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Surgery

Fronto-Ethmoidal Osteoma in the Context of Gardner's Syndrome: Case Report

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Abstract | **Case Report**

Gardner syndrome is a rare autosomal dominant disorder caused by mutations in the APC gene. It is characterized by a triad of colorectal polyps with a high risk of malignant transformation, soft tissue tumors, and bone lesions, particularly osteomas of the maxillofacial region. Dental anomalies, including supernumerary teeth, odontomas, and dentigerous cysts, are highly prevalent and often serve as early indicators of the disease. The diagnosis is primarily clinical, supported by genetic testing and imaging. While surgical resection may be necessary for symptomatic osteomas, asymptomatic lesions are generally managed conservatively. Early recognition of oral and maxillofacial manifestations is crucial for timely diagnosis, preventive care, and appropriate surveillance, particularly for colorectal malignancies. A multidisciplinary approach involving dental, surgical, and gastroenterological teams is essential for optimal patient management. This study aims to discuss the clinical presentation, diagnostic challenges, and management of maxilla-facial symptoms. We report a case of a patient diagnosed with Gardner syndrome with exophthalmos, treated successfully.

Keywords: Gardner Syndrome, Orbiatal Osteoma, Odontoma, Jaw Tumors, Familial Adenomatous Polyposis, Craniofacial Surgery, Genetic Counseling.

INTRODUCTION

Gardner syndrome is a rare, autosomal dominant genetic disorder characterized by the development of multiple colorectal adenomatous polyps, as well as various extracolonic manifestations, including osteomas, dental abnormalities, epidermoid cysts, and soft tissue tumors. It is considered a variant of familial adenomatous polyposis (FAP) and is caused by mutations in the APC (Adenomatous Polyposis Coli) gene located on chromosome 5q21 [1].

One of the hallmark features of Gardner syndrome is the presence of osteomas, particularly in the skull and facial bones, including the frontal and ethmoidal sinuses. These benign bone tumors can be asymptomatic or may cause complications such as sinus obstruction and facial deformities [2]. The syndrome also predisposes individuals to colorectal cancer, typically developing from adenomatous polyps if prophylactic colectomy is not performed [3].

Early diagnosis is crucial due to the high risk of malignant transformation of colonic polyps. Diagnosis is typically based on genetic testing, clinical evaluation, and imaging studies such as CT scans for osteomas and dental abnormalities [4].

CASE REPORT

This is a 33-year-old female patient who underwent surgery four years ago for a frontal and left mandibular osteoma and is now presenting with exophthalmos of the left eye.

On extraoral examination, the patient exhibits multiple bilateral mandibular swellings, located at the inferior orbital rim and the scalp. She comes from a consanguineous marriage and has a family history of Gardner syndrome among her siblings, characterized by multiple osteomas and familial adenomatous polyposis.

Palpation reveals that these tumors are hard, well-defined, non-adherent to the skin, and firmly attached to the mandibular cortex. The associated lymph node areas are free.

On intraoral examination, no lesions are detected. CT imaging reveals multiple mandibular osteomas of varying sizes and a retro-orbital fronto-ethmoidal osteoma, responsible for the exophthalmos of the left eye. She had a preserved Visual Acuity.

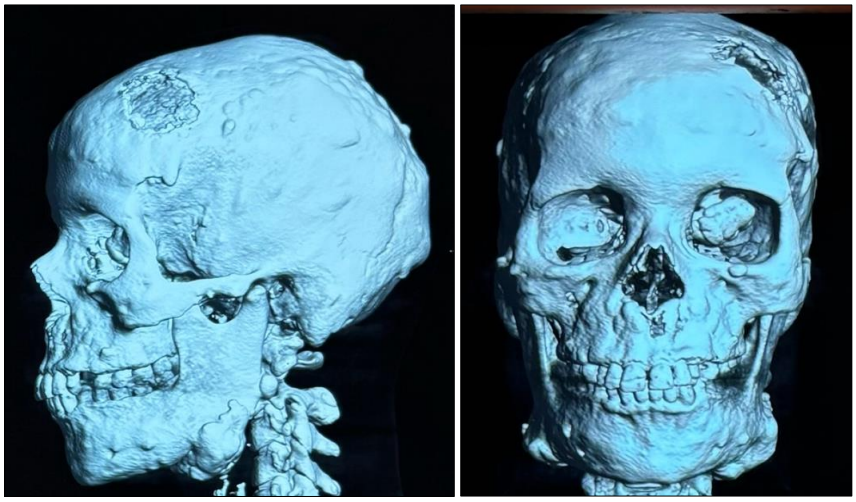

Figure 1: CT scan image showing multiple craniofacial osteomas

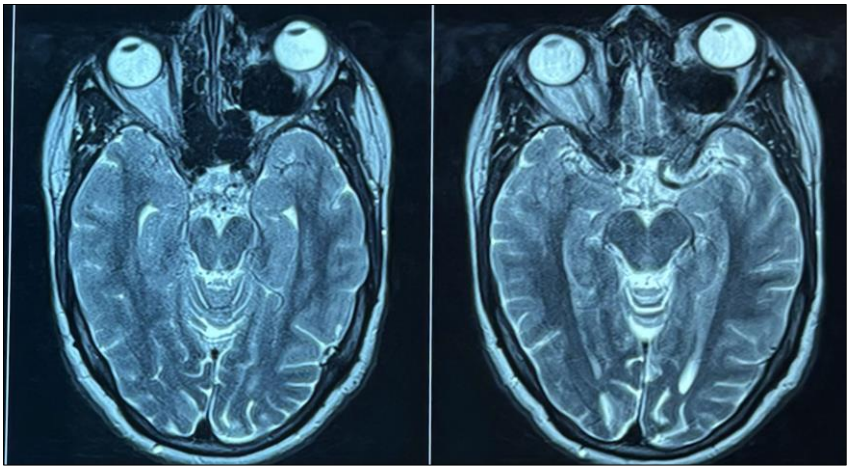

Figure 2: MRI showing intra-orbital tumor of the left eye responsible for the exophthalmos

A para-latero-nasal approach surgery was performed under general anesthesia to excise the left retro-orbital osteoma**.**

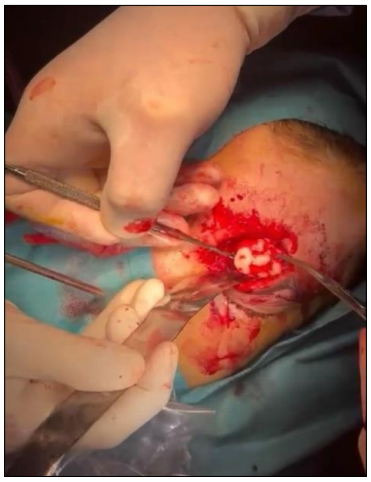

Figure 3: Per-operatory picture of the osteoma

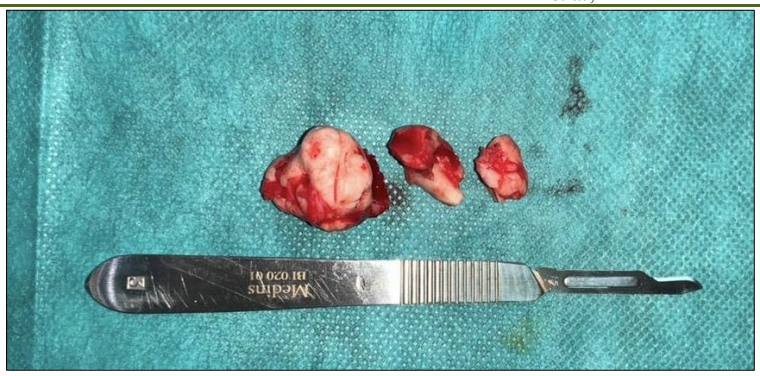

Figure 4: Resected osteoma

The clinical follow-up three months after surgery was satisfying. The patient didn"t show any signs of complication or recurrence.

DISCUSSION

Gardner syndrome is characterized by a triad consisting of colorectal polyps, which frequently undergo malignant transformation, soft tissue tumors, and bone tumors that primarily, but not exclusively, affect the bones of the face and skull. The disease, caused by a mutation in the APC gene, is autosomal dominant with complete penetrance and variable expressivity.

The rich dento-maxillary symptomatology and family history suggest a syndromic condition and confirm Gardner syndrome.

In most cases of Gardner syndrome reported in the dental literature, the dento-maxillofacial manifestations are described as part of an already known syndrome [5]. More rarely, it is the dento-maxillary manifestations that have led to the discovery of an unrecognized syndrome [6].

An earlier diagnosis should be possible, given the presence of stomatological manifestations from childhood or adolescence [7].

Bone lesions are predominantly osteomas. These benign tumors, which are almost exclusively located in the maxillofacial region, affect all the bones of the face, the paranasal sinuses, but especially the mandible [8].

Osteomas can be single or multiple, appearing as small, well-defined opacities, primarily located in the alveolar portion, forming enostoses or exostoses. They must be distinguished from odontomas or cementomas, as these two entities are also part of the syndrome. Peripheral formations (exostoses) are most commonly found on the basal border, where they create palpable or visible protrusions when they push against the overlying skin [9].

These osteomas are found in 80% to 90% of patients with Gardner's syndrome between 10 and 30 years old, whereas their prevalence in the general population is below 10% [10]. They exhibit the radiodensity and ultrastructure of mature compact bone, with a well-developed Haversian system. In some cases, osteomas may lead to late-onset functional complications, such as ocular or nerve compression, nasal obstruction, or reduced mobility. However, they are most often of cosmetic concern, which prompts patients to seek medical advice. They can develop early and may even precede the diagnosis of polyposis [11].

From a therapeutic perspective, despite their tendency to recur [5], osteomas may be eligible for surgical resection. However, when they are asymptomatic, especially in the case of enostoses, whose removal could lead to significant bone damage, there is no indication for intervention [12].

Dental anomalies are observed in 70% of cases, whereas only 1% of the general population presents dental anomalies [13]. These include dental agenesis, supernumerary teeth, multiple impactions, dentigerous cysts, odontomas, hypercementosis, and microdontia. These anomalies represent the most consistent feature of the syndrome. The incidence of supernumerary teeth in the general population ranges from 0.1% to 3.6%, whereas it reaches 38% in Gardner syndrome [14].

Generally, osteomas are of moderate size with limited growth potential. Their number and size may increase with age, so long-term monitoring is recommended [15].

This study highlights the importance of maxillofacial surgery, in cooperation with other specialties, in managing both the functional and aesthetic aspects of Gardner's syndrome. Patients with large osteomas often seek surgical removal. The excision technique depends on the location, size, and number of osteomas, requiring a tailored approach for each lesion and patient. The aesthetic and functional outcomes vary from case to case. When a patient has significant functional impairment, aesthetic considerations become secondary.

Certain oral and dental signs can aid in diagnosing this condition. The detection of radiopaque lesions in the jaws, suggestive of osteomas, or the presence of supernumerary teeth in a young patient should prompt a gastroenterology consultation to confirm or rule out Gardner's syndrome. Osteomas typically appear before the onset of intestinal polyposis. Early diagnosis is crucial, as it allows for regular monitoring of affected individuals, given the high risk of colorectal cancer in these patients.

CONCLUSION

Gardner syndrome is a rare genetic disorder characterized by a triad of colorectal polyps with malignant potential, soft tissue tumors, and bone lesions, primarily osteomas affecting the maxillofacial region. Dental anomalies, present in the majority of cases, play a crucial role in early diagnosis, as they often appear in childhood or adolescence. The high prevalence of supernumerary teeth, odontomas, and other dental abnormalities distinguishes this syndrome from the general population. While surgical management may be necessary for symptomatic osteomas, asymptomatic lesions typically do not require intervention. Early recognition of oral and maxillofacial manifestations is essential for timely diagnosis and management, helping to prevent serious complications, particularly colorectal cancer. Multidisciplinary follow-up involving dentists, oral surgeons, and gastroenterologists is critical for comprehensive care.

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