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內文：

1. Introduction

·Gorlin-Goltz syndrome is an infrequent multisystemic disease
→inherited in a dominant autosomal way
·1894, this syndrome first descriptions
→the presence：multiple basocellular carcinomas

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1960：Gorlin and Goltz established characterizes of this syndrome
→multiple basocellular epitheliomas,
keratocysts in the jaws,
bifid ribs
→associated with a spectrum of other
neurological, ophthalmic,
endocrine, and genital manifestations

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in general population：this syndrome is estimated to be 1 in 50,000 to 150,000
Males and females are equally affected

2. Case Report：

Patient ：25 years old /female
Chief complaint ：Swelling in bilateral cheeks
Duration ：10 month and the growth was slow
Past medical history ：She was 12 years old
→similar bilateral swellings
→had underwent surgery

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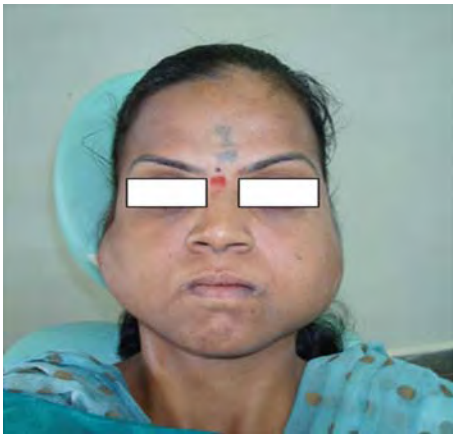
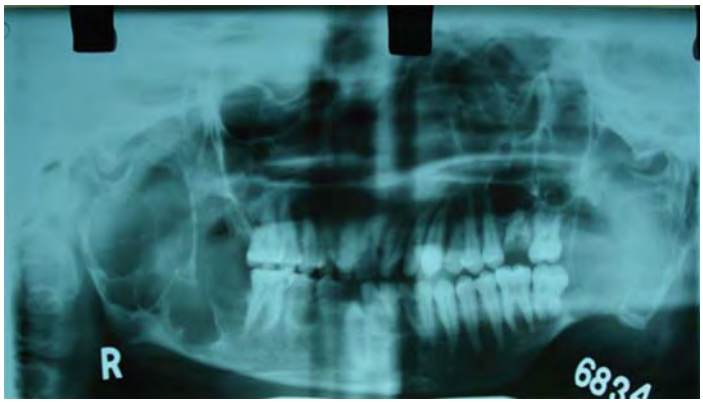
On examination ： The swelling was firm and slightly tender on right side
→face showed frontal bossing, broad nasal bridge, hypertelorism, and mandibular prognathism



Orthopantomograph：Multiple multilocular well-defined radiolucencies with sclerotic border located in maxilla and mandible



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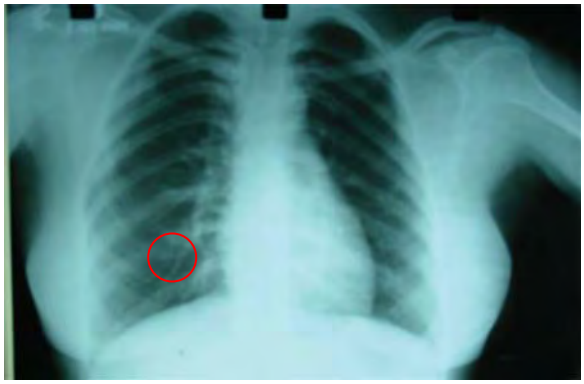
Suspicion of Gorlin syndrome

→ So, other relevant investigations were done

→Chest radiograph ：showed a bifid fifth rib



Biopsy---location：the swelling in left and right side of mandible

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↓

Histopathological examination ： parakeratinised stratified squamous epithelium palisading pattern of columnar cells few giant cells inflammatory cells (found in the specimen of the left side of the mandible)

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↓

Suggestive right side：odontogenic keratocyst
left side：odontogenic keratocyst with secondary infection

↓

↓

Surgical enucleation was performed
→skin lesions：not seen like basal cell nevus or keratosis
the parents of the patient：showed no signs of Gorlin-Goltz syndrome
But the patient was diagnosed as having Gorlin-Goltz syndrome
←the clinical, radiographic, and histologic findings
the diagnostic criteria for nevoid BCC syndrome
→established by Evans et al
modified by Kimonos et al. in 1997

3. Discussion

The Gorlin-Goltz syndrome：an autosomal dominant inherited syndrome
→multiple defects involving the skin,
nervous system, eyes,
endocrine system, and bones
=Basal cell nevus syndrome
=Multiple basal cell carcinoma syndrome
=Gorlin syndrome
=hereditary cutaneomandibular polyonocosis
=multiple nevoid basal cell epithelioma-jaw cysts
=bifid rib syndrome

→ → → →The diagnostic criteria for nevoid BCC
→ established by Evans et al., and modified by Kimonis et al

Major Criteria are as follows:

(1) more than 2 BCCs or one under age of 20 year,
(2) odontogenic keratocyst,
(3) three or more palmar pits,
(4) bilamellar calcification of falx cerebri,
(5) bifid, fused, or splayed ribs,
(6) first-degree relative with NBCCS.

Minor Criteria are as follows:

(1) macrocephaly adjusted for height,
(2) fontal bossing, cleft lip/palate, and hypertelorism,
(3) sprengel deformity, pectus, and syndactyly of digits,
(4) bridging of sella turcica, hemivertebrae, and flameshaped radiolucencies,
(5) ovarian fibroma,
(6) medulloblastoma

→Two major or one major and two minor are present
→diagnosis of Gorlin-Goltz syndrome

Odontogenic Keratocysts

Two types：

Parakeratotic odontogenic keratocyst (P-OKC)…more common
Orthokeratotic odontogenic keratocyst (O-OKC)…less common
↓
P-OKC：more aggressive growth potential and
a higher recurrence
rate than the O-OKC and other odontogenic cysts
→P-OKC is a part of the Jaw cyst-Basal cell nevus-Bifid rib syndrome
→Gorlin syndrome
→benign, the recurrence rate：high
ranging from 12% to 62.5%

↓

Before concluding：detailed examination and X-ray investigation of the relatives should be undertaken

EX. Table 1: Diagnostic protocols in NBCCS.

Family history
Past medical and dental history
Clinical examinations
Oral
Skin
Central nervous system
Head circumference
Interpupillar distance
Eyes
Genitourinary system
Cardiovascular system
Respiratory system
Skeletal system
Genetic testing
X-ray
Chest
A.P. and lateral skull
Panoramic radiograph
Cervical and thoracic spine
Hands (for pseudocysts)
Pelvic (female)

4. Conclusion

Gorlin-Goltz syndrome
-its malignant potential
→In order to be able to establish early diagnosis of NBCCS,
→specialists should carry out clinical and imaging examinations

題號	題目
1	以下何者最不可能是 Gorlin syndrom 的特徵？ (A) Odontogenic keratocyst (B) Basal cell carcinoma of the skin (C) Calcification of falx cerebri (D) Café au lait spots
答案(D)	出處：Oral and Maxillofacial pathology,2nd,Nevielle,p598,599
題號	題目
2	與 Gorlin syndrom 最有相關的惡性腫瘤是以下何者？ (A) Verrucous cacinoma (B) 分化良好的 Squamous cell carcinoma (C) Basal cell carcinoma (D) Lymphoma
答案(C)	出處：Oral and Maxillofacial pathology,2nd,Nevielle,p598