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# Multiple cemental lesions in the jaw bones of a patient with Gardner's syndrome

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**Summary.** Unusual multiple lesions of jaws in a middle-aged woman with all classic manifestations of Gardner's syndrome are reported. Clinical examination revealed a diffuse swelling of the mandible and maxilla with bone-like hardness and numerous radiopaque lesions scattered throughout both mandible and maxilla. Impaction of the molar teeth was revealed by roentogenographic examination. Histopathologically, the multiple jaw lesions consisted of trabecular proliferation of hard-tissue which more closely resembled cementum than immature bone or osteoid. It was found to be united with the cementum of tooth-roots through obliteration of the periodontal ligament. The present lesions do not fit into any of the distinct entities of cemental lesions described.

**Key words:** Gardner's syndrome – Cemental lesions – Mandible – Maxilla

In 1912, Devic and Bussy first reported the association of generalized intestinal polyposis, osteomas of the mandible and multiple sebaceous cysts and lipomas. This was obviously Gardner's syndrome, in the light of present day knowledge. In subsequent years, other scattered case reports appeared in the literature (Cabot 1935; Fitzgerald 1943). Gardner and Richards (1953) described a study of a family group of 51 individuals, six of whom exhibited a triad of abnormalitis consisting of cutaneous lesions, osteomas and intestinal polyposis and concluded that the condition was a dominantly transmitted inheritable disorder with at least 80 per cent penetrance. The condition has since been known as Gardner's syndrome. Osteomas of Gardner's syndrome may involve various sites of bones, but are mainly detected in the jaw bones. In addition, odontomas, impacted teeth and/or hypercementosis have been described in about 50 per cent of cases (Fader et al. 1962; Gardner 1962; Jones et al. 1966; Chang et al. 1968; Duncan et al. 1968; Coli et al. 1970). However, heretofore, multiple cemental lesions of jaw bones have not been documented in any of the preceding reports of the syndrome. The aim of this paper is to report a case of multiple cemental lesions found in the mandible and maxilla of a female patient with Gardner's syndrome.

## Case report

*Clinical course.* A 38-year-old female was referred to our hospital with swelling of the left molar region of her mandible with pain, on December, 1976. The patient stated that she had first noted painless swelling of the mandible in childhood. She had not experienced any serious or chronic illnesses but her family history was unknown since she was an orphan. Clinical examination revealed that diffuse swelling of both mandible and maxilla with bone-like hardness in consistency and the overlying mucosa was normal in colour. Her lower left premolar and molar teeth were missing, and a few fistulous openings with purulent drainage were formed on the gingiva of the lower left molar region. Radiographs revealed numerous radiopaque lesions scattered throughout the mandible and maxilla and impaction of lower left premolar and molar teeth (Fig. 1). A provisional diagnosis of multiple osteomas of jaw bones was made, and Gardner's syndrome was assumed to be present in view of such diagnosis. Roentgenograms of bones showed osteomatous radiopaque lesions in the skull and pelvis, and multiple polyposis of the rectum, colon, ileum, jejunum, duodenum and stomach was found by gastointestinal x-ray examination. Biopsy specimens were taken from some of the intestinal polyps; their histopathological diagnosis was tubular adenoma without malignancy. Up to thumb tip-sized fibromatous tumours of the skin were seen in the breast, hypochondrium, umbilical and inguinal regions, and histopathological examination of some of these tumours revealed a well demarcated neoplasm, covered by pigmented squamous epithelium, composed mostly of fibro-connective tissue with no evidence of malignancy. There were non-remarkable changes on investigations of renal and liver function tests, haematological and blood chemi-

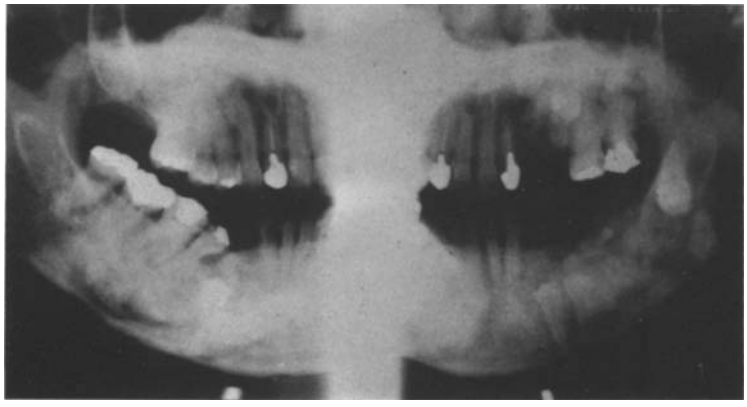

**Fig. 1.** Panoramic x-ray photograph showing multiple radiopaque lesions scattered throughout both mandible and maxilla, and impaction of lower left premolar and molar teeth

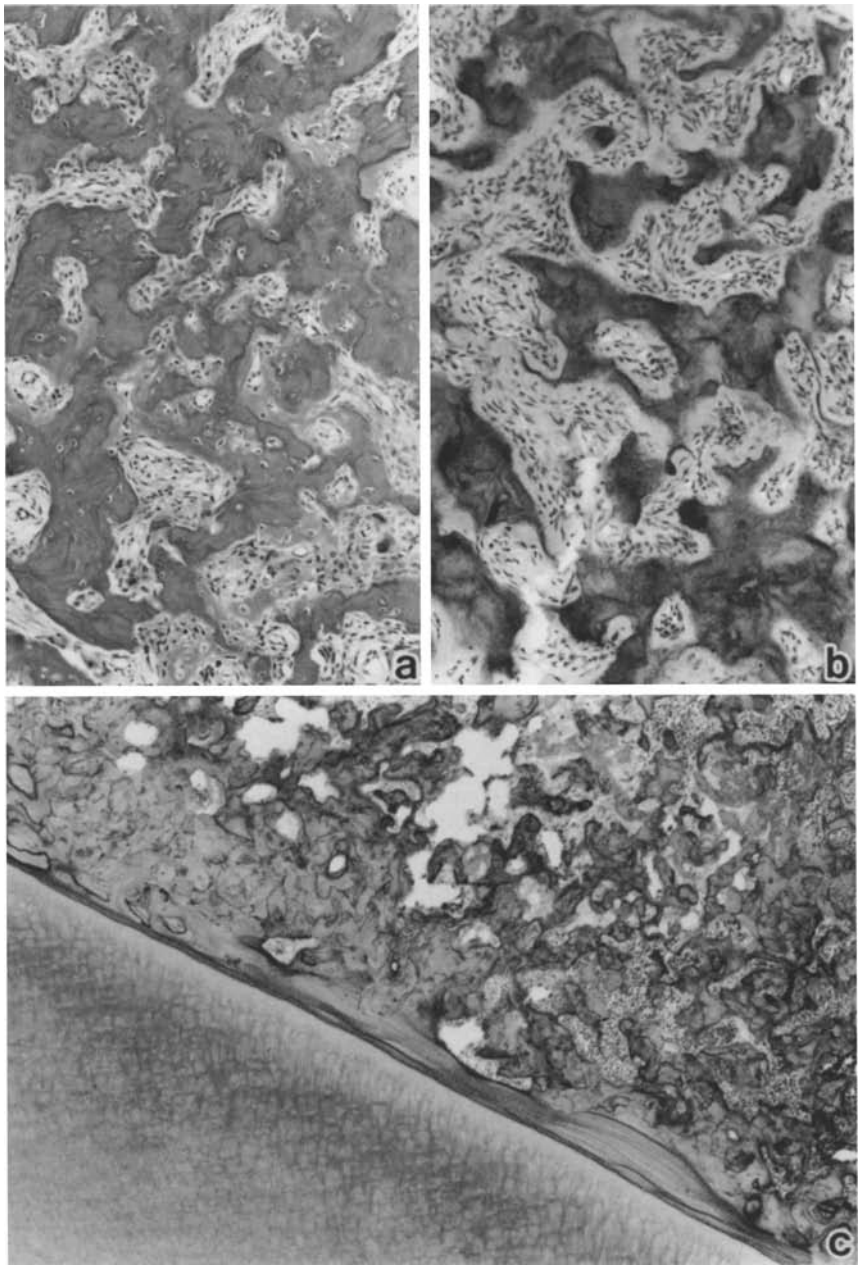


**Fig. 2.** Trabecular proliferation of hard tissues in the mandible, some of them resembling secondary cellular cementum (**a** ×300) but other resembling primary acellular cementum (**b** ×300). Numerous plump mononuclear cells in intertrabecular spaces. Cementum-like trabeculae uniting with the cementum of tooth root (**c** ×40)

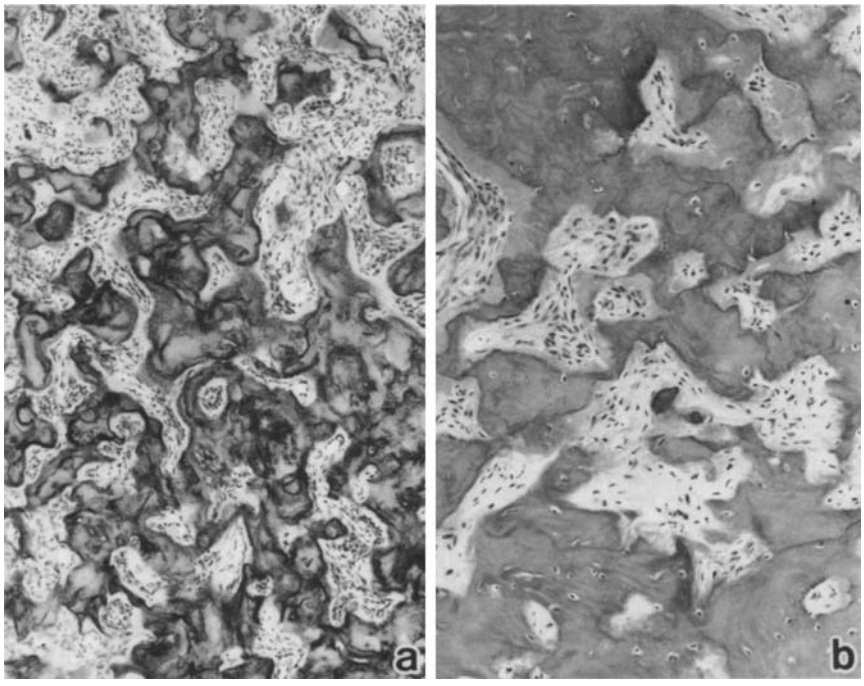


**Fig. 3.** Maxillary lesions showing similar pathological findings to those of mandibular lesions (**a** ×200; **b** ×300)

cal screens and urinalysis, with the exception of elevation of the white blood cell count. From these data the case was diagnosed as Gardner's syndrome.

The patient was treated with antibiotics and symptomatic care and clinical symptom at the lower left molar region regresed. However, suppurative inflammation of the same region with severe pain appeared again on August, 1978, and surgical curettage of the mandibular radiopaque lesions and extractions of impacted lower left permanent teeth were performed. The histopathological diagnosis of the curetted tissue was a cemental lesion.

She was lost to follow-up for the next 8 years, and she returned with diffuse swelling of the maxilla and a pulm-sized abdominal tumour, in May, 1986. Surgical curettage of the maxillary radiopaque lesions and resection of abdominal tumour were performed. The pathological diagnosis of the maxilla was a cemental lesion, and the latter was a desmoid. Furthermore, the colon was removed surgically because of colonic ileus due to polyposis and intestinal continuity was reestablished by an end-to-end ileorectostomy. Histological sections of available material revealed no evidence of malignant transformation of tubular adenoma in any of the polyps.

*Histopathological findings.* The curetted radiopaque lesions of the mandible consisted of trabecular proliferation of basophilic hard tissues which had reversal lines, in a cellular connective tissue stroma (Figs. 2a, b). Trabecular hard tissues showed a cementum-like appearance, some of them resembling secondary cellular cementum (Fig. 2a) but others primary acellular cementum (Fig. 2b). Collagenous fibers from the cementum-like hard tissues streamed into stromal fibrous tissues. The intertrabecular stroma was composed of a large number of plump mononuclear cells, polyhedral in shape and irregular in arrangement, and stromal vascularity was poor. Multinucleated giant cells were few. Cell atypia and mitoses were absent in both the mononuclear plump cells of the stroma and the inclusion cells in the cementum-like trabeculae. Cementum-like trabeculae were found united to the tooth-roots through obliteration of the periodontal ligament, and transition between cementum-like trabeculae and the normal cementum of tooth-roots was evident (Fig. 2c).

In some areas of the lesions, dense irregular or lobulated masses of dense calcification with a few empty lacunae were found. These findings closely resembled those of gigantiform cementoma.

The border of the lesions was distinct, but neither fibrous capusule nor a poorly-mineralized zone were seen between the lesions and the normal bone tissue. Histopathological findings of the maxillary lesions were similar to those of the mandible (Fig. 3).

From these histopathological findings this case was diagnosed as multiple cemental lesions (unusual variant).

## Discussion

Unlike gastrointestinal polyposis, it is thought that the bone lesions in patients with Gardner's syndrome are harmless and are usually of only cosmetic significance. To date, no bone lesions have been shown to undergo malignant transformation with distant metastases (Coli et al. 1970). However, if multiple radiopaque lesions in the jaw bones are found, one should consider the possibility that it is a manifestation of Gardner's syndrome and that the patient should be evaluated for gastrointestinal polyposis (Utsunomiya and Nakamura 1975). Furthermore, multiple bone lesions appear around puberty and precede the appearance of gastrointestinal polyposis (Chang et al. 1968).

Bone lesions of the jaws in Gardner's syndrome are mostly located in the tooth-bearing areas, how-

ever, most of them are entirely separated from tooth-roots. Some of lesions may be found near the tooth-root apex but do not fuse with them. It is well known that such lesions are histopathologically compatible with osteoma, consisting of mature bone tissue with well-developed Haversian systems (Gorlin et al. 1976). However, no other tumours or tumour-like conditions have occurred in a multiple form in the jaw of patients with Gardner's syndrome. The present case showed all the classic manifestations of Gardner's syndrome, i.e., gastrointestinal polyposis, multiple radiopaque lesions in mandible and maxilla, impacted permanent teeth, and multiple cutaneous lesions, but histopathological examination revealed unusual multiple bone lesions in the jaws. These were composed of trabecular calcified tissue with or without cellular inclusions and gradual transition between trabecular calcified tissue and normal cementum of tooth-roots was evident. Furthermore, dense irregular or lobulated masses of highly calcification with few cellular inclusions were seen in part of the lesion. This calcified tissue more closely resembled cementum than immature bone or osteoid and it was thought that the histopathological diagnosis of cemental lesion was appropriate. The diagnostic term, cementoma, has encompassed four distinct entities, namely: benign cementoblastoma (true cementoma), cementifying fibroma, periapical cemental dysplasia (periapical fibrous dysplasia), and gigantiform cementoma (familial multiple cementomas, florid fibro-osseous dysplasia), but it is acknowledged that many cases do not fit accurately into any one of these entities (Pindborg et al. 1971). The present multiple jaw lesions resembled benign cementoblastoma or active stage of periapical cemental dysplasia histopathologically, and had partial resemblance to gigantiform cementoma. However, the radiographic features and histopathological findings indicated that the present lesions did not fit into any one of the distinct entities of cementoma. The present case was finally diagnosed as cemental lesion (unusual type) though such multiple lesions of jaw bones have not been documented previously in patients with Gardner's syndrome. Some of the difficulties in classification are discussed by Waldron et al. (1975) in their report on cemental lesions. These authors would regard these lesions as part of a spectrum of fibro-osseous lesions of peridontal ligament origin.

In general, it is thought that multiple bone lesions of jaws in Gardner's syndrome are pathologically compatible with osteoma. But, in fact, histopathological examination of multiple radiopaque lesions in jaw bones have not been carried out in most of reported cases with Gardner's syndrome, because of the impossibility of obtaining sufficient material from lesions deeply hidden in the jaw bones without extensive damage to an asymptomatic part of the patients. Secondly, the lesions are not subjected to histopathological examination since this could not provide any more conclusive information than that provided by the radiology (Utsunomiya and Nakamura 1975). However, pathological findings in the present case suggest that various types of fibro-osseous lesions may be found in jaw bones by further detailed pathological examinations of multiple radiopaque lesions in Gardner's syndrome.

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