry, and dysfunction such as limited mouth opening[2,3]. Radiographic and CT scan features are characterized by round or oval radiopaque masses. De Oliveira Ribas et al.[7] reported a case of limited

mouth opening due to bilateral coronoid hyperplasia, later diagnosed to be GS. Osteomas that cause dysfunction, such as restriction of mandibular movement, may require resection[4]. This appears to be the first report of craniomaxillofacial manifestations of GS treated with bilateral customized TMJ-TJR, suggesting promising prospects for the treatment of this complex pathology.

The importance of clinicians' role in the early diagnosis of GS cannot be underestimated. The removal of the major osteomas with bilateral customized TMJ-TJR and coronoidectomy has emerged as an important treatment option to restore the patient's normal jaw function and improve quality of life.

According to Seehra et al.[2], a maxillofacial pathology such as osteoma always appears to precede the presentation of intestinal polyps. The patient in this case report presented maxillofacial manifestations many years before the gastrointestinal lesions.

It is important to highlight that the success of rehabilitation with customized TMJ-TJR in patients with GS and limited mouth opening will depend on careful patient selection, meticulous surgical planning, and precise prosthesis fabrication. Additionally, long-term monitoring is crucial to identify potential complications or prosthetic-related issues that may arise over time. Moreover, each patient should be evaluated carefully on an individual basis, with treatment planning considering the severity of the condition, patient age, associated comorbidities, and patient concerns.

Further research and long-term results are warranted to assess the durability and long-term success of customized total alloplastic temporomandibular joint replacement in the treatment of limited mouth opening in patients with Gardner syndrome.

Ethical approval

Not required.

Patient consent

Written informed consent obtained.

Funding

None.

Competing interests

None.

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