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內文：

### INTRODUCTION

- Odontogenic keratocyst (OKC) is a developmental odontogenic cysts
- originate from the dental lamina.
- first noted in 1956 by Phillipson
- OKC is well recognized by its aggressive behavior, rapid growth and high tendency to invade the adjacent tissues including bone.
- It tends to recur and occasionally is associated with the basal cell nevus syndrome.
- odontogenic keratocyst (OKC) is characterized by specific histopathologic features and clinical behavior which is more suggestive of tumor rather than cyst. Several investigators believe that it must be classified as a benign cystic neoplasm.
- In the latest WHO classification of odontogenic tumors in 2005, these lesions have been given the name “Keratocystic Odontogenic Tumors” (KCOTs)
- Multiple KCOTs are usually seen with cutaneous, skeletal, ocular and neurologic abnormalities as a component of Gorlin-Goltz syndrome

### ASSOCIATION WITH GORLIN-GOLLTZ SYNDROME

- rare multi-system disease
- characterized by neoplasms and other 發育異常
- 體染色體顯性遺傳 and caused by abnormalities in the PTCH1 (Patched1) gene which is traced to the long arm of chromosome 9q22.3-q31
- different name throughout the time:
  1. nevoid basal cell carcinoma syndrome
  2. Gorlin syndrome
  3. fifth phacomatosis
  4. multiple basilioma syndrome
  5. hereditary cutaneomandibular polyoncosis
  6. ‘Jaw-cyst-basal-cell-nevus, bifid rib syndrome
- Diagnosis of Gorlin-Goltz syndrome:
  - two major criteria
  - one major and two minor criteria
- The major criteria includes:
  1. odontogenic keratocysts of the jaws
  2. more than two sites of basal cell carcinomas or one site of basal cell carcinoma in persons younger than 20 years
  3. three or more palmar or plantar pits(手掌及腳底出現小凹陷)
  4. calcification of falx cerebri(大腦廉)
  5. bifid(分岔) or fused ribs
- The incidence of this disorder lies in the range of 50,000 to 150,000 in the general population, 受區域分配影響.好發率 female=male.
- The age of diagnosis of NBCCS(nevoid base cell carcinoma syndrome) is usually 13 years while average age for basal cell carcinoma syndrome is 20 years

-Gorlin-Goltz syndrome 的臨床表現是 different from individuals within the same family

-如果出現 Multiple keratocysts 就應該要特別注意是否有這個 syndrome 的 possibility

#### THE MANAGEMENT AND PROGNOSIS OF KCOT IN CHILDREN

-review 目的:發現是否有關於 KCOT in children 的預後及行為的具體特徵

-Odontogenic cysts have low prevalence in children

- KCOT usually appears in the second, third and fourth decades(10-40 歲) of life (54.2%) and rare cases reported as early as the first(10 歲), and as late as the ninth decade(90 歲) of life

-Because of their great tendency to recur(高復發率), KCOTs 的 tx 仍然是爭議性的議題

-decompression(減壓) or 造袋術(marsupialization) are the first surgical manipulations in KCOT

-when the volume becomes smaller in size, enucleation has to be performed

#### Case 1

-Ravi et al revealed a case of 11 years old boy which was diagnosed as Gorlin-Goltz syndrome according to 臨床病史, radiographic imaging and 組織病理.

-The cyst was surgically enucleated with good prognosis.

#### Case 2

-Maria et al reported a case of a 10-year old boy with KCOT on the right side of mandible

-為了保留恆牙, the patient was submitted to 透過 extraction of the primary molars and insertion of iodoform gauze 來達到 decompression procedure

-The permanent teeth went to natural eruption and 2 years after the first visit lesion had healed.

-Patient was referred to orthodontic treatment and radiographic follow-up of 7 years showed no lesion recurrence

-He concluded, 特別是在 young p't 的 the treatment option should consider 保守治療 with low morbidity(發病率)

#### Case 3

-Vijay reported a case of a 6-years old boy KCOT in left posterior side of mandible

- tumor was surgically enucleated 然後 application of Carnoy's solution for 5 minutes

-good healing and bone regeneration was achieved, the lesion showed no evidence of recurrence during the first year of follow up

-He concluded that, the first priority is to the preservation of adjacent vital structures, 日後才有 less postoperative complications.

-Enucleation with or without chemical cautery using Carnoy's solution followed by iodoform dressing can be considered as a more conservative treatment modality with a low tendency to recurrence ,and fairly good healing for KCOT.

-由於 KCOT 的 high recurrence,也建議要 OP 後要觀察比較長的一段時間.

#### Case 4

-Singh et al. reported a KCOT in an 11-years old female which he treated by surgical enucleation of the cyst, according to the principle of Partsch II

-The cystic linings were removed and curettage was done with Carnoy's solution

-The site was closed with 優碘浸泡過的 gauze pack and their borders were then sutured to create an open cavity that communicates with the oral cavity

#### Case5

-Rakesh et al. reported a case of KCOT associated with Gorlin-Goltz syndrome in a male patient aged 12 years of age.

-Marsupialization followed by enucleation of upper odontogenic keratocyst and 用 surgical bur 及 curette 去刮 bone cavity.

-the cyst was surgically enucleated followed by application of Carnoy's solution as chemical cauterization to reduce the recurrence

#### DISCUSSION

-OKC is regarded as an aggressive lesion due to its characteristic high tendency to recur, and to invade adjacent tissues

-In 1967, Toller suggested that 根據臨床特徵 OKC should be classified as a benign tumor rather than a cyst.

-之後 WHO categorize the lesion as a tumor.

-This decision was based on several factors:

1. Behavior: OKC is locally aggressive and highly recurrent.

2. Histopathology:

-The basal layer of the OKC (KCOT) budding into connective tissue.

-Mitotic figures(核分裂) were commonly seen in the suprabasal layers.

3. Genetics:

-PTCH ('patched')(與 nevoid basal cell carcinoma syndrome 及 sporadic(type) KCOTs 相關的 tumor 抑制器 gene) commonly found on chromosome 9q22.3-q31.36-40 normally

-PTCH 加上 oncogene(致癌基因)SMO ('smoothened')合成 SHH ('sonic hedgehog') ligand(SHH 配體)的 receptor complex

-PTCH binding to SMO 會抑制 growth signal transduction

-而 SHH binding to PTCH 會造成這種 growth signal transduction 的抑制

-如果 PTCH 的功能 lost,那 SMO 的 proliferation-stimulating 效用就會成為主導

-The decision on the treatment option should be established on the size and site of the lesion, recurrence status and cortical 破壞的 x 光判讀及組織的變異

-parakeratotic type is more common in young age

-根據前述特徵,要用 aggressive surgical approaches with complete lesion eradication(根除)

-但 These aggressive 的操作 may lead to deformities, which may lead to 嚴重的心理問題

-因此 reduction of complications as much as possible 是 priority 的

-許多研究指出如果只用 enucleation, KCOT 是容易復發的

-如果加上像是 Peripheral Ostectomy, Carnoy's solution, and Cryotherapy(冷凍治療)是可以降低復發率的

-但這些治療仍有其風險

-例如 Carnoy's solution and other chemical cauterization might cause damage to nearby vital structures such as the IAN and/or permanent tooth buds

-When the cystic lesion 在 vital structures 附近, marsupialization or decompression 為更明智的做法

-但如果 lesions are away from these vital structures, then 5 minutes of application 化學腐蝕劑例如 Carnoy's solution would be more acceptable given its ability to permeate(滲透) and 修復 tissues up to a depth of 1.54 mm into the bone.

-Cryotherapy(冷凍療法)也是個可以達到類似的結果, but may produce complications such as wound dehiscence(裂開)noted in most cases.

-Cases where cysts are large owing to the presence of very thin bony walls Peripheral Osteotomy is impractical(不切實際的)

-enucleation 後伴隨著 a large cavity is more prudent(謹慎) to allow it to heal by secondary intention rather than primary wound closure to avoid the hazards of postoperative infection

-The ideal properties of dressing material used for dressing wounds in the oral cavity should consist of following:

1. 緩解 postoperative pain
2. 提升 healing 能力
3. safeguard against infection

-Iodoform is regarded as a dressing material owing to

1. its capability to reduce wound fluids by Fibrinolytic(纖維蛋白溶解) activity
2. exhibits antimicrobial(抗菌) activity, 也可降低疼痛

-Recently investigators have shown that decompression and marsupialization change the epithelial lining of KCOT into a less aggressive form, and some tumors have undergone complete resolution with these treatment modalities

-Based on the above results, they recommended performing enucleation with or without chemical cauterization utilizing Carnoy's solution followed by iodoform gauze dressing for all cases especially in children

-In children with unerupted teeth, aggressive manipulations endanger the eruption process and the development of the involved jaw

-10 歲前應用 conservative approach

-en bloc resection (較 aggressive)is thought of in the following cases:

1. 先前做過 enucleation 加上其他處置但復發的 cases
2. 先前做過 marsupialization followed by enucleation 但復發的 cases
3. In cases of multiple nonsyndromic or syndromic odontogenic keratocysts of NBCCS(nevoid basal cell carcinoma syndrome)
4. 當被診斷出特別 aggressive 的 clinical behavior 時,需要 radical resection(根治性切除) as the first treatment option

**CONCLUSION**

-Treatment of 全方位治療團隊 is important if associated with Gorlin's syndrome.

-Postoperative follow up is advised every 6 months

題號	題目
1	Karatocystic Odontogenic Tumors(KCOT)好發在哪個年齡? (A) 10 歲以下 (B) 10-40 歲 (C) 50-60 歲 (D) 70 歲以上
答案 (B)	出處 oral and maxillofacial pathology,3 rd edition, p685
題號	題目

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2	下列關於 Keratocystic Odontogenic Tumors(KCOT)的處置,何種最適當?
	(A) Enucleation and curettage (B) decompression (C) marsupialization (D) Peripheral Osteotomy
答案 (A)	出處：oral and maxillofacial pathology,3 rd edition, p686