Gardner Syndrome Associated with Late Mandibular Osteoma

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. 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. As in the study of Kashima et al., muscle traction may play a role in development of osteoma. 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. The lesions were considered to be osteomas due to the presence of polyposis 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.
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).

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. According to a study conducted in 2002, osteomas do not vary in accordance with age and gender. 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. 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. 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-
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
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.

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**Conflict of Interest**

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

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**References**