CASE REPORT

DOI: 10.5336/caserep.2018-63701

Gardner Syndrome Associated with Late Mandibular Osteoma

Orkhan ISMAYILOV[a],
Ali EKEMEN[a],
Haydar CELASİN[b],
Hakan Alpay KARASU[a],
İbrahim Ethem GEÇİM[c]

[a]Department of Oral and Maxillofacial Surgery, Ankara University Faculty of Densitry,
[b]Department of General Surgery, Lokman Hekim University Faculty of Medicine,
[c]Department of General Surgery, Ankara University Faculty of Medicine, Ankara, TURKEY

Received: 14 Nov 2018
Received in revised form: 07 Mar 2019
Accepted: 11 Mar 2019
Available online: 15 Mar 2019

Correspondence:
Orkhan ISMAYILOV
Ankara University Faculty of Dentistry, Department of Oral and Maxillofacial Surgery, Ankara, TURKEY
dr.orhan.ismayilov@gmail.com

ABSTRACT Osteomas are well-differentiated, slowly-growing and asymptomatic benign tumors of mature bone. Osteomas are usually observed on the jaw and classified into two types as central and peripheral osteoma. Diagnosis is made by observing radio-opacities on computed tomography. While follow-up is recommended for minor cases, surgical excision should be performed for major cases. Gardner syndrome is an autosomal dominant disease characterized by intestinal polyps and multiple osteomas. Due to the malignancy potential of intestinal polyps, early diagnosis and treatment are important. In this case report, diagnosis, surgical treatment and two years follow-up of a patient with Gardner syndrome are presented.

Keywords: Gardner syndrome; familial adenomatosis polyposis; protocolectomy; late osteoma

Osteoma is a slowly-growing, asymptomatic and benign bone tumor.[1] Although its pathogenesis is not clearly understood, there are several theories have been proposed. Some authors consider that the development of osteoma is associated with stimulation of osteogenic cells by muscle contraction due to trauma.[2] As in the study of Kashima et al., muscle traction may play a role in development of osteoma.[3] Periosteal ossifying fibroma, exostosis, chondroma, osteosarcoma, fibrosis dysplasia, Paget's disease and chronic osteomyelitis should be considered for the differential diagnosis of osteomas. Osteomas are most often confused with exostosis.[4] The lesions were considered to be osteomas due to the presence of polypozis coli. Although osteomas are generally diagnosed in the early period of Gardner Syndrome; they may also be detected in the late period, as in this case. Therefore, long-term follow-up is recommended.

Gardner syndrome should be considered for the patients presenting with osteoma, in accordance with family history and the patient should be referred to colonoscopic examination for the presence of intestinal polyps. Likewise, patients diagnosed with intestinal polyp should be referred to the department of oral and maxillofacial surgery considering the presence of osteoma in the mandible. The aim of this study was to describe the diagnosis and treatment of Gardner syndrome.

CASE REPORT

The mother of the patient was learned to have been diagnosed with familial adenomatosis polyposis (FAP) and had been, treated with total colectomy and ileorectal anastomosis and a secondary liver cancer had been developed thereafter. During this treatment, patients family was invited for screening and her son was diagnosed with FAP at the age of 20. Restorative proctocolectomy was performed due to multiple adenomatous polyps detected on colonoscopy. During follow-up, millimetric polyps were detected in the anal transition zone (ATZ) and repeated polypectomies were performed during follow-up periods. During the last physical examination, after 16 years from the initial surgery, a solid mass was detected on the lower part of the mandible and he was referred to the Department of Oral and Maxillofacial Surgery in the Faculty of Dentistry of Ankara University for the evaluation of the palpable mass and asymmetry on the angle of the mandible. After the physical examination, computerized tomography scan revealed four radiopaque and oval bone lesions in the mandible; three small lesions on the right side and one large lesion on the lower part of the left side (Figure 1). The patient underwent surgery under general anesthesia and local anesthetic with vasoconstrictor was administered to incision line. Subsequently, cutaneous and subcutaneous tissues were incised to platysma muscle layer in parallel with the lower edge of the mandible and 2 cm distant from the bone with no 10 scalpel. The deep cervical fascia was exposed by blunt dissection through superior-inferior fibers of platysma muscle. By preserving the marginal mandibular branch of the facial nerve, an incision measuring the same with the skin incision was performed on the platysma muscle. The superficial and deep layers of cervical fascia were dissected and periosteum was exposed (Figure 2). The periosteum was dissected with no 15 scalpel blade to reveal the mass. Osteoma was completely excised with drill and osteotomes (Figure 3). The lower edge of the mandible was corrected with round drills and the layers were sutured anatomically (periosteum, fascia, muscle, subcutaneous and cutaneous tissues in order). In order not to lead the pathologist to the diagnosis of osteoma, the removed specimen was sent to the laboratory without inform of pre-diagnosis of Gardner syndrome. The pathology result of the specimen was reported as osteoma.

The patient is under being followed up for 24 months and no intestinal recurrence was detected. An X-ray was performed during follow-up after 2 years and the incision site was detected to have anatomic borders and no recurrence was detected (Figure 4).

DISCUSSION

Isolated osteomas are very rare in the stomatognathic system and they most commonly occur at the angle and body of the mandible. On the other hand, multiple lesions are associated with Gardner syndrome.[5]

According to a study conducted in 2002, osteomas do not vary in accordance with age and gender.[6] Nevertheless, Kaplan et al. showed that osteoma has a higher incidence in males. They are frequently observed between 16 and 74 years and occur at 3rd or 5th decade of life.[7] Our patient's being an adult is consistent with literature.

In clinical practice, osteomas may be observed as well-demarcated oval shaped tumors with pedicles or unilateral, slowly growing asymptomatic adherent tumors and externally adherent to the bone surface. Osteoma may be observed as a radiopaque mass with bone density and with a thin radiolucent line around. Three-dimensional computed tomography was shown to be the best radiographic method for diagnosis of osteomas.[8] The presented case is clinically and radiographically consistent with literature data.

Gardner syndrome is considered as a version of familial adenomatous polyposis (FAP); however, the presence of osteomas should be excluded. FAP is characterized by the presence of hundreds of intestinal polyps. Gardner syndrome is the association of polyposis, osteomas and soft tissue tumors (desmoid tumors, fibromas, epidermoid cysts). As the malignancy risk of intestinal polyps is very high, early diagnosis has vital importance. The dis-

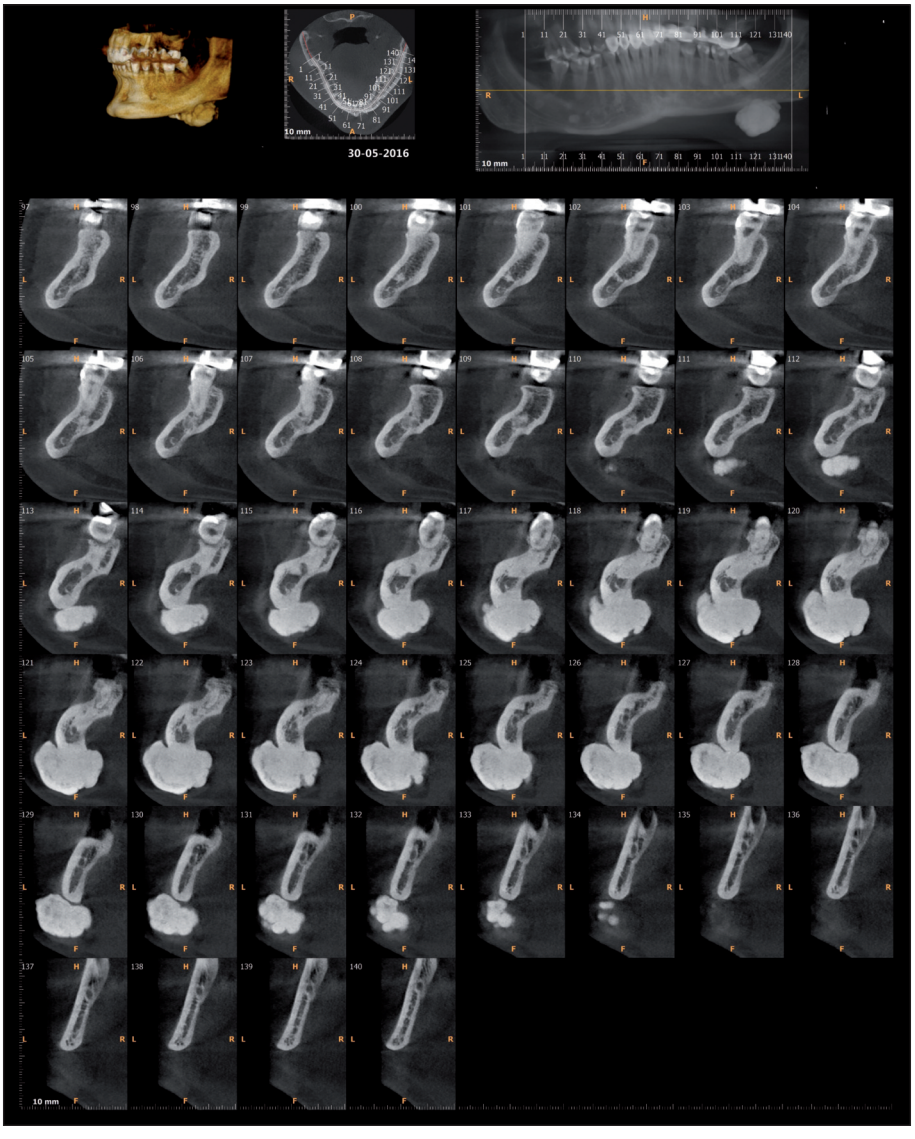

FIGURE 1: Preoperative CBCT.

ease is autosomal dominant and colorectal adenocarcinoma may develop in untreated patients. Although pathological lesions, such as osteomas, skin cysts, etc., may be asymptomatic for a long time; polyposis lesions may lead to symptoms, such as rectal bleeding, diarrhea, and abdominal pain.[9] Also Gardner's syndrome (Familial adenomatous polyposis) is associated with papillary thyroid cancer in 89% of the cases.[10,11] In the present case, symptoms of Gardner syndrome were noted.

If osteoma is diagnosed during head and neck examination, upper and lower endoscopic examination should be recommended. In patients with

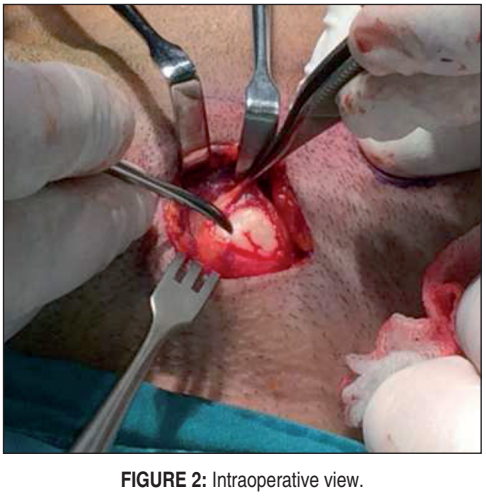

FIGURE 2: Intraoperative view.

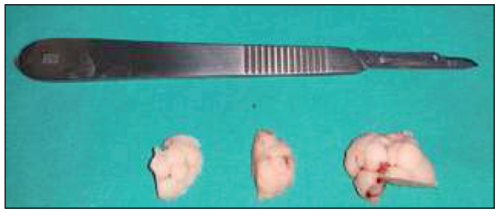

FIGURE 3: Removed Lesion.

colonic or duodenal polyposis, the investigation of osteoma and papillary thyroid cancer by performing head and neck examination will be useful.

Informed Consent

Informed consent was obtained from the patient prior to the operation.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or

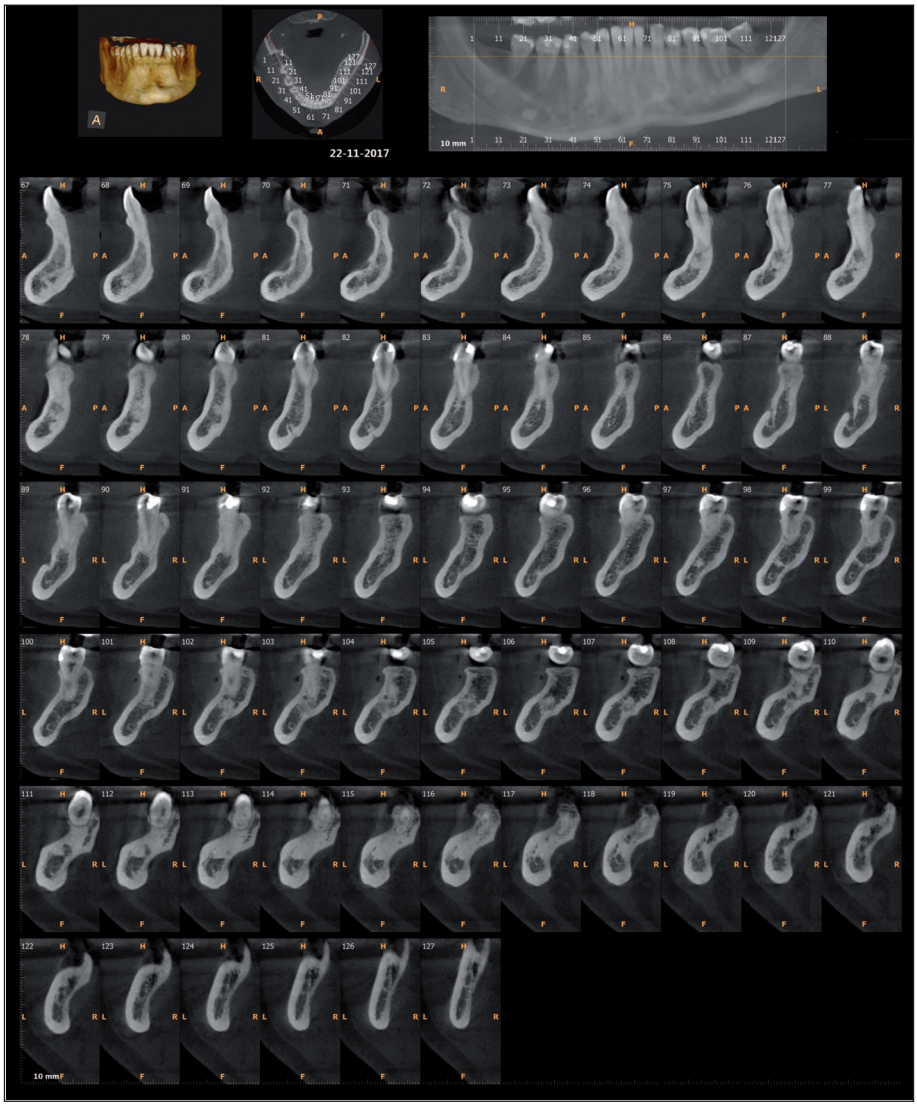

FIGURE 4: Postoperative 24-month control CBCT.

members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Ibrahim Ethem Gecim, Hakan Alpay Karasu, Orkhan Ismayilov;* ***Design:*** *Haydar Celasin, Ali Ekemen;* ***Control/Supervision:*** *Hakan Alpay Karasu, Ali Ekemen, Orkhan Ismayilov;* ***Data Collection and/or Processing:*** *Haydar Celasin, Ibrahim Ethem Gecim, Ali Ekemen;* ***Analysis and/or Interpretation:*** *Orkhan Ismayilov, Ali Ekemen;* ***Literature Review:*** *Ibrahim Ethem Gecim, Hakan Alpay Karasu, Haydar Celasin;* ***Writing the Article:*** *Orkhan Ismayilov, Ibrahim Ethem Gecim, Hakan Alpay Karasu;* ***Critical Review:*** *Hakan Alpay Karasu, Ali Ekemen;* ***References and Fundings:*** *Haydar Celasin, Orkhan Ismayilov;* ***Materials:*** *Ibrahim Ethem Gecim, Hakan Alpay Karasu, Orkhan Ismayilov.*

REFERENCES

1. Kerckhaert A, Wolvius E, van der Wal K, Oosterhuis JW. A giant osteoma of the mandible: case report. J Craniomaxillofac Surg. 2005;33(4):282-5. [Crossref] [PubMed]
2. Kaplan I, Nicolaou Z, Hatuel D, Calderon S. Solitary central osteoma of the jaws: a diagnostic dilemma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2008;106(3):e22-9. [Crossref] [PubMed]
3. Kashima K, Rahman OI, Sakoda S, Shiba R. Unusual peripheral osteoma of the mandible: report of 2 cases. J Oral Maxillofac Surg. 2000;58(8):911-3. [Crossref] [PubMed]
4. Guimarães KB, Cavalcante JR, Ferraro-Bezerra M, Silva DN, de Holanda Vasconcellos RJ, do Egito Vasconcelos BC. Peripheral osteoma bilateral of the mandible without association with Gardner syndrome. J Craniofac Surg. 2012;23(2):e83-6. [Crossref] [PubMed]
5. Zouloumis L, Lazaridis N, Maria P, Epivatianos A. Osteoma of the ethmoidal sinus: a rare case of recurrence. Br J Oral Maxillofac Surg. 2005;43(6):520-2. [Crossref] [PubMed]
6. Sayan NB, Uçok C, Karasu HA, Günhan O. Peripheral osteoma of the oral and maxillofacial region: a study of 35 new cases. J Oral Maxillofac Surg. 2002;60(11):1299-301. [Crossref] [PubMed]
7. Kaplan I, Calderon S, Buchner A. Peripheral osteoma of the mandible: a study of 10 new cases and analysis of the literature. J Oral Maxillofac Surg. 1994;52(5):467-70. [Crossref]
8. Ogbureke KU, Nashed MN, Ayoub AF. Huge peripheral osteoma of the mandible: a case report and review of the literature. Pathol Res Pract. 2007;203(3):185-8. [Crossref] [PubMed]
9. Waller A, Findeis S, Lee MJ. Familial adenomatous polyposis. J Pediatr Genet. 2016;5(2):78-83. [Crossref] [PubMed] [PMC]
10. Bell B, Mazzaferri EL. Thyroid cancer in familial polyposis coli: case report and literature review. Dig Dis Sci. 1993; 38(1):185-90. [Crossref] [PubMed]
11. Isik A, Firat D, Yilmaz I, Peker K, Idiz O, Yilmaz B, et al. A survey of current approaches to thyroid nodules and thyroid operations. Int J Surg. 2018;54(Pt A):100-4. [Crossref] [PubMed]