原文題目(出處)：	Oral and maxillofacial manifestations of familial adenomatous polyposis (Gardner's syndrome): A report of two cases. J Contemp Dent Pract 2009 10;1:82-90
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報告日期：	98/04/10

內文：

Aim:

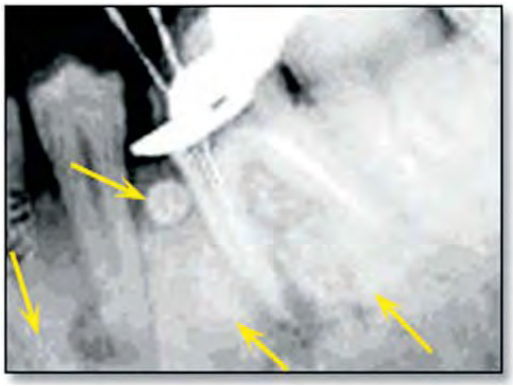
- Chimenos-Kustner et al.and Wijn et al: the importance of an early diagnosis of Gardner's syndrome through the detection of lesions appearing in the oral and maxillofacial area.

Introduction:

- familial adenomatous polyposis (FAP) and Gardner's syndrome were considered as a single condition and classified under a single OMIM number (OMIM 175000) which describes Gardner's syndrome as a variant of FAP.
- a family with intestinal polyposis and multiple bony and soft tissue tumors which were subsequently characterized as multiple osteomas, cutaneous and subcutaneous fibromas, and epidermoid cysts.
- The triad of osteomas, soft tissue lesions, and FAP has now been expanded to include other lesions, including desmoids tumors, retinal pigmentation, carcinomas, sebaceous cysts, gastric and upper intestinal adenomas, pancreaticobiliary adenomas, hepatoblastomas, and dental abnormalities.
- Turcot et al.: malignant central nervous system tumors were associated with FAP
- Oral and maxillofacial symptoms of FAP include an increased risk of osteomas in the jaws, odontomas, supernumerary, or unerupted teeth.
- The oral and maxillofacial hallmark of Gardner's syndrome is the osteoma. (mandible, but they may occur in the skull, long bones, and paranasal sinus cavities.)
- Takeuchi et al.: oral and maxillofacial lesions in 22 of 23 patients with Gardner's syndrome.

Case Report: (CASE 1)

- A 25-year-old Caucasian man was referred by an endodontist for evaluation of a dental abscess in tooth #47 that had been treated.

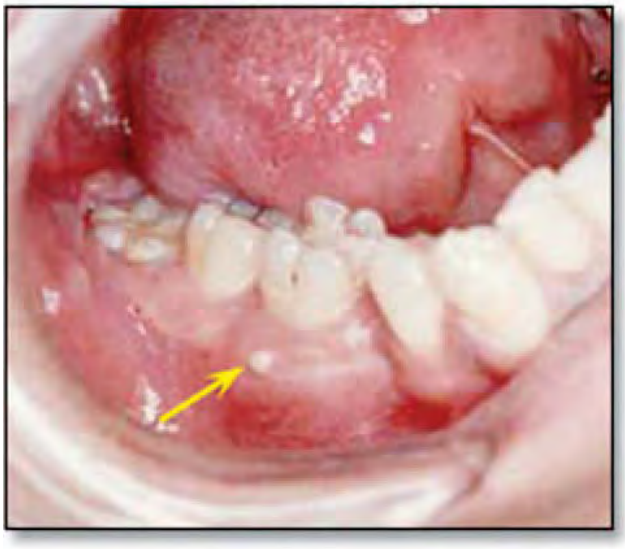


- P.I.:
  - several previous surgeries for removal of "sebaceous cysts" in his body.
  - suffered from occasional rectal bleeding and mucus discharge with

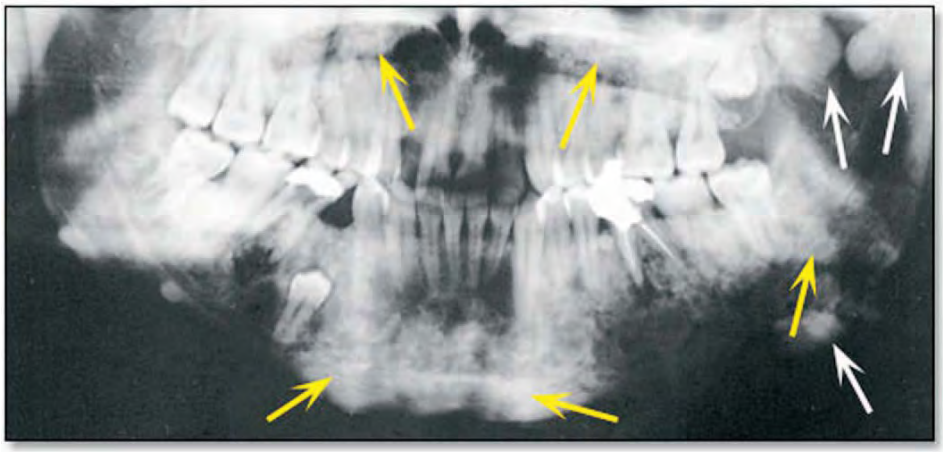
defecation.

- Family History:
  - father and brother had died “with bowel cancer.”
  - 12-year-old niece was presenting several symptoms of Gardner’s syndrome
- P.E.:
  - A 3 cm mobile subcutaneous nodule on his scalp

  - Multiple painless hard masses on the mandible and skull were present. The largest palpable hard mass was located on the left mandibular angle.
  - various submucosal hard masses, mainly in the mandibular external cortex. A small round pearllike structure was observed in the right molar region.



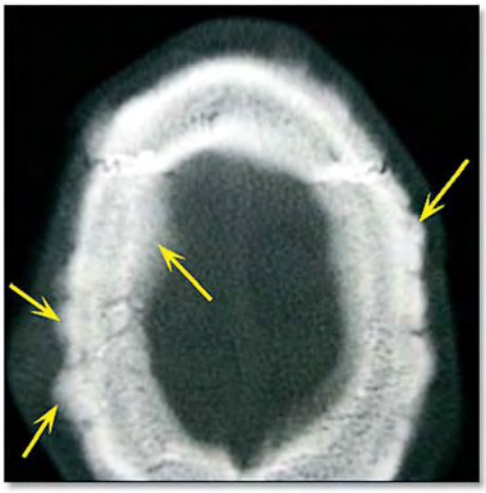
- Panoramic radiograph findings:

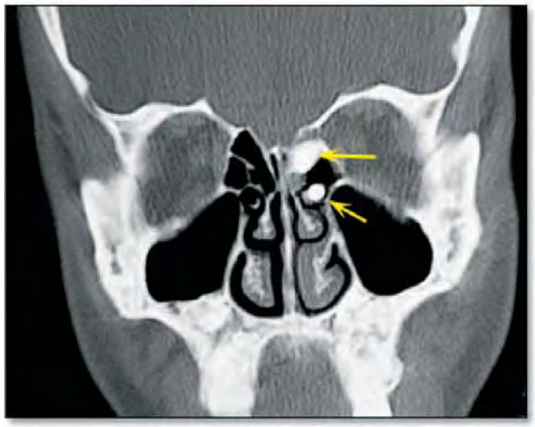


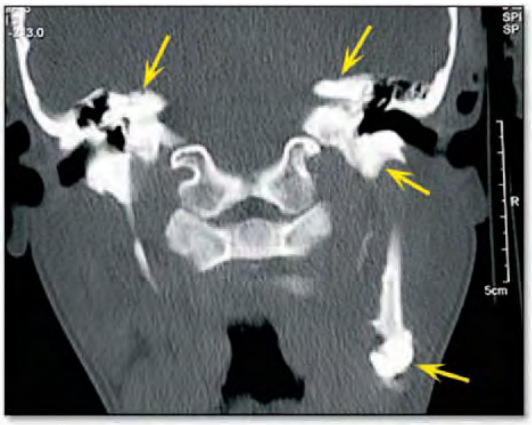
  - Cotton wool-like radiopaque images in both the maxilla and mandible and

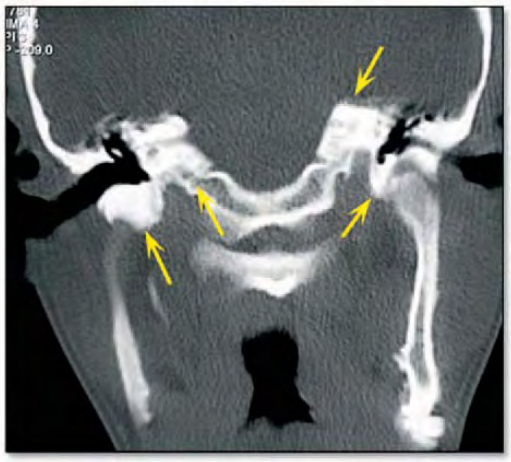
maxillary sinus (yellow arrows). Round or ovoid dense radiopaque images were in the temporomandibular areas and in the left mandibular angle (white arrows).

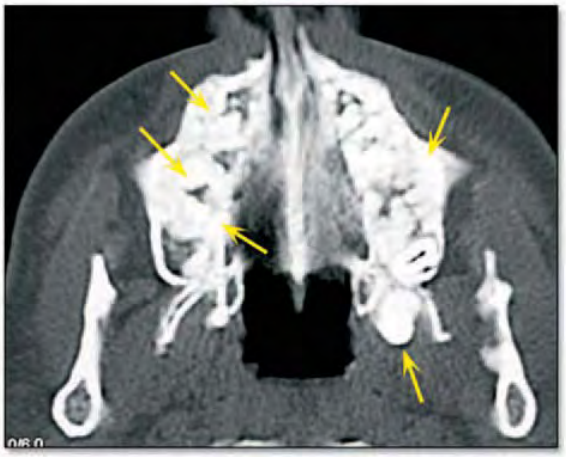
- Computed tomography:







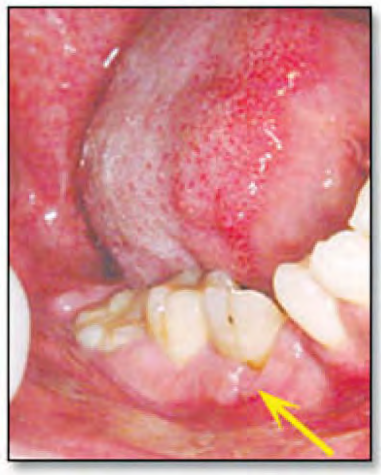


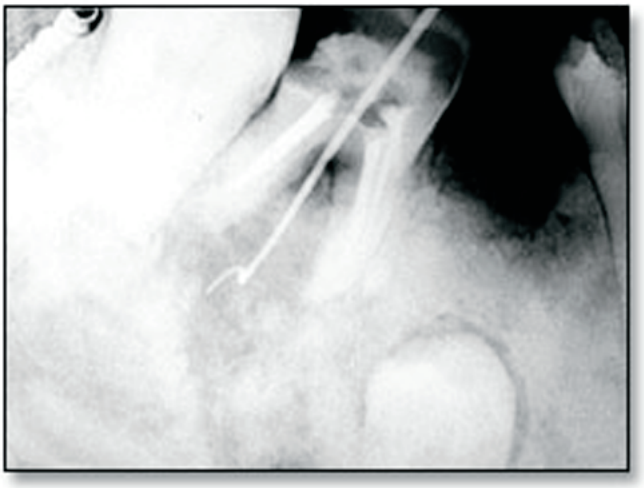


- Several well circumscribed radiopaque area were detected in the ethmoid region and in the nasal walls near the upper conchae and zigomatic areas, sphenoid bone, skull base, and bilateral temporomandibular joint condyles and the maxilla.
- Colonoscopy examination:
  - Numerous polyps carpeting the colon but the rectum was relatively polyp free. A small ulcerating mass was found in the distal portion of the colon.
- Histologic examination:
  - Adenocarcinoma --A colectomy and an ileorectal anastomosis were

performed by general surgeons.

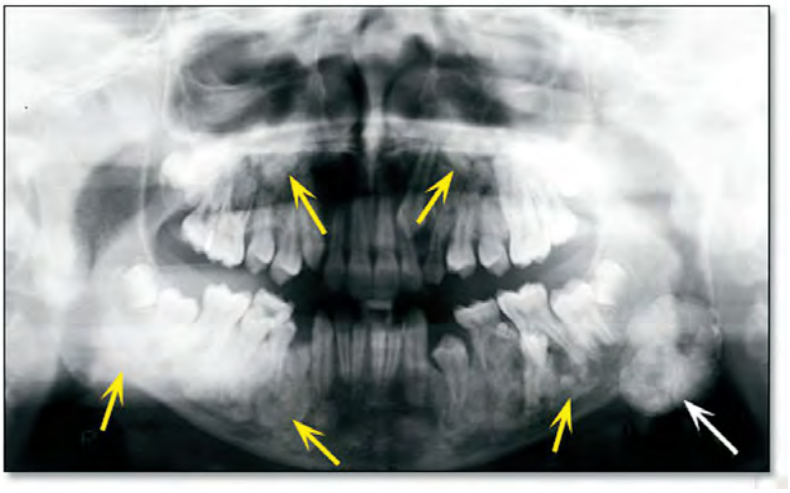
- Microscopic examination of the colon confirmed more than 50 adenomatous polyps, none of which had undergone malignant change.
- Patient was proposed to surgically remove some of the bony masses of the mandible-- refused.
- Two years later, a spontaneous elimination of a 3 x 5 mm bony fragment from the right mandibular molar region, resulting in a periodontal pocket in the first molar.
  - Histologically, the specimen showed trabeculae of lamellar bone with a fibrofatty marrow, confirming the diagnostic of peripheral osteoma.

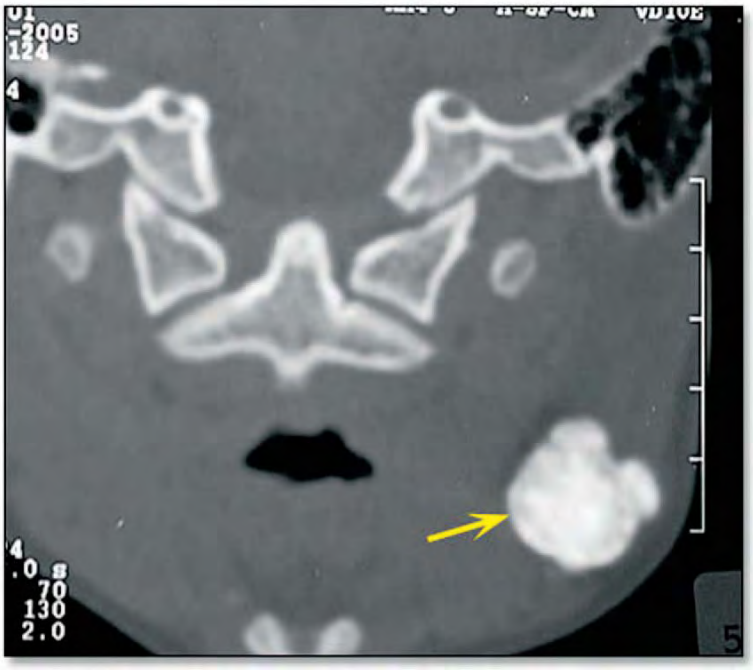




(CASE 2):

- A 12-year-old Caucasian girl presented for consultation with a history of progressive difficulty with mouth opening but no complaints of digestive problems.
  - A panoramic radiograph showed multiple radiopacities in the maxilla and mandible. Some were diffuse (right mandibular body, left and right maxillary tuberosities) with a cotton-wool appearance while others were well circumscribed (left mandibular angle).
  - Multiple impacted teeth were also present.
  - Extensive tumor located in the left mandibular angle.





- A colonoscopy was suggested, but the patient's parents refused.

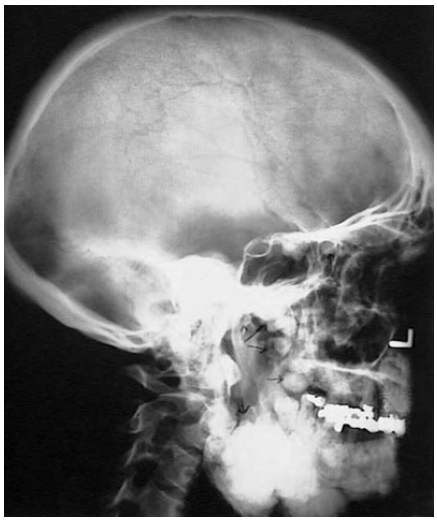
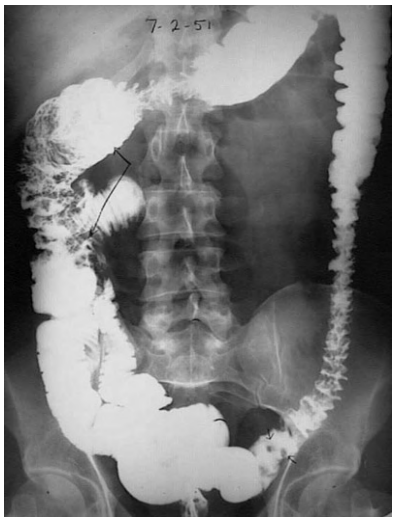
Discussion:

- FAP is the most common polyposis syndrome with a prevalence of about 1 in 8,000.
- In general, the cutaneous and bone abnormalities develop approximately 10 years prior to polyposis.
- Polyp formation starts at puberty but diagnosis is usually in the third decade.
- Unless surgical transaction is performed, gastrointestinal polyps may progress to malignancy in almost 100% of patients (rates vary from 58%-100%).
- Progression to malignancy is usually observed in patients aged 30 to 50 years. The average age by which malignancy is diagnosed is 39.2 years.

Summary:

- Since osteoma formation (the most common accompanying bone lesion seen in Gardner's syndrome) and other maxillofacial abnormalities usually precede polyposis, the general dental practitioner may be the first healthcare professional to suspect the diagnosis.
- Panoramic radiography is the most useful technique for screening patients and affected family members since it can demonstrate subtle opacities at an early age.
- All first and second degree relatives of patients with FAP should be evaluated for the possibility of Gardner's syndrome because an early diagnosis of this disease can lead to a better prognosis for the patient.

題號	題目
1	下列症候群中，何者易出現骨瘤(osteoma)和多生牙(hyperdontia)的情形? (A) 黎蓋氏症候群 (Riegel syndrome) (B) 戈林症候群 (Gorlin syndrome) (C) 外胚層發育異常 (ectodermal dysplasia) (D) 嘉德耐氏症候群 (Gardner's syndrome)
答案(D)	出處：96年第一次高等考試 基礎(二)_3 A:facial bone defect, frontal bone發育不足，上顎前牙先天性缺牙，角

	膜和虹膜發育不全 B:自體顯性遺傳疾病，odontogenic keratocyst, 異位性的大腦鐮鈣化，分岔的肋骨，basal cell carcinoma C:缺牙，無汗症，毛髮稀少 D: 自體顯性遺傳疾病，家族遺傳性結腸息肉，骨瘤，supernumerary teeth
題號	題目
2	一28歲男性因牙齒擁擠欲接受矯正治療，在資料收集X-ray分析中發現病人多處呈現irregular 的dense radiopaque，亦發現阻生牙的情形。經過詳細檢查，也發現multiple colonic adenomas。請問最有可能的診斷為?   (A) Cleidocranial dysplasia (B) Paget's disease (C) Gardner's syndrome (D) Florid cemento-osseous dysplasia
答案(C)	出處：http://library.med.utah.edu/WebPath/COW/COW032.html