

原文題目(出處)：	Gardener syndrome: A rare case report. Dentistry 2014, 4:5
原文作者姓名：	Arora M, Deora SS, Arora P, Saluja
通訊作者學校：	SGT Dental College and Hospital, Gurgaon, India
報告者姓名(組別)：	陳育苹 (Intern A 組)
報告日期：	103.8.12

內文：

Introduction

1. First described in 1951, Gardner's syndrome is a fairly rare condition sharing similarity with polyposis(息肉) coli
2. Gardner syndrome (GS) is a group of diseases
 - (1) **Familial adenomatous polyposis** :
 - A. presence of large numbers of adenomatous polyps(**20 decades**) in the colon with a marked tendency to malignant transformation(**40 decades**)
 - B. APC (Adenomatous Polyposis Coli) gene, located on chromosome 5 (30% of individuals with FAC have a new dominant mutation)
 - C. Early diagnosis in at-risk individuals is usually performed by colonoscopy for detection of intestinal polyposis
 - (2) osteoma :
 - A. is necessary for the diagnosis of Gardner syndrome
 - B. appears in : mandible, skull, long bone, paranasal sinus
 - C. Osteomas usually precede the intestinal polyposis by often as much as 10 years
 - (3) ocular lesions (90%)
 - (4) several dental abnormalities
 - A. odontoma
 - B. supernumerary teeth
 - C. impacted teeth
 - (5) abdominal desmoid tumors(10% in all,3 times more frequent in female)
 - (6) epidermoid cyst of skin

→**FAC with osteomas and benign tumors of the soft tissues is referred to as Gardner's syndrome**
3. Prevalence : varies from 1: 8,300 to 1: 16,000 live births in varying literatures (不分種族)
4. Early identification and surgical intervention of the disease are important to prolong the life of the patient

Case report

A **64 year** old patient

1. C.C. : Swelling on the right lower part of face for past 6 years and in front of right ear region for past 3 months
2. History :

Swelling had **gradually increased** from small in size to attain the present size. The swelling was **painless** upon palpation and **not difficulty in mouth opening**. Patient also complained of another swelling in **right temple region** 請看下方 A 圖



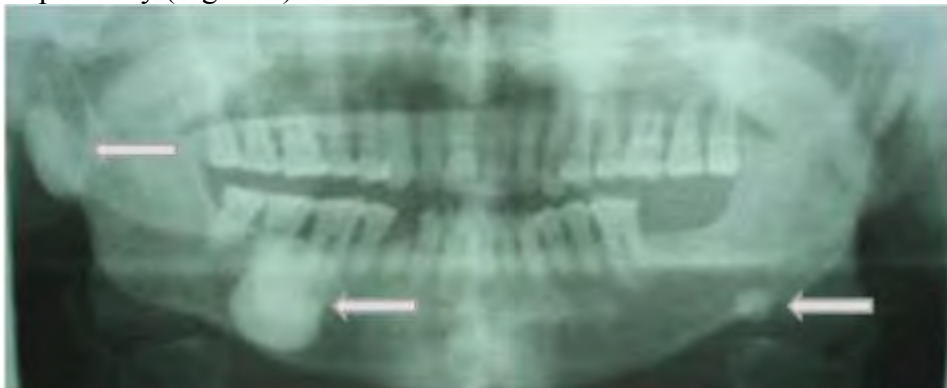
3. Intra Oral Examination :

Revealed two oval well circumscribed swellings measuring approximately 2.5 x 2.5cms and approximately 1.5 x 1.5 cms, seen on the **buccal aspect of 46, 47 and 35, 36 with obliteration of the buccal sulcus**. Overlying mucosa was normal in color and texture 請看上方 B 圖

4. X-ray findings :

Pano

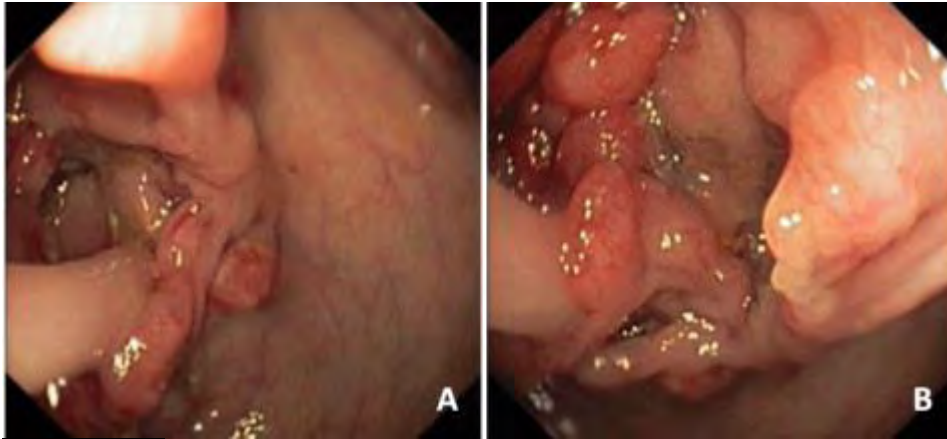
There is **three well defined** dense **radio- opaque masses at right condyle region, right body of the mandible and in the region of lower border of the mandible on left side**, measuring approximately 2 x 1.5 cms, 2.5 x 2 cms, and 0.3 x 0.3 cms respectively (Figure 2)



Water's view, obtained to inspect other facial regions, revealed a **radio-opaque mass in the left frontal sinus** measuring approximately 0.2 x 0.2 cms (Figure 3)



5. Colonoscopy showed **sessile polyps** in the **rectum and descending colon** (Figure 4a and 4b). **Biopsy revealed benign growths and no features of malignancy**



Discussion

1. Familial adenomatous polyposis (FAP) is an autosomal dominant colon cancer syndrome characterized by the presence of polyps in the colon and rectum
2. Gardner syndrome (GS) is a variant of FAP showing extracolonic manifestations such as **osteomas**, **dental anomalies**, **epidermoid cysts** and congenital hypertrophy of the **retinal pigmented epithelium**
3. The intestinal polyps carry as high as **100% potential of malignant change**, and early diagnosis is therefore essential
 → **Several of these extra-colonic manifestations occur in the oral and maxillofacial region precede the colonic polyps and may be discovered during routine dental examination**
4. Dental abnormalities :
 - (1) **Tooth extraction is likely difficult**
 - A. thickening of interdental cortical bone
 - B. extensive hypercementosis
 - (2) multiple impacted permanent teeth, supernumerary teeth and root anomalies
 - (3) Odontomas and cementomas are also frequent
 → as seen in our case at mandible regions and also in frontal sinus
5. Skeletal abnormalities
 - (1) Osteomas :
 - A. slow, continuous growth, and occur most frequently in the mandible, the outer cortex of the skull and the paranasal sinuses
 - B. peripheral exostoses are detectable only radiographically
 - C. two type radiography

localized radiopaque lesion with a sharp border
a large and diffuse radiopaque cotton-wool-like area
widespread radiopaque lesion (in either jaw)
 - (2) mandible angle → facial deformity
 → as seen in our case localized radiopaque lesion with a sharp border
6. The limitation mouth opening caused by
 - (1) **osteoma of the condyle**
 - (2) maxilla in close proximity to the ascending ramus, the external body of the mandible, the coronoid process, and the inferior border of zygomatic arch
 → no seen in our case
7. **Epidermoid cysts**
 - (1) precede the intestinal polyposis
 - (2) multiple and asymptomatic
 - (3) limbs, face, and scalp

- no seen in our case
8. Congenital hypertrophy of the retinal-pigmented epithelium (CHRPE) is reported in 80% of patients with GS and occurs early after birth so it **may represent the first detected sign of the syndrome**
- no mention in our case
9. Desmoid tumours
- (1) histologically benign
 - (2) may behave aggressively and can cause death
- no found in our case
10. neoplasms :
- hepatocellular carcinoma, desmoid tumor, fibromas, lipomas, leiomyomas, neurofibromata, and pigmented skin lesions
- no found in our case
11. In addition, several studies have reported an association between GS and **endocrine disorders** like Cushing's syndrome, thyroid gland nodules, craniopharyngioma, and thyroid cancer
12. **Prophylactic colectomy** is usually recommended. (良性時)
13. Removal of jaw osteomas and epidermoid cysts for cosmetic reasons sometimes may be indicated, **but the long term prognosis depends on the behavior of the bowl adenocarcinomas** (惡性變化時)

1	題目
1	Gardner syndrome 的 clinical finding 下列何者不屬之? (A) Supernumerary teeth (B) Colonic polyps (C) Oral ulcer (D) Osteoma over mandible angle
答案(C)	出處：Oral and Maxillofacial PATHOLOGY,3rd edition , Neville, et al P.651
2	題目
2	Gardner syndrome 的 osteoma 可以在以下哪些部位發現? (A) Long bone (B) Mandible (C) Maxillary sinus (D) Skull bone
答案(A)	出處：Oral and Maxillofacial PATHOLOGY,3rd edition , Neville, et al P.651