Gardner Syndrome: Presurgical Planning and Surgical Management of Craniomaxillofacial Osteomas

Matteo Brucoli, MD, Mariangela Giarda, MD, and Arnaldo Benech, PhMD

Abstract: Gardner syndrome, a variant of familial adenomatous polyposis, is an autosomal dominant genetic disease characterized by the combined presence of multiple intestinal polyps and extraintestinal manifestations. The extraintestinal manifestations include multiple osteomas, connective tissue tumors, thyroid carcinomas, and hypertrophy of the pigmented epithelium of the retina. Osteoma is a benign neoplasm of bone tissue characterized by slow continuous growth that usually affects the long bones and cranial bones and is a major symptom for Gardner syndrome. The authors report the extraintestinal lesions affecting the maxillofacial regions in 2 male patients (father and son) with Gardner syndrome. The presurgical planning and surgical management of these lesions are described.

Key Words: Osteomas, Gardner syndrome, computerized tomography, familial adenomatous polyposis

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Gardner syndrome, a variant of familial adenomatous polyposis,[1] is an autosomal dominant genetic disorder characterized by the combination of multiple intestinal polyps and extraintestinal manifestations. It is caused by a mutation of the APC gene on chromosome 5, has an early onset, and affects both sexes.[1] The extraintestinal manifestations differentiating Gardner syndrome from other forms of familial adenomatous polyposis generally precede the appearance of the intestinal lesions and include connective tissue tumors, bone lesions, dental alterations, thyroid carcinomas, and characteristic hypertrophy of the pigmented epithelium of the retina.[2] The bone lesions are osteomas usually affecting the long bones and cranial bones and are necessary for making the diagnosis[2]; in particular, a location in the mandibular angle is characteristic.[3] The maxillofacial osteomas are responsible for facial asymmetry, altered dental eruption, and interference with mastication.[4] Radiologically, they appear as roundish or oval radiopaque, bone-thickening lesions whose base is united with the underlying bone.[5] All patients with osteomas must be carefully assessed to exclude the presence of Gardner syndrome, thus allowing an early intervention aimed at avoiding malignant evolution of the intestinal lesions.[4,6] The only available means of avoiding this is early diagnosis and preventive colectomy. The imaging methods used are orthopantomography and computed axial tomography, which, also by means of three-dimensional image reconstructions, allow a more precise characterization that is useful when programming the corrective intervention. The bone lesions, particularly the facial areas, should be treated surgically if they lead to major deformity or interfere with normal function.[4,7] We report the extraintestinal lesions affecting the maxillofacial region found in 2 members of the same family with Gardner syndrome. We underline the importance of preoperative radiologic imaging in planning the correct surgical approach and describe surgical management.

CLINICAL REPORT

Two male patients (a son and father aged 25 and 52 years) were diagnosed as having Gardner syndrome in 1992. Both patients came to our attention because of multiple, painless, maxillofacial neoformations. The son, 25 years old, with a history of directed polypectomies, had a number of hard and painless swellings on the lower mandibular border and lacked a large number of teeth; his father, 52 years old, had previously undergone total colectomy and had various hard, painless, and nontender swellings at the level of the right frontal bone, the left lower edge of the orbit, and bilaterally at the mandibular angle. Orthopantomograms (Fig. 1) and computed tomography (CT) images (LightSpeed Plus; GE Medical Systems, Milwaukee, WI) were taken for planning the operation. Closely spaced, 1.25-mm-thin axial scans of the maxillofacial region and a high-definition bone image processing were applied (Figs. 2 and 4); this was completed by three-dimensional reconstructions (three-dimensional volume rendering [VR]) (Figs. 3 and 5) using a dedicated software (Advantage Workstation 4.0; GE Medical Systems). Orthopantomography was positive in both cases: there were numerous, well-circumscribed radiopaque lesions and dental alterations, a picture that was compatible with mandibular osteomas (Fig. 1), a typical extracolic localization in Gardner syndrome. The

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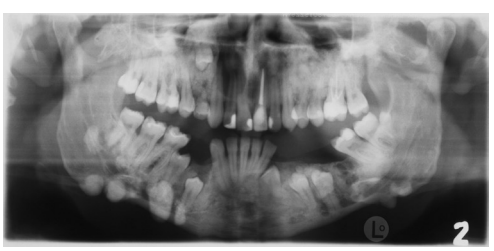

FIGURE 1. Patient 1, orthopantomography: multiple osteomas, odontomas, and unerupted teeth.

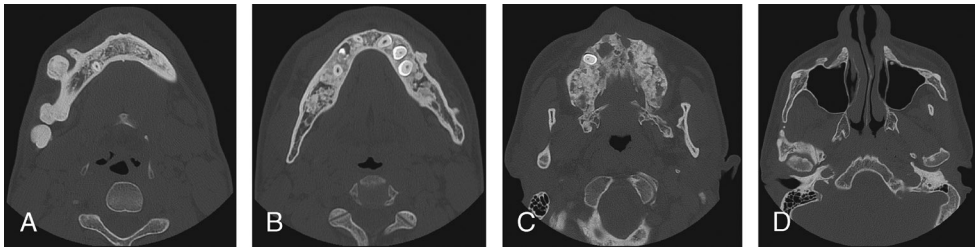

FIGURE 2. A–D, Patient 1, MDCT: axial images showing many mandibular and maxillary osteomas.

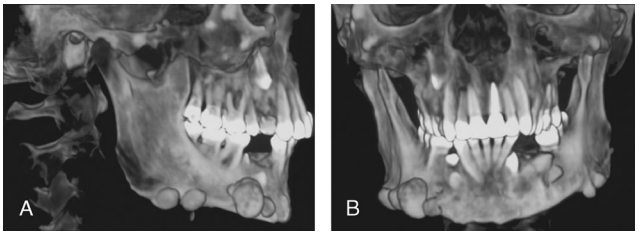

FIGURE 3. Patient 1, MDCT: VR reformat in lateral and anteroposterior views, showing mandibular and maxillary osteomas.

CT examination of the first patient (Fig. 2) confirmed the presence of mandibular osteomas and also revealed initial maxillary lesions, osteo-thickening bone remodeling of the spongy bone of the mandible and the maxillary bone, and initial millimetrical lesions involving both condyles. Volume-rendering reconstruction (Fig. 3) confirmed these findings, provided three-dimensional views at different angles of rotation, and showed the morphology of the lesions. In the second patient, the CT (Fig. 4) and VR reconstructions (Fig. 5) revealed similar alterations involving both mandibular angles, the left mandibular subcondyle, and the ethmoid bone, but not the osteo-thickening spongy bone remodeling visible in the first. Both patients underwent surgical resection of mandibular lesions under general anesthesia. Mandibular osteomas were approached by an intraoral approach. After exposure of mandibular bone, a reciprocating saw and osteotomes were used to resect mandibular osteomas. Histological examination (Fig. 6) confirmed the osseous nature of the lesions in both cases, which seemed to consist of trabeculae made of compact and mature lamellar bone tissue associated with remodeling phenomena. The postoperative courses were uneventful, and 1 year postoperatively, no recurrence was noted.

DISCUSSION

Gardner syndrome, a variant of intestinal adenomatous polyposis, is an autosomal dominant disease characterized by intestinal polyposis, connective tissue, and multiple osteomatoses most frequently affecting the cranium, facial bones, and teeth.[1] Osteomas are required to make the diagnosis of Gardner syndrome.[8] In particular, it is typical to find even large pedunculate osteomas in the mandibular angle.[3] Osteomas may precede clinical and radiologic evidence of colonic polyposis, so patients with osteomas should be evaluated for Gardner syndrome.[9] Osteoma is a benign neoplasm consisting of well-differentiated compact bone that increases in size by continuous osseous growth.[3] In our cases, the osteomas prevalently involved the mandible and had the characteristic pedunculate appearance; furthermore, numerous small osteomas were visible in the alveolar process of the mandible, the upper maxilla, and the paranasal and ethmoid sinuses. The osteo-thickening alveolar remodeling can be interpreted as the hypercementosis reported as being typical of the syndrome.[3] No previous reports have been made in literature concerning the contribution of multi–detector row CT (MDCT) and three-dimensional postprocessing of the images in patients with Gardner syndrome. The three-dimensional reconstructions of the CT images were very useful for planning the corrective removal of the largest osteomas, which would otherwise have led to alteration in the facial profile and interfered with mastication. In the case of Gardner syndrome, it is necessary to study

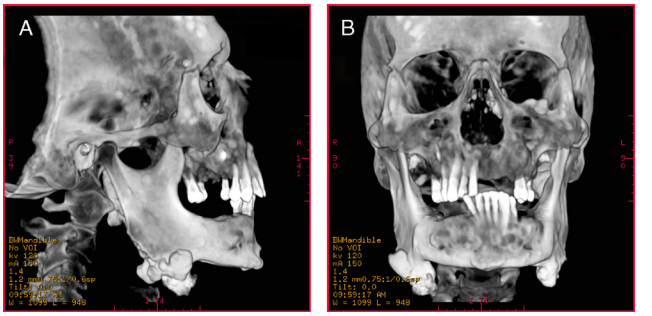

FIGURE 5. Patient 2, MDCT: VR lateral (A) and anteroposterior (B) views reconstructions; ethmoidal and mandibular osteomas can be recognized. Classic localization at the mandibular angle of osteomas.

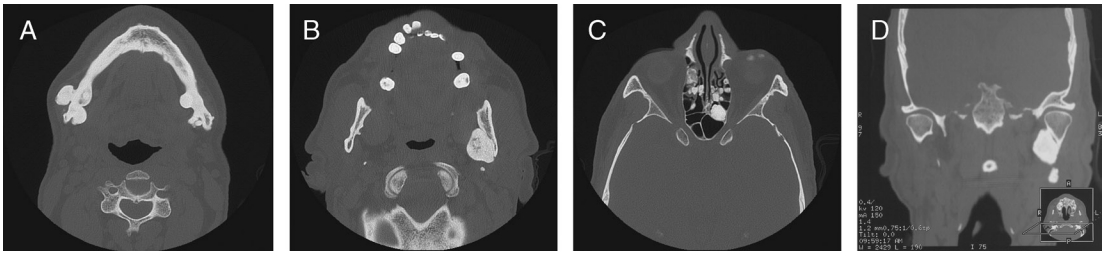

FIGURE 4. A–F, Patient 2, MDCT: axial (A–C) and coronal (D) reconstructed images showing mandibular, condylar, and ethmoidal osteomas.

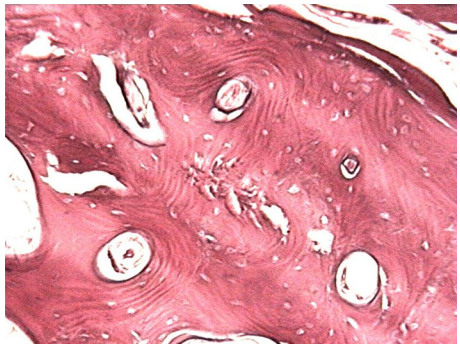

FIGURE 6. Histologic analysis: lamellar bone tissue associated with bone remodeling.

the maxillary bone compartment to check for the possible presence of osteomas. The diagnostic workup in such cases includes orthopantomography followed by multiplanar spiral CT and three-dimensional reconstructions of the images to allow precise preoperative planning. Treatment of the osteoma consists of complete surgical removal at the base where it unites with cortical bone.[10] The osteoma recurrence after removal is rare; furthermore, it is appropriate to provide periodic clinical and radiographic follow-up after surgical excision.

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