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Gardner's syndrome with bilateral osteomas of coronoid process resulting in limited opening

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Abstract

Gardner's syndrome is characterized by a triad of intestinal polyposis, which ultimately become malignant, soft-tissue neoplasms such as desmoid tumors and fibromas, and osteomas, particularly of the skull and facial bones. A case report of bilateral osteomas of the coronoid process resulting in limited incisal opening in a 12-year-old girl and review of the pertinent literature is presented.

Gardner's syndrome is a clinicopathologic entity of interest to the dental and medical professions and is characterized by intestinal polyposis, osteomatosis, and multiple soft-tissue neoplasms (Gardner and Richards 1953). Although many case reports described patients with some or all of the features of the triad, it was not until Gardner and others[1] documented several kindred in the early 1950s that the syndrome was well defined. Fader et al. (1962) and Fitzgerald (1943) described the facial and oral aspects of the disease which includes osteomas, impacted primary and permanent teeth, and supernumerary teeth. The etiology is through an autosomal dominant pattern of inheritance, with a high degree of penetrance.[2] The disease has been diagnosed from ages 2 through 70, with the prevalence reported at 1:14,000 (Bochetto et al. 1963; Pierce et al. 1970). Since the oral findings serve as an indicator of the underlying intestinal polyposis which has a malignant propensity of 100% by age 50, the syndrome is of acute interest to the dental profession.[3] This report describes an unusual presenting oral feature which ultimately led to the diagnosis of Gardner's syndrome.

Clinical Manifestations

The diagnosis of Gardner's syndrome can be confirmed on the basis of 3 major symptoms: intestinal polyposis, osteomatosis, and soft-tissue neoplasms. This diagnosis is further supported by abnormal dental findings, the hereditary nature of the disease, and ophthalmologic abnormalities.

The most serious aspect of the syndrome is intestinal polyposis which is predisposed to malignant transformation to adenocarcinoma at approximately 34 years (Dukes 1952). The polyps occur before puberty and become generalized in the late teens and early twenties (Coli 1970). The polyps are multiple and scattered, occurring at any location in the gastrointestinal tract, particularly in the distal colon, with a few cases of small bowel mucosal involvement.[4] The symptoms may include diarrhea, passage of blood or mucosa, and cramp-like abdominal pain (Gumpel and Carballo 1956). Surgical intervention is required as a preventive measure (Jones and Cornell 1966).

Osteomas consist of dense bony proliferations of histologically normal membranous bone which vary from slight thickening to large masses and may affect all parts of the skeleton (Weary et al. 1964; Chang et al. 1968). Between 50 and 80% of patients are affected, showing an average number of 5.9 osteomas per person with the frontal bones as the most frequent site.[5] The 2 different general types of osteoma are: a protuberant mass with a broad base which presents as a palpable lump; and a sessile mass which projects into the paranasal sinus. In the mandible, the 2 types of

[1] Gardner 1951, 1962; Gardner and Richards 1953; Gardner and Stephens 1950; Gardner and Woolf 1952; Gardner and Plenk 1952; Rayne 1968.

[2] Gardner 1951; Coli et al. 1970; Collins 1959; Duncan et al. 1968; Hughes and Heuston 1960.

[3] Lindqvist et al. 1983; Jones and Cornell 1966; Ida et al. 1981; Jarvinen et al. 1982.

[4] Coli et al. 1970; MacDonald et al. 1967; Gumpel and Carballo 1956.

[5] Rayne 1968; Ida et al. 1981; Chang et al. 1968.

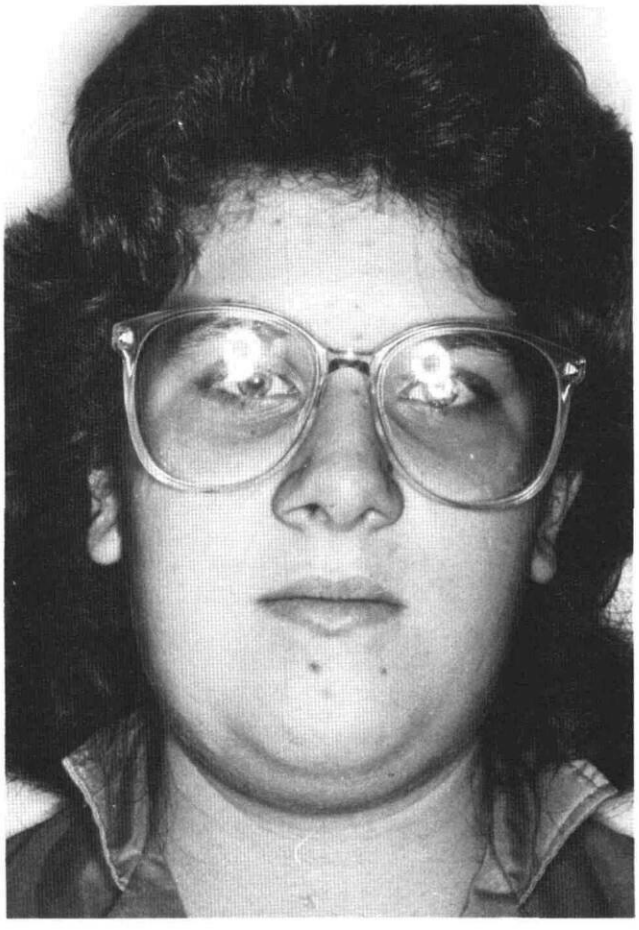


FIG 1. Normal facies in a 12-year-old with Gardner's syndrome.

osteomas occur centrally or lobulated on the cortex. A centrally located osteoma appears as a mass near the roots of teeth, whereas a lobulated osteoma arises from the cortex, characteristically at the mandibular angle (Chang et al. 1968). Generally, the osteomas grow slowly, reaching a stationary size, and then become dormant, although actively growing fibro-osseous tumors of the mandible have been reported.[6] Since osteomas may precede polyp formation and are a useful predictor, it has been recommended to include a panoramic radiograph as a screening procedure in all patients suspected of having the syndrome (Ida et al. 1981; Jarvinen et al. 1982).

The most frequent soft-tissue neoplasm is the epidermoid inclusion cyst, usually found on the face or extremities (Jones and Cornell 1966; Jarvinen et al. 1982). Also reported are lipomas, leiomyomas, and neurofibromas (Gumpel and Carballo 1956; Laberge et al. 1957). Desmoid tumors or aggressive fibromatosis, which frequently appear in abdominal scar tissue, are found in 17% of patients with Gardner's syndrome.[7] Since they can compress and infiltrate muscle, desmoid tumors are considered locally invasive (Coli et al. 1970; Jones and Cornell 1966). The skin cysts may appear before intestinal polyps and are useful as a predictor (Leppard and Bussey 1975).

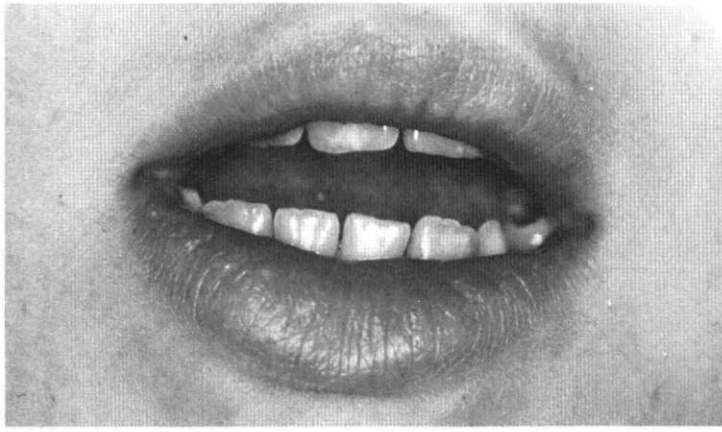


FIG 2. Limited incisal opening of 12 mm.

More than 50% of patients with Gardner's syndrome exhibit abnormal dental findings (Chang et al. 1968). Single or multiple supernumerary teeth are a common finding as well as the presence of hypercementosis.[8] Changes in trabecular bone pattern, impacted and unerupted teeth have been reported.[9] Ankylosis and difficulty in extraction of erupted teeth have been reported (Fitzgerald 1943; Amato and Small 1970). Several reports of chronic osteomyelitis are suggestive of an association with Gardner's syndrome (Fader et al. 1962; Calhoun et al. 1957).

Multiple and bilateral patches of congenital hypertrophy of the retinal pigment epithelium have been related to the Gardner's syndrome gene and may be useful in identifying patients at risk (Lewis et al. 1984; Blair and Trempe 1980).

Since the etiology is through an autosomal dominant pleiomorphic gene with a high degree of penetrance, a complete family history is very important to the diagnosis.[10] Other genetic neoplasms have been reported in Gardner's syndrome, such as 1 case of thyroid papillary adenocarcinoma in a family (Comiel et al. 1968).

Case Report

A 12-year-old white female was admitted to Children's Hospital of Michigan with a chief complaint of an acute problem which included limited jaw opening of approximately 12 mm and multiple, grossly

[6] Coli et al. 1970; Weary et al. 1964; Small et al. 1980.

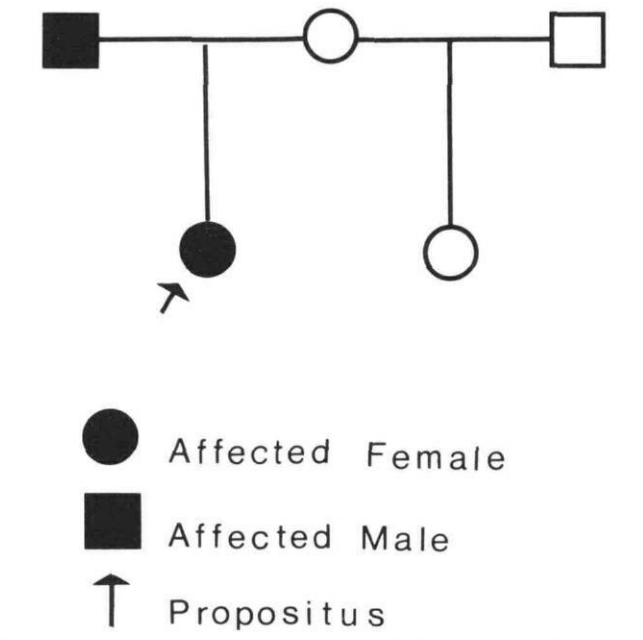
[7] Gardner 1962; Hughes and Heuston 1960; Jones and Cornell 1966; Jarvinen et al. 1982; Gorlin and Chaudhry 1960.

[8] Fader et al. 1962; Fitzgerald 1943; Chang et al. 1968.

[9] Fader et al. 1962; Fitzgerald 1943; Davies 1970; Chang et al. 1968; Bessler et al. 1984.

[10] Gardner 1951; Coli et al. 1970; Duncan et al. 1968; McKusick 1962.

TABLE 1. Pedigree of Family With Gardner's Syndrome



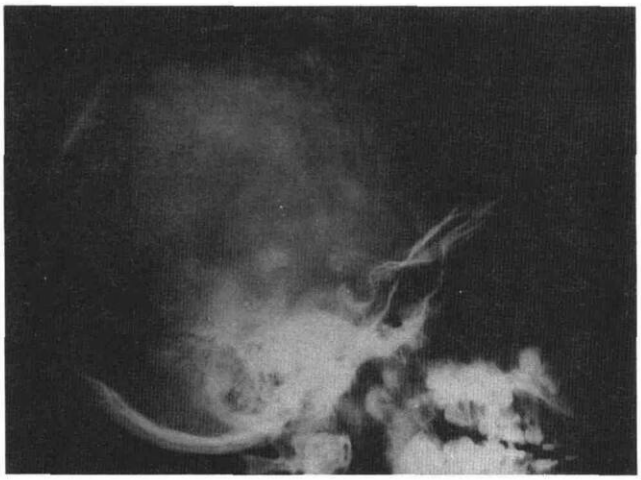


FIG 3. Skull radiograph reveals multiple osteomas, particularly of the parietal region, which measured 8–15 mm in diameter.

carious and abscessed teeth (Figs 1, 2). Approximately 7 months prior, she was seen by her family physician who referred her for a complete evaluation for Gardner's syndrome.

She presented as a well developed, cooperative child with normal facies and a positive family history in her father for Gardner's syndrome (Table 1). No other family members were known to have the disease. She presented with a 2 × 2 mm sebaceous cyst of the scalp and bony protrusions of her heels. Benign polyps of the sigmoid colon had been removed by her physician 2 months prior to her admission.

A series of radiographs of the upper and lower gastrointestinal tract, including a barium enema, and air contrast techniques were obtained. Significant findings included at least 6 polyps, 3–5 mm in size, in the rectosigmoid area with probable lymphoid hyperplasia of the ascending colon.

The full skeletal survey revealed osteomas only in the skull area. The skull radiographs revealed multiple osteomas of the bony calvarium, especially in the parietal region, which measured 8–15 mm in diameter (Fig 3). In the floor of the sphenoid sinus appeared a 7.0 mm osteoma. In the mandible, at least 6 separate areas of osteomas were found, measuring 1–3 cm in diameter, most prominent near the angle of the mandible.

Her temporomandibular joint, defined by computed tomography (CT), revealed osteomas which were not detected initially on the skull radiograph (Fig 4). The joint space was normal, with evidence of osteomas projecting from the coronoid process of the mandible. Osteomas were noted on the left ascending ramus, in the body of the mandible, the sphenoid bone, lateral and medial pterygoid plates, and medial temporal bone. Both rami were deformed.

The oral examination revealed an early mixed dentition, with a dental age delayed in comparison to her chronologic age. A panoramic radiograph revealed the presence of multiple impacted permanent teeth, including the maxillary and mandibular canines and premolars. Many supernumerary teeth were noted, which appeared between the overretained primary molars and the impacted premolars (Fig 5). The supernumerary teeth may have caused deflection of the premolars, resulting in an ectopic eruption pattern. The mandibular right second primary molar and the left and mandibular right first permanent molars were abscessed and unrestorable. The limited incisal opening was due to an increased enlargement and impingement of the bilateral osteomas located on the coronoid processes. These rapidly growing osteomas caused a mandibular shift to the left which affected her speech and ability to masticate.

The treatment indicated was a bilateral coronoidectomy and extraction of the first permanent molars. The patient presented initially as an anesthesia management problem due to her limited interincisal opening, and preoperatively it was anticipated that a fibro-optic intubation may be necessary. Under IV sedation, a blind nasal intubation was performed successfully. Postoperatively she received physical therapy instruction to exercise daily with a rubber bite block and tongue blade appliance; however, compliance was poor. The resultant incisal opening was maintained at 32 mm. The histopathology of the osteomas indicated dense, viable bone.

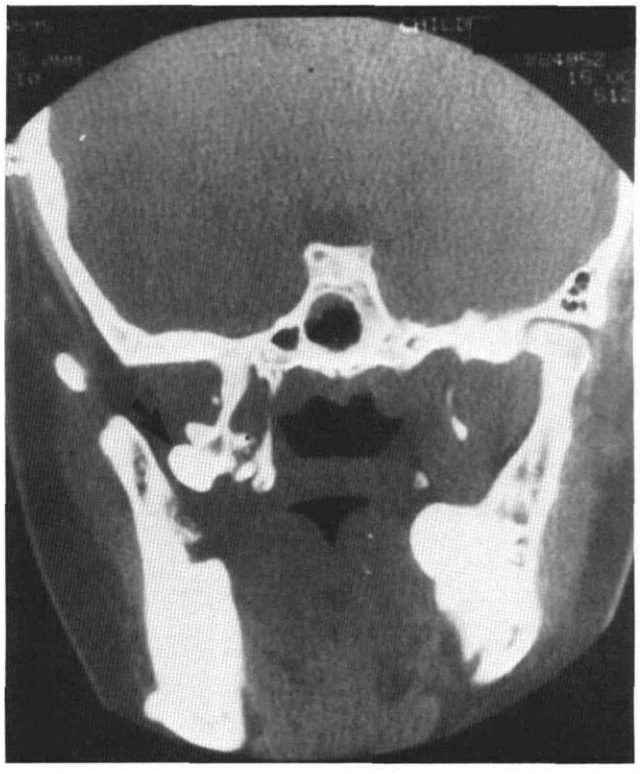


FIG 4. CT scan of skull with arrow pointing to an osteoma located off the lateral and medial pterygoid plate in the maxillary sinus.

Discussion

The patient exhibited the classic clinical picture found in Gardner's syndrome. She presented with minimal intestinal polyposis of the rectosigmoid area. Unless treated with a total colectomy or a partial one with ileoproctostomy (Duncan et al. 1968), all patients will die of colon adenocarcinoma by age 50 (MacDonald et al. 1967). At this time, the patient moved out of state and is being followed for gastrointestinal changes.

Another early sign of Gardner's syndrome is the presence of osteomas. The osteomas were limited to her skull and more than average in number (Ida et al. 1981). She presented with the classic feature of an osteoma located at the angle of the mandible (Chang et al. 1968). The majority of osteomas are slow growing, but may alter the facies thereby requiring surgical intervention (Rayne 1968). While the location of the osteomas varies and may interfere with incisal opening, the presence of bilateral osteomas on the coronoid process is an unusual finding. Limited opening has been reported due to an osteoma attached to the inferior border of the zygomatic arch, over the coronoid process (Fitzgerald 1943). Another report cited the presence of an osteoma in the right maxilla, which was in close proximity to the ascending ramus and interfered with function (Rayne 1968). Restricted

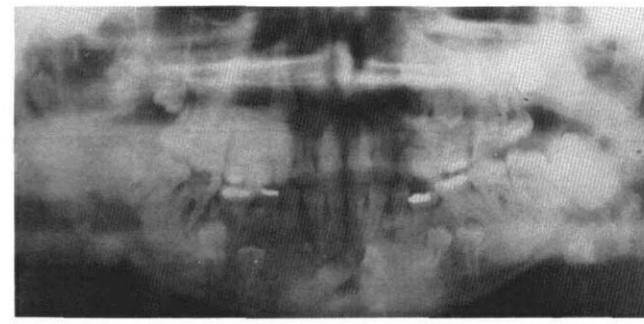


FIG 5. Panoramic radiograph reveals impacted permanent canines and premolars, supernumerary teeth, ankylosed primary molars, abscessed permanent molars, and multiple scattered osteomas.

mandibular movement was caused by an osteoma on the external body of the mandible (Fader et al. 1962). Difficulties in mastication and swallowing were described due to a pedunculated osteoma projecting from the retromolar pad (Lindqvist et al. 1983).

The finding of a sebaceous cyst on the back of the patient's skull was the only evidence of a soft-tissue tumor and completed the triad of the syndrome.

The abnormal dental findings supported the diagnosis of Gardner's syndrome and included the presence of multiple impacted supernumerary teeth, impacted permanent teeth, and ankylosed primary molars.

The patient's pedigree was positive for Gardner's syndrome and confirmed the autosomal dominant pattern of inheritance.

Summary

An unusual case of bilateral osteomas which caused decreased incisal opening in a 12-year-old girl with Gardner's syndrome is reported. The stigmata of the syndrome included intestinal polyposis, soft-tissue cyst, and osteomas of the skull and mandible. The diagnosis was supported by the presence of abnormal dental findings, which included multiple impacted supernumerary teeth, impacted permanent teeth, and ankylosis of primary molars.

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