

# Case Report

報告者 : Intern Group B

指導醫師 : 陳玉昆 主任

林立民 醫師

及口腔病理科全體醫師

2015/09/29



# 工作分配



- ◆ General data: 陳暉翔
- ◆ X-ray finding: 邱仲凡
- ◆ Discussion: 廖信昱
- ◆ 醫學倫理 : 許永瀲
- ◆ PPT製作 : 全體組員
- ◆ 報告 : 全體組員



# General data

- ❖ Name: 陳X
- ❖ Sex: Male
- ❖ Age: 19 y/o
- ❖ Native: 高雄市
- ❖ Marital status: single
- ❖ Attending staff: OOO醫師
- ❖ First visit : 94/11/22



# Chief Complaint

- ❖ Came with his mother, 12 months s/p enucleation of the large cyst, came with his mother for follow-up of his multiple lesions.

# Present Illness

- ◆ This is a 19 year-old male who first visited our out-patient-department in 94/11/22 and was found multiple radiolucences over bimaxillary and arranged operation (biopsy & decompression), and histopathologic report showed keratocystic odontogenic tumor over right mandible; left maxillary & mandible. Then after a period of follow up, the patient came back in 95/02/07 for extraction tooth 83 residual root at Pediatric Dentistry Department

# Present Illness



- ❖ Came back in 95/02/21 for enlargement of right mandible cyst orifice (tooth 83) and obturator fabrication, came back in 95/03/14 to extract tooth 84 for enlarging right mandible fistula, and in 95/05/16 for re-shaping the decompression button (tooth 84), then in 95/07/18 for removing decompression button over left mandibular angle area, and in 95/11/21 for removing right obturator.



# Present Illness

- ❖ Then in 97/01/31, 38 area cystic lesion persisted, we did extraction tooth 53 & 75 as decompression in 97/07/24, and 97/08/14 we found right maxilla keratocystic odontogenic tumor, left mandible odontogenic cyst and tooth 38 area dentigerous cyst. In 97/01/23, the patient got 13 impaction and consult orthodontic department, the doctor of orthodontic department suggested orthodontic treatment right now or until 31 eruption

# Present Illness



- ◆ In 98/07/14, patient was diagnosed of keratocystic odontogenic tumor over left posterior mandible, and we placed decompression button and biopsy. In 99/07/03, the patient had increased size of tooth 38 dentigerous cyst and a biopsy & decompression button was set, also the histopathologic report: infected odontogenic cyst.



# Present Illness

- ❖ Then, in 99/02/01 we did the keratocystic odontogenic tumor enucleation over tooth 42, 43 area, and in 99/07/06, we found new cyst of tooth 23 area but only followed up at OPD. In 100/08/09, we found tooth 38, 48 swelling and 47 pushed posterior and we did decompression & biopsy histopathologic report: inflammatory cell infiltration, then in 100/10/25, we found a new cyst over tooth 18 and kept following up.

# Present Illness



- ◆ In 101/08/27, two compression buttons were observed at left maxilla and cystic lesion at right maxilla so we arranged sedation anesthesia at dental out-patient-department for special need for biopsy and place decompression button in 101/09/27. The operation was at sleeping mode but cough reflex severe, so we placed compression button, penrose and set bacterial culture, also arranged operation (multiple keratocystic odontogenic tumor over

# Present Illness



- ❖ Maxilla & mandible with impaction s/p Debulky with tooth 17, 18 removed + decompression over right maxilla and cyst enucleation with tooth 27, 28, 38, 47, and 48 removed over left maxilla + mandible in 101/10/26. And histopathologic report was hyperparakeratosis of tooth 46 distal, and keratocystic odontogenic tumor of bilateral mandible and maxilla.

# Present Illness



- ❖ This time after the patient has been followed up at out-patient-department for 4 months, we kept monitoring the operation outcome of enucleation of the large cyst over right maxilla. In 102/01/26, restoration of tooth 16 drop, the local-dental-clinic suggested extraction of tooth 16, so the extraction was performed in 102/04/11.

# Present Illness



- ❖ In 102/05/02, we arranged operation in 102/07/03, and the post-operative pathology report: keratocystic odontogenic tumor over right and right posterior medial pterygoid plate, keratocystic odontogenic tumor over right infraorbital and posterior, fibrotic tissue with multiple odontogenic epithelial rest over right mandible.

# Present Illness



- ❖ In 102/12/12, we arrange CT, check BUN, Cr for him, and CT report: 1) Shrinkage yet persistence of odontogenic cyst in the right aspect of the maxilla status post drainage tube insertion. 2) Absence of odontogenic cyst in the left aspect of the maxilla. 3) Mild bilateral ethmoid and left maxillary sinusitis. Non-specific small lymph nodes (<1cm) in the submental, the bilateral submandibular, jugulo-digastric, and the posterior cervical spaces

# Present Illness



- ❖ In 103/02/13, we arrange operation in 103/03/12 for him.
- ❖ Pt was suspected with Gorlin syndrome (Basal cell nevus syndrome). Pt denied the history of drug allgery. Pt denied the oral habits of cigarette smoking, betel quid chewing and alcohol drinking. 病人表示打完造影劑之後會有過敏的反應 病人表示他的頭頂上有長了一顆新的痣 高雄榮總皮膚科意見為觀察 No evidence of active progression



# Personal History

- ❖ Past medical history
  - ❖ Underlying disease: Denied
  - ❖ Hospitalization (+), odontogenic cyst
  - ❖ Surgery under GA (+), odontogenic cyst
  - ❖ Allergy: contrast medium (Ultravist)



# Personal History

- ❖ Past Dental History
  - ❖ General routine dental treatment
- ❖ Attitude to dental treatment : co-operative
- ❖ Risk factors related to malignancy
  - ❖ Alcohol (-)
  - ❖ Betel quid (-)
  - ❖ Cigarette (-)
- ❖ Special oral habits: Denied
- ❖ Irritation: Denied



# X-ray findings

◆ 2015/9/10



# X-ray findings



- ◆ Mandible:

There are two well-defined unilocular round shaped radiolucence with a corticated margin lesions. One is at right posterior region of mandible body and with bone expanding extending to cortical margin, measuring in about 3x3 cm in size. Another one is at right mandible ramus region and extends from retromolar pad region to mandibular canal region, measuring in about 2x1 cm in diameter.

# X-ray findings



## ◆ Maxilla:

There is a well-defined unilocular round shaped radiolucency without a corticated margin over maxilla extending from distal side of lateral incisor to mesial side of canine and from ridge between tooth 13 and 12 up to right maxilla sinus, measuring about 1x3 cm in size.

# X-ray findings



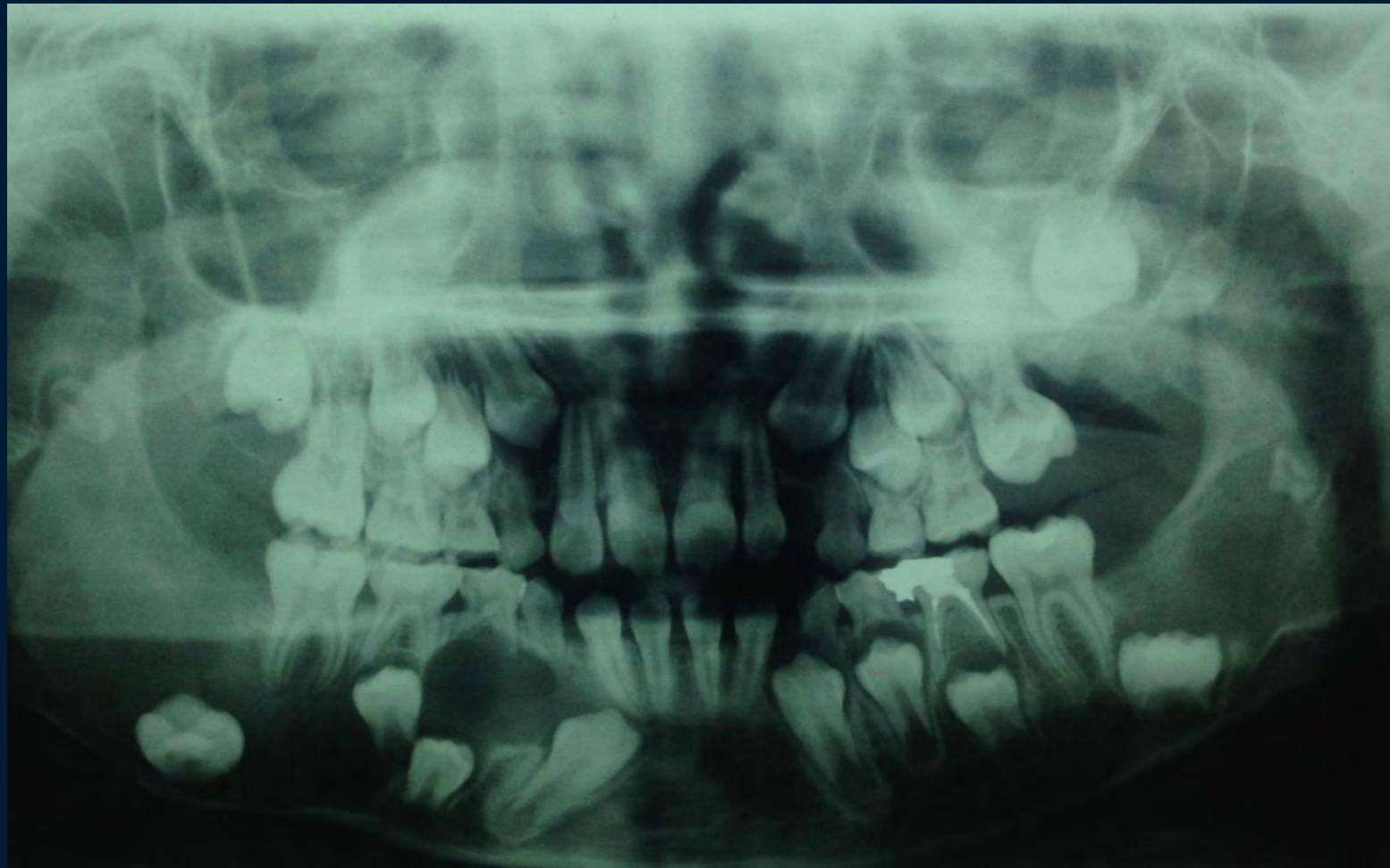
## ❖ Dental findings:

1. Missing tooth: 16, 17, 18, 27, 28, 38, 46, 47, 48
2. Restoration: tooth 36 (MO)
3. Tooth is mesial tilting



# X-ray findings

► 2005/12/13





# X-ray findings

◆ 2006/9/19





# X-ray findings

◆ 2006/11/21





# X-ray findings

◆ 2007/2/13





# X-ray findings

❖ 2007/5/31





# X-ray findings

❖ 2008/1/31





# X-ray findings

❖ 2008/7/24





# X-ray findings

❖ 2009/1/20



Z: 30

# X-ray findings



❖ 2009/7/3



# X-ray findings



◆ 2009/8/18





# X-ray findings

❖ 2010/1/19



# X-ray findings



❖ 2010/4/24





# X-ray findings

❖ 2010/8/17



# X-ray findings



❖ 2011/1/25





# X-ray findings

◆ 2011/8/2





# X-ray findings

◆ 2011/11/25





# X-ray findings

◆ 2012/1/17





# X-ray findings

◆ 2012/8/27



Z:35%



# X-ray findings

◆ 2012/10/27





# X-ray findings

❖ 2012/10/27





# X-ray findings

◆ 2012/12/6





# X-ray findings

◆ 2013/1/17





# X-ray findings

◆ 2013/7/4





# X-ray findings

❖ 2013/10/3





# X-ray findings

◆ 2014/2/6

7/2106





# X-ray findings

❖ 2014/3/13





# X-ray findings

◆ 2014/5/15





# X-ray findings

❖ 2015/2/12



7·35%



# X-ray findings

❖ 2015/9/10



# X-ray findings

❖ 2015/9/10



# X-ray findings

◆ 2015/9/10



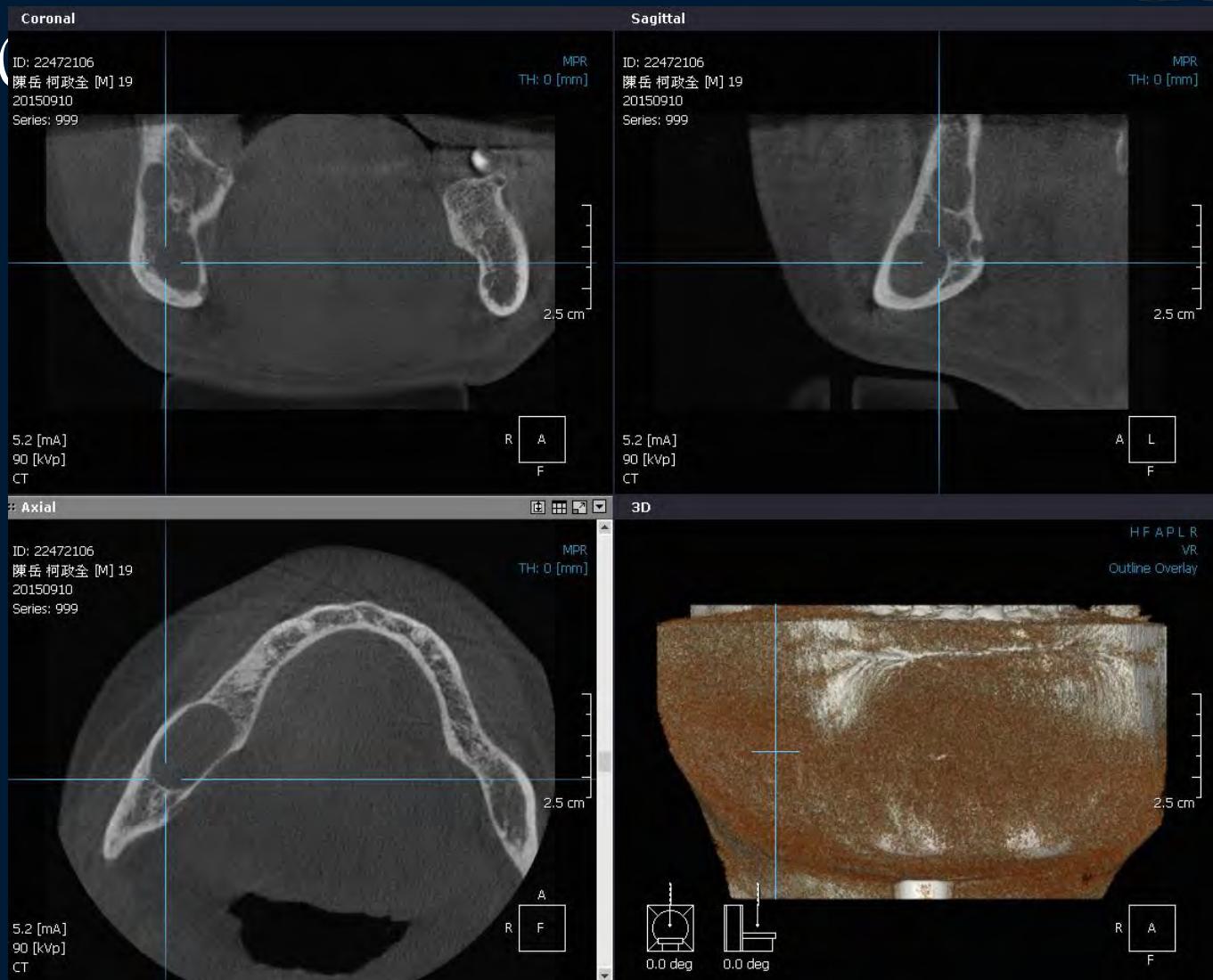
# X-ray findings

◆ 2015/9/10



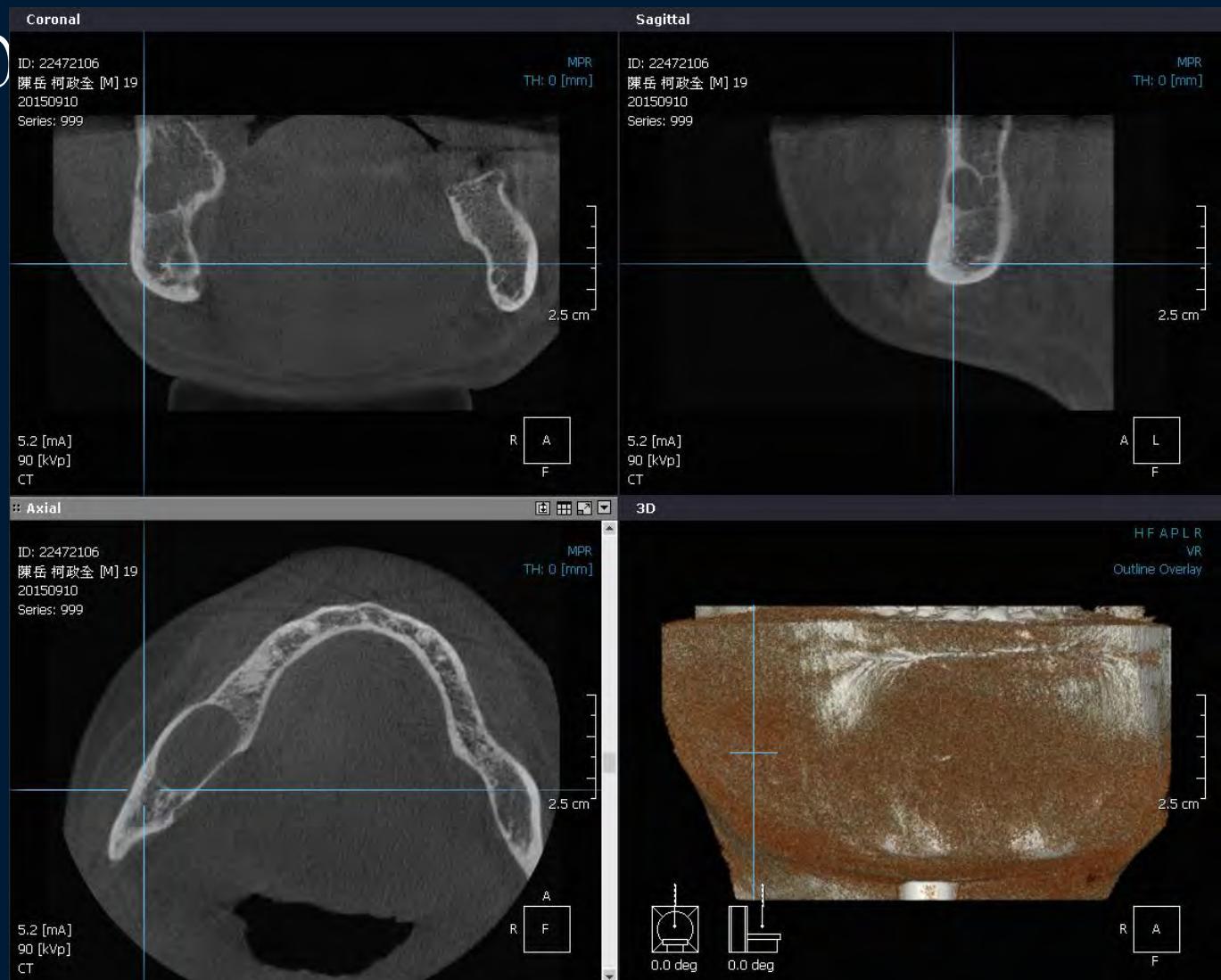
# X-ray findings

❖ 2015/9/10



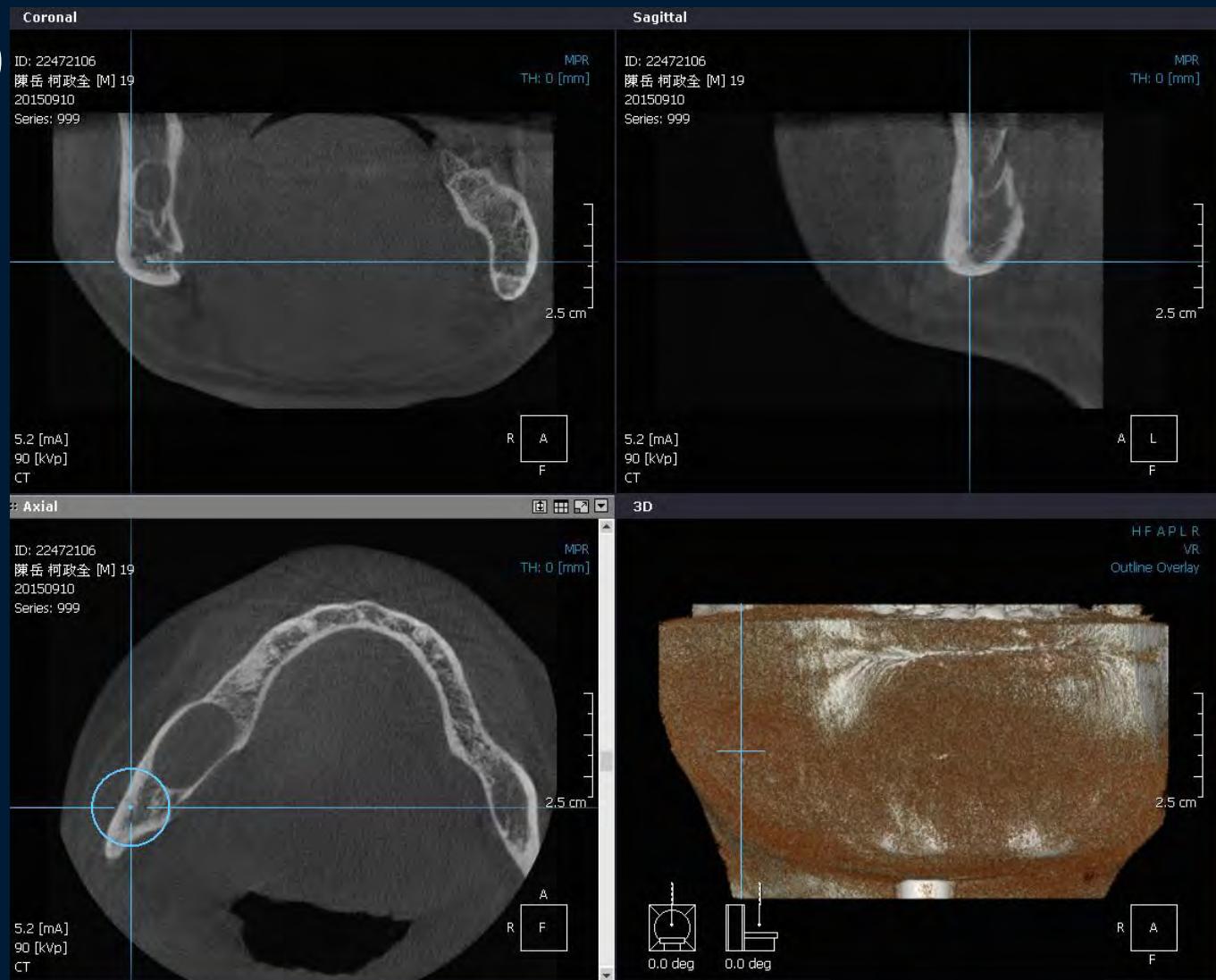
# X-ray findings

❖ 2015/9/10



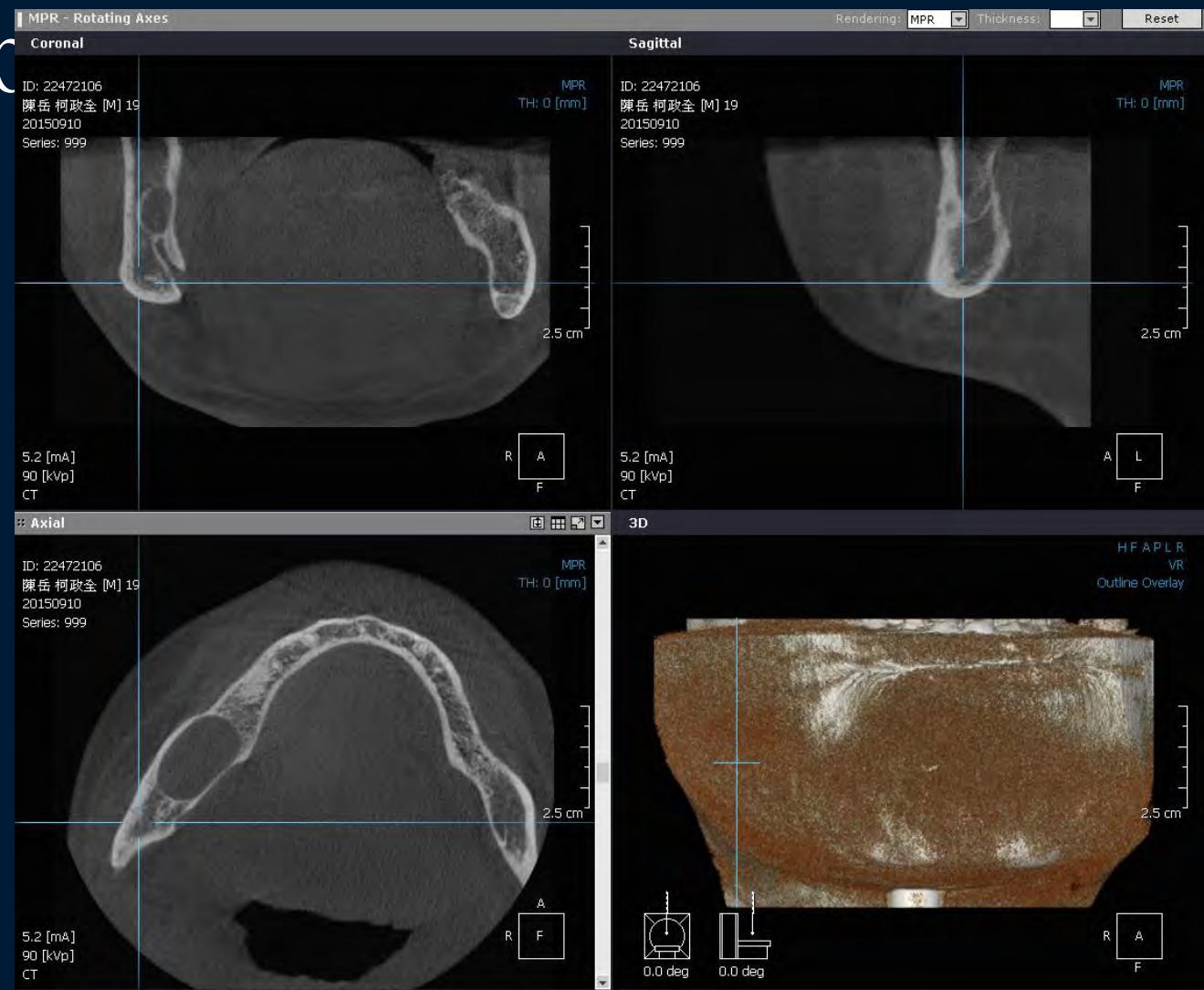
# X-ray findings

❖ 2015/9/10



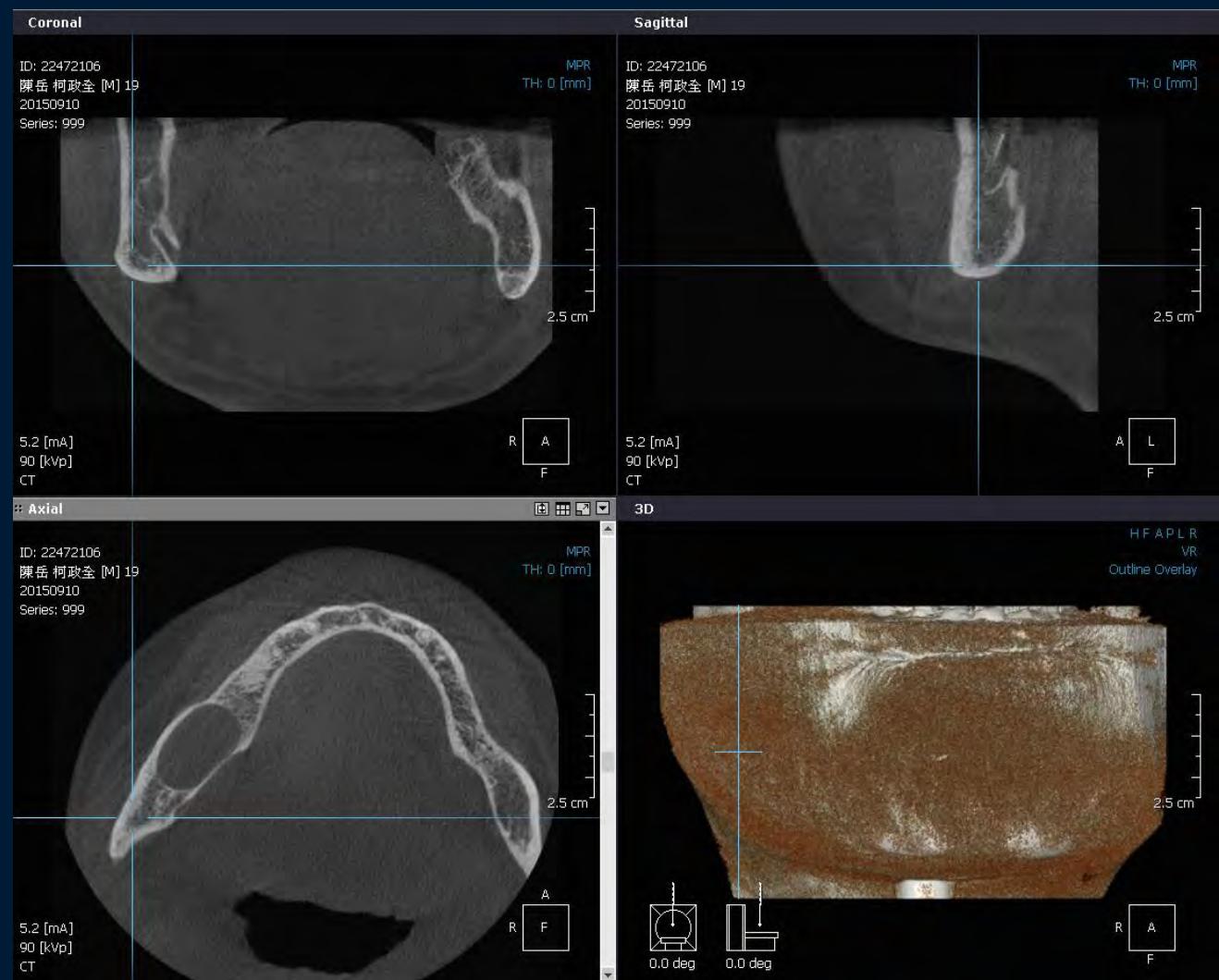
# X-ray findings

❖ 2015/9/10



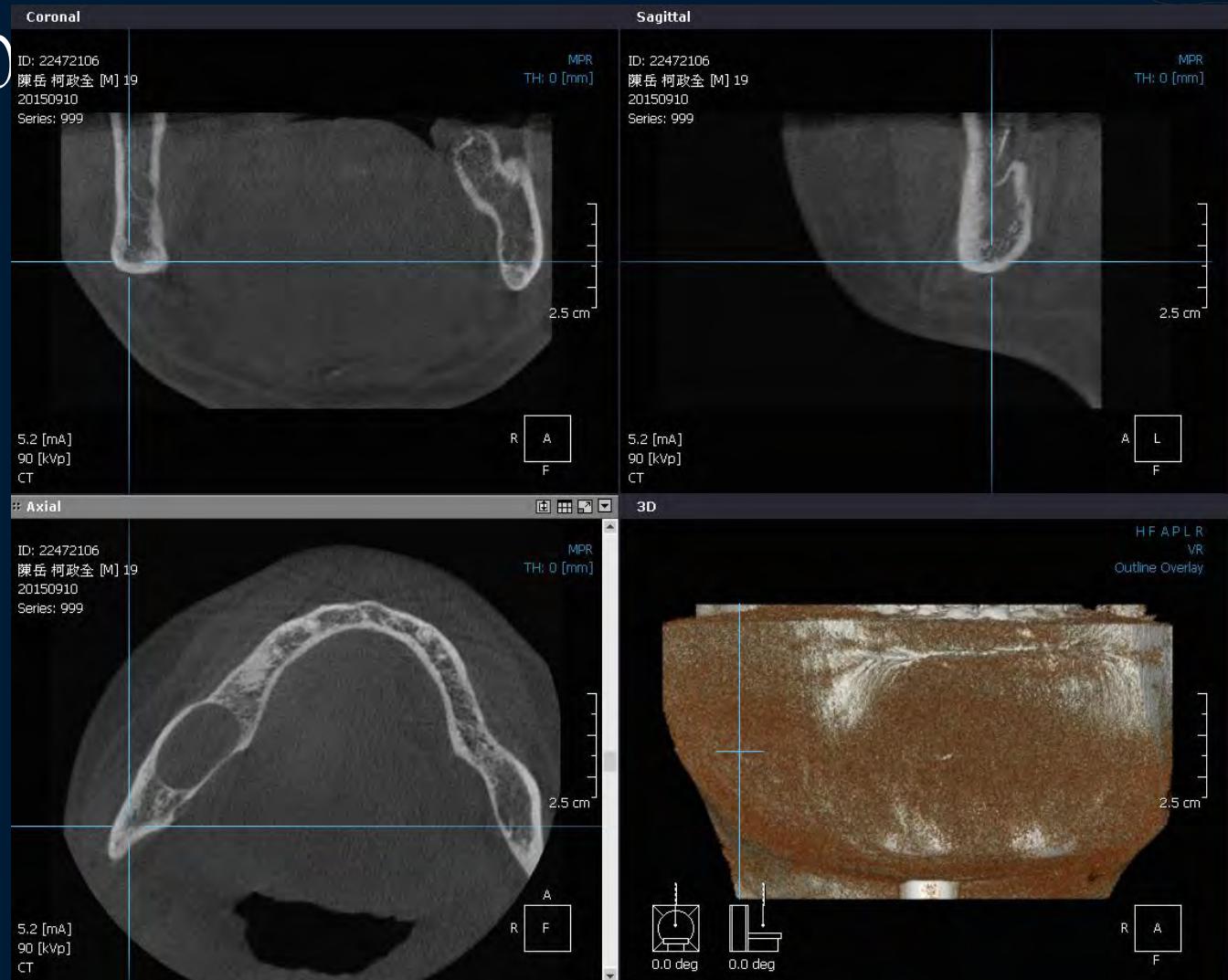
# X-ray findings

❖ 2015/9/10



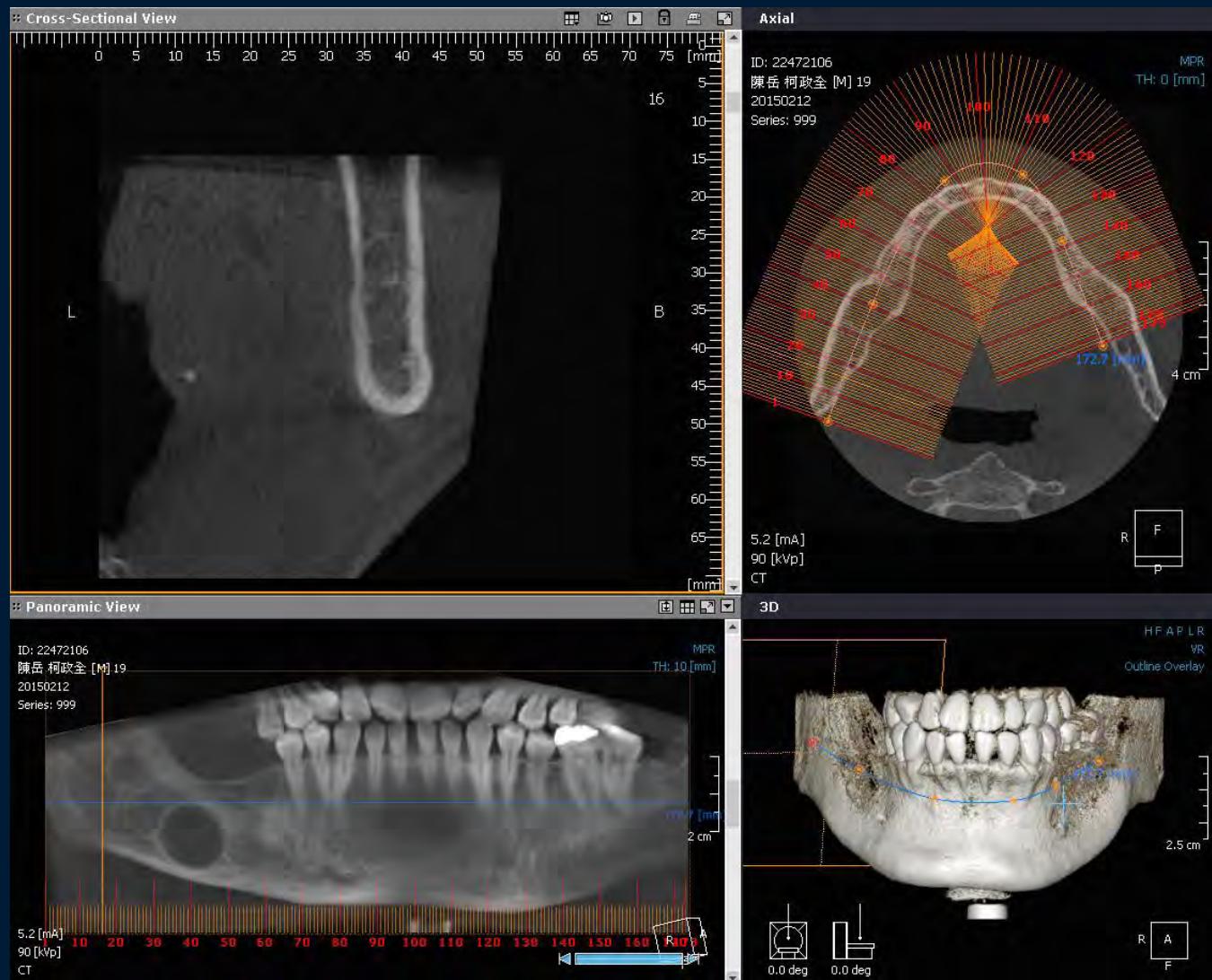
# X-ray findings

❖ 2015/9/10



# X-ray findings

❖ 2015/2/12



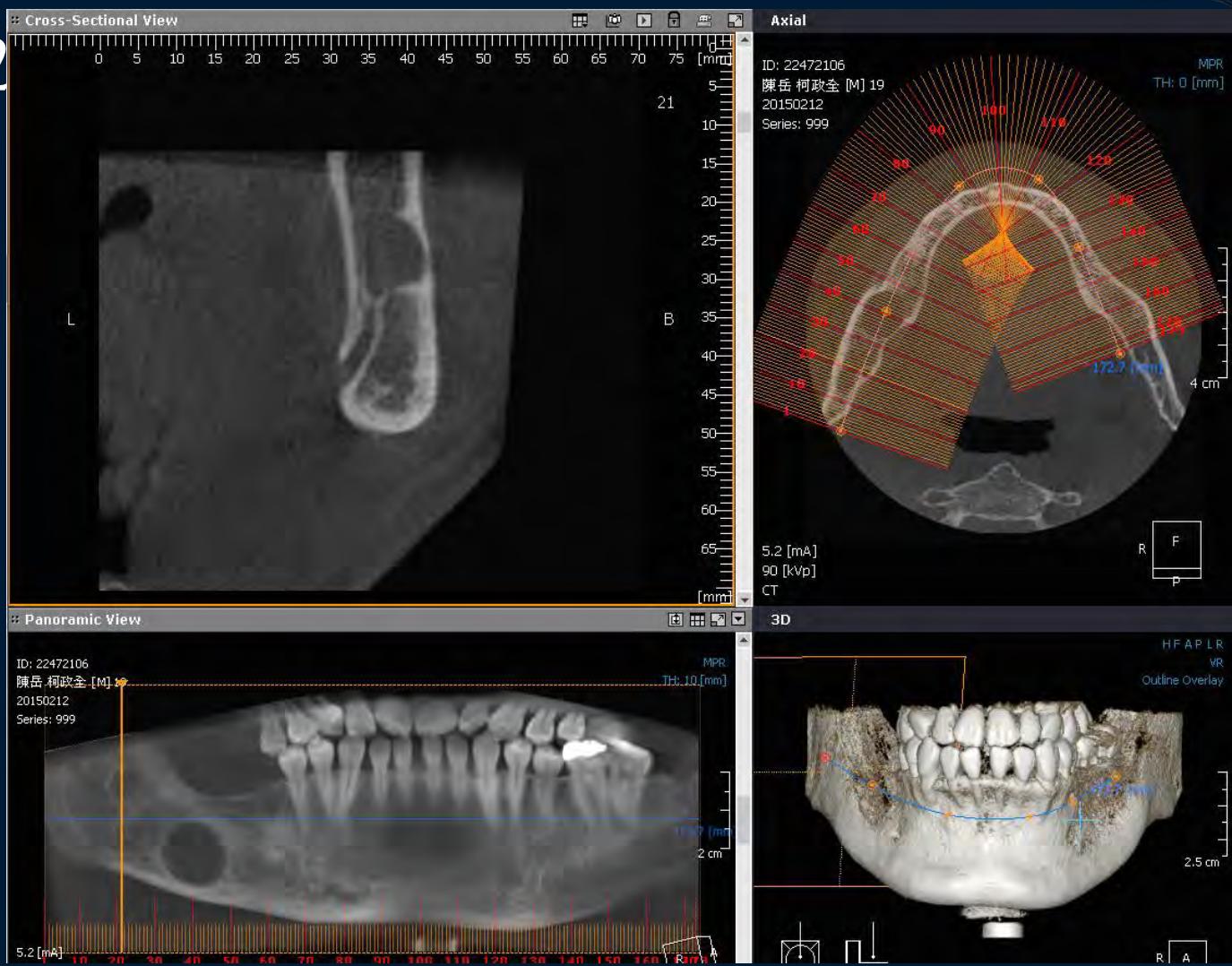
# X-ray findings

◆ 2015/2/12



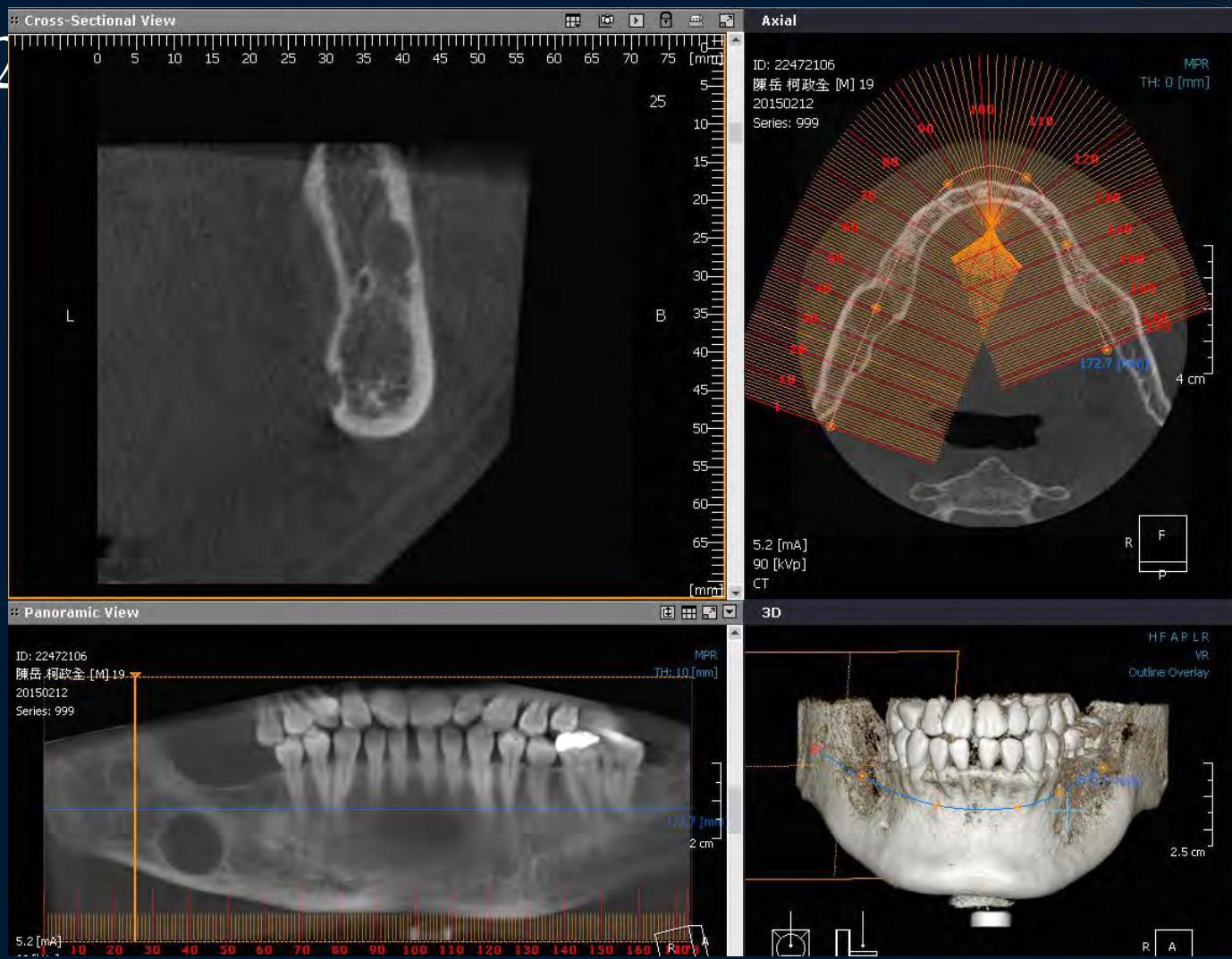
# X-ray findings

◆ 2015/2/12



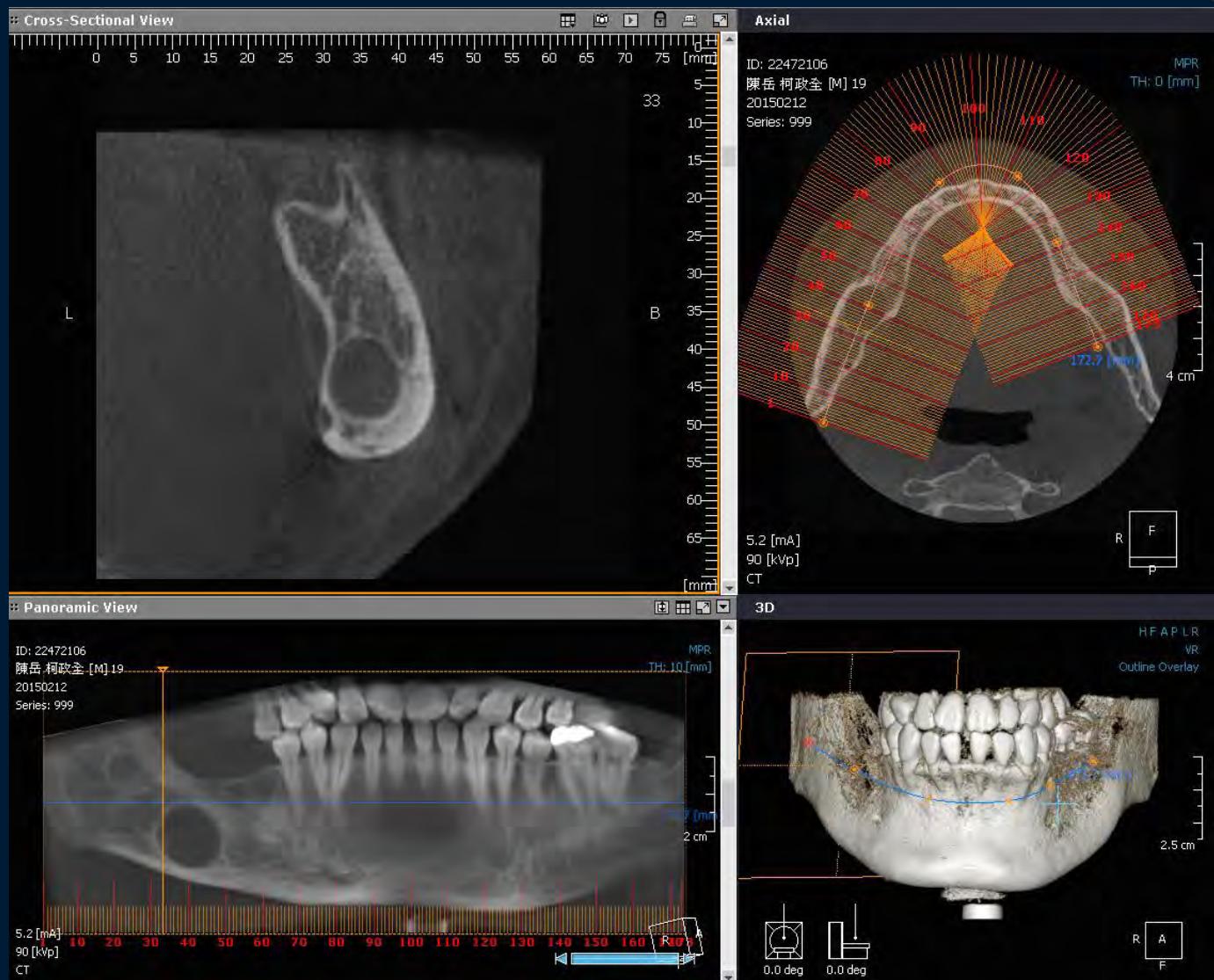
# X-ray findings

❖ 2015/2/12



# X-ray findings

◆ 2015/2/12



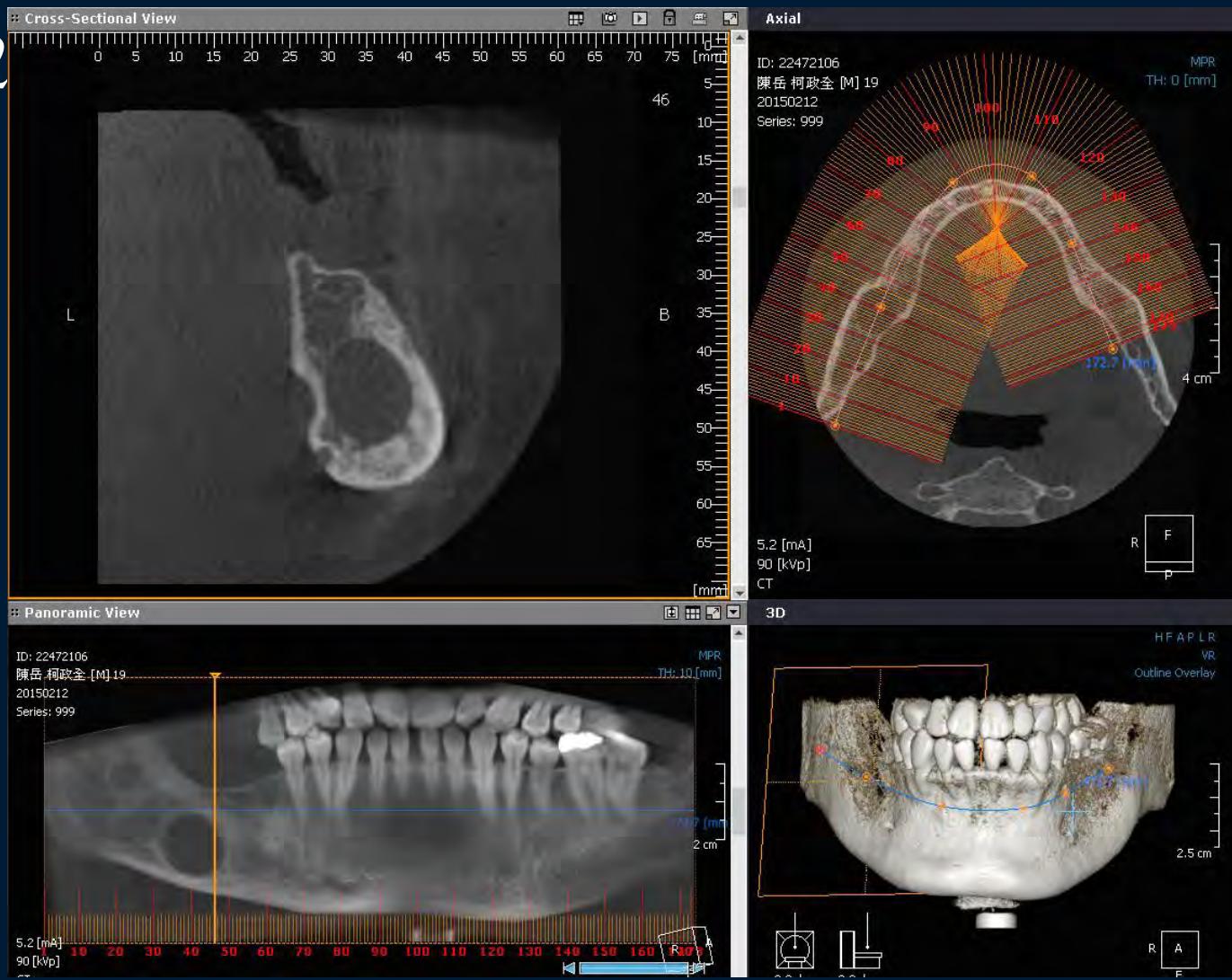
# X-ray findings

❖ 2015/2/12



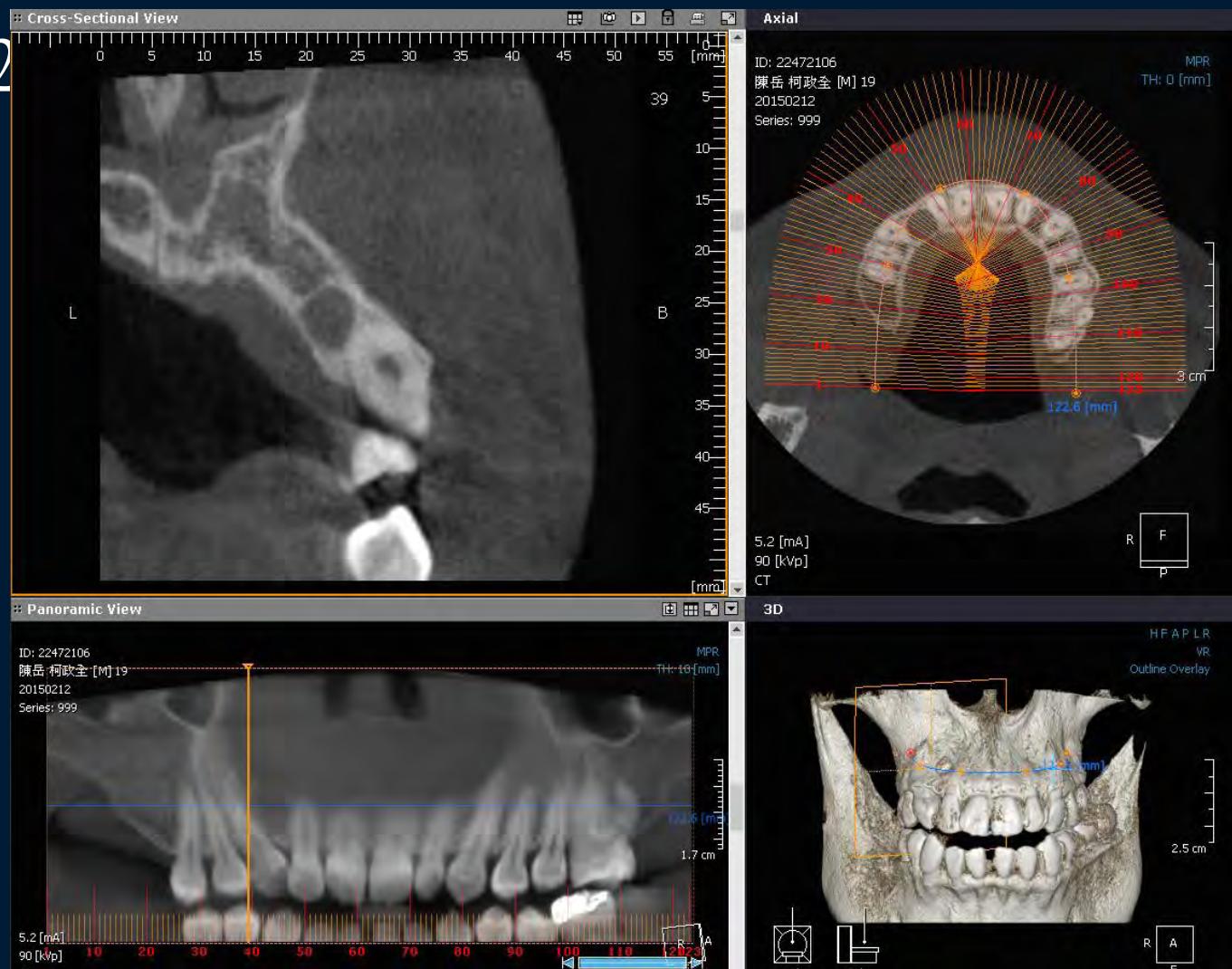
# X-ray findings

◆ 2015/2/12



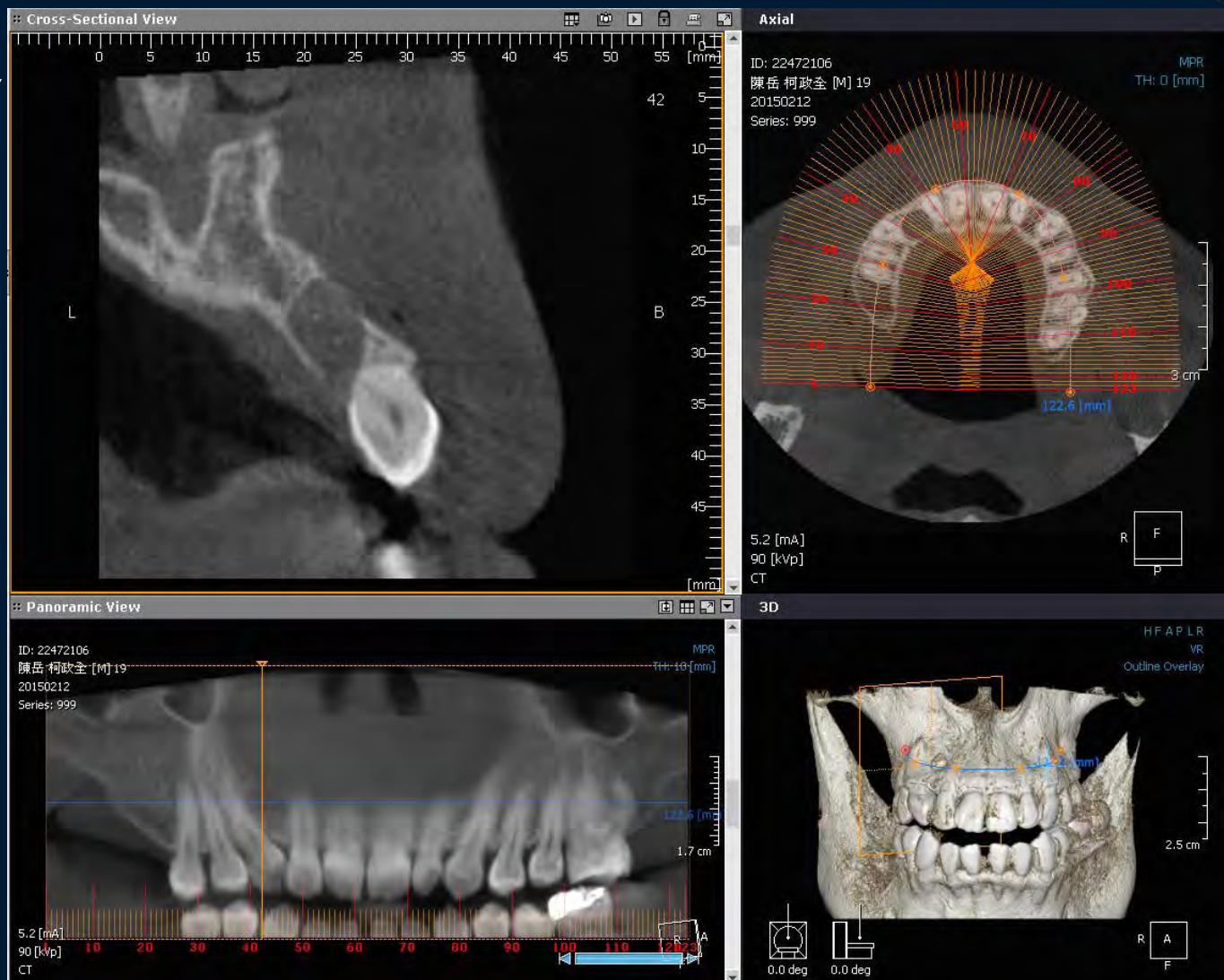
# X-ray findings

◆ 2015/2/12



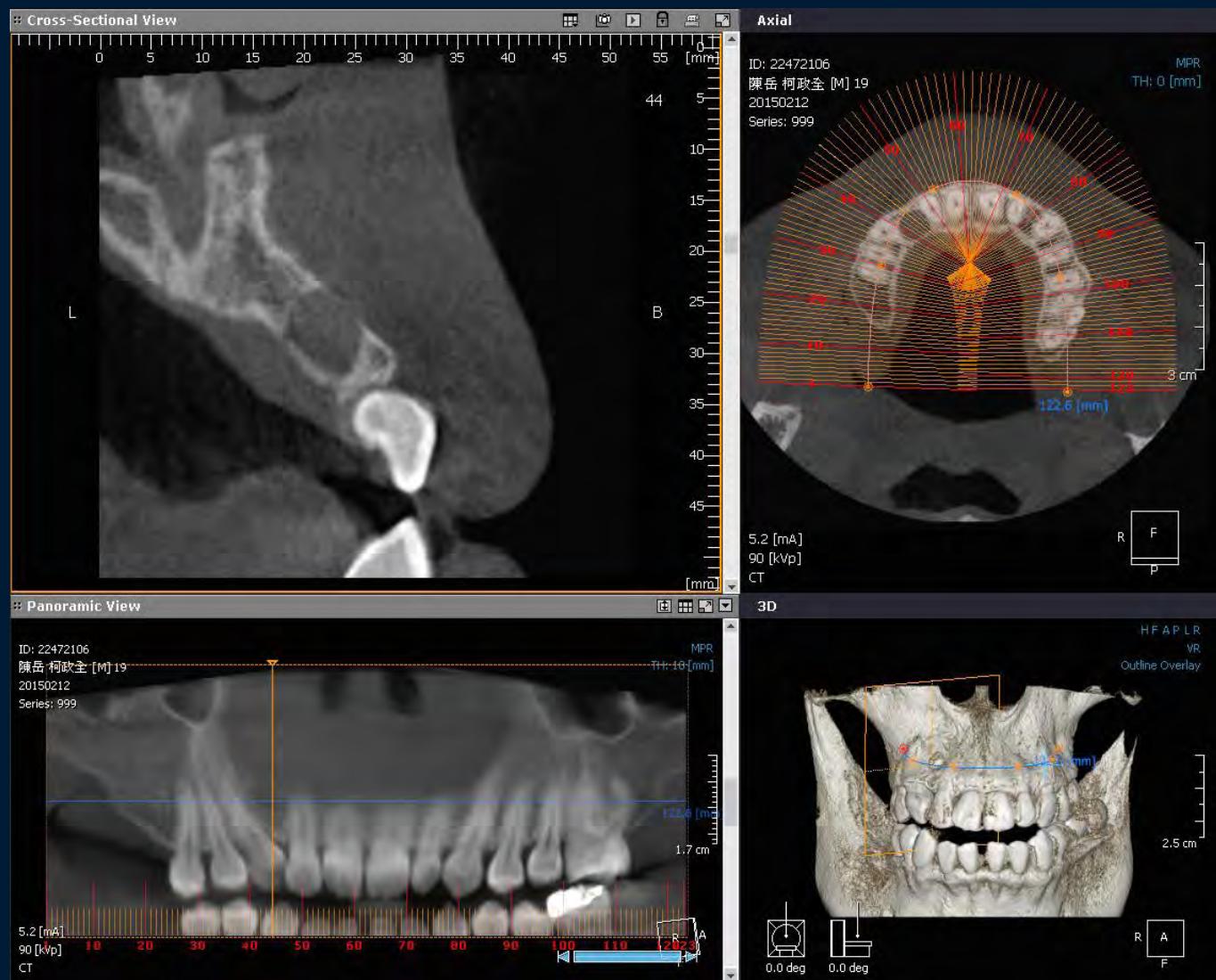
# X-ray findings

◆ 2015/2/12



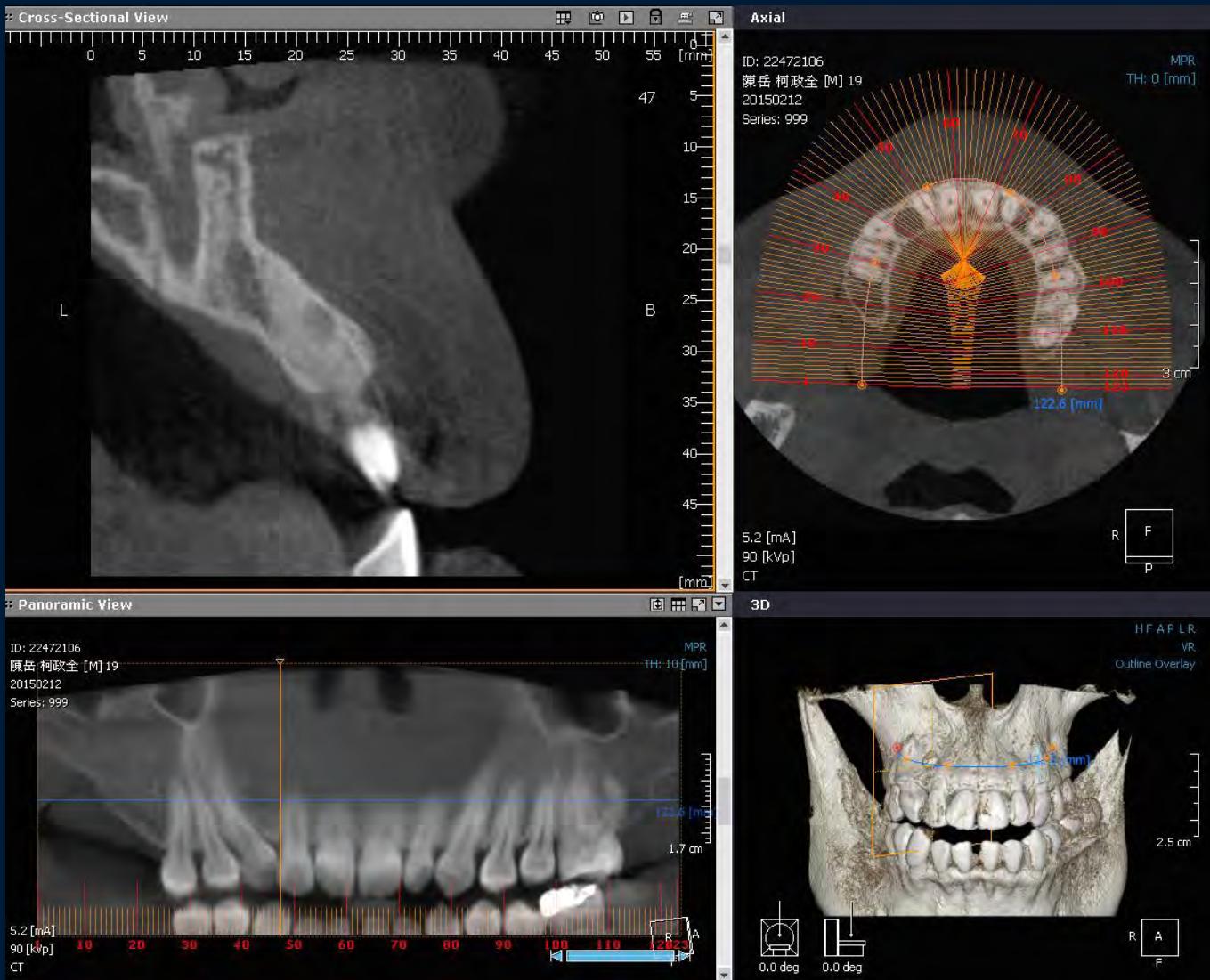
# X-ray findings

◆ 2015/2/12



# X-ray findings

◆ 2015/2/12





# X-ray findings

Mandible:

- ❖ Bone expansion at buccal-lingual and up-down direction.
- ❖ The large one compress right mandibular canal
- ❖ The smaller one is fused with mandibular canal

Maxilla:

- ❖ The lesion invades in right nasal cavity.



# Differential diagnosis

# Differential diagnosis

- Keratocystic odontogenic tumor
- Orthokeratinized odontogenic cyst
- Unicystic ameloblastoma

# Keratocystic odontogenic tumor



	<b>Our case</b>	<b>Keratocystic odontogenic tumor</b>	
Gender	Male	Male > female	
Age	20 y/o	10~40 yrs (60%)	V
Site	Mandible (third molar)	Posterior Mandibular, Mostly molar area(49%)	V
S/S	No	Usually asymptomatic Large: pain, swelling or drainage.	V
size	1 x 2 cm in diameter	varies	
X-ray features	Well-defined unilocular ovoid shaped radiolucency with a sclerotic margins	Well-defined unilocular radiolucency with smooth and often corticated margin 25~40% unerupted tooth involved	V
Clinical features	Unknown	Usually asymptomatic	V

# Orthokeratinized odontogenic cyst



	<b>Our case</b>	<b>Orthokeratinized odontogenic cyst</b>	
Gender	Male	Male	V
Age	20 y/o	Young adult	V
Site	Mandible (third molar)	Posterior mandible	V
S/S	No	No	V
Size	1 x 2 cm in diameter	Less than 1 cm to greater than 7 cm in diameter	V
X-ray features	Well-defined unilocular ovoid shaped radiolucency with a sclerotic margins	Unilocular RL, but occasional examples have been multilocular. Most often involve an unerupted mandibular third molar	
Clinical features	Unknown	Usually asymptomatic	75

# Unicystic ameloblastoma



	Our case	Unicystic ameloblastoma	
Gender	Male	None	
Age	20 y/o	Young age, average 23	
Site	Mandible (third molar)	Post .Mandible	V
S/S	no	Nil	
size	1 x 2 cm in diameter	Average size 4.3 cm~6.3 cm	
X-ray features	Well-defined unilocular ovoid shaped radiolucency with a sclerotic margins	Well-defined, smooth, unilocular radiolucency with corticated margin	V
Clinical features	Unknown	Color: pink Pain (-)	V

# Clinical impression

Keratocystic odontogenetic tumor  
(suspected Gorlin syndrome)



# Discussion

KCOT and Gorlin Syndrome



# Keratocystic odontogenetic tumor



- ❖ Etiology and Pathogenesis
- A distinctive form of developmental odontogenic cyst that deserves special consideration because of its specific histopathologic features and clinical behavior



## ❖ Clinical features

- Sex predilection: ♂ ≥ ♀
- Age predilection : 10~40 years old (60%)
- Racial predilection: None
- Site predilection: Mandible(60~80%)

	Anterior	Premolar	Molar
Maxilla	13%	2%	20%
Mandible	9%	7%	49%

- Symptom and Sign

1. 多半沿著anterior-posterior方向長，就算是很大的lesion也不會有expansion
2. 用針筒抽取病灶，可抽出草綠色或黃綠色的液體



## ❖ Radiographic Features

- 通常是一個well defined且有sclerotic bone圍繞的unilocular RL，25%~40%包裹著一個impacted tooth
- 比起dentigerous cyst跟radicular cyst，KCOT比較不會造成鄰接牙齒的牙根吸收



## ❖ Histopathologic Features

- 可見cyst的三種特徵：cyst wall、lining epithelium、lumen/cavity
- Cavity中可見到很多類似keratin的東西
- Lining epithelium：
  1. 均勻厚度為6~8層的stratified squamous cell
  2. epithelium 與wall接面平坦，rete ridge不明顯
  3. basal cell呈現柵狀排列(palisaded)，為cuboidal或columnar
  4. 最靠近cavity的cell為parakeratinizing，呈現wavy appearance
- 若發炎狀況發生，上皮會出現rete ridge且parakeratinizing和palisaded basal cell都消失

# Nevoid Basal Cell Carcinoma syndrome(Gorlin syndrome)



- ❖ Etiology and Pathogenesis
- 為體顯性遺傳(autosomal dominant)
- 與第九對染色體的PTCH(一種tumor suppressor gene)基因的變異有關



## ❖ Clinical Features

- 頭圍增加(increased cranial circumference)
- 眼距變大(hypertelorism)
- 輕微下顎前突(mandible prognathism)
- 表皮多發的basal cell carcinoma，特別是在中臉部，30歲前發生
- 手掌或腳掌有凹陷(Palmar and plantar)
- 女性有卵巢纖維瘤(Ovarian fibroma)
- 肋骨分叉或外展(rib bifid or splayed)
- 多發性的odontogenic keratocyst：與一般OKC比，p53和cyclinD1(Bcl-1)過度表現



## ◆ 治療照護

戈林症候群需要有經驗的專科醫師來進行治療及追蹤照護，重點如下：

- \* 早發的角化腫瘤藉手術切除。
- \* 基底細胞瘤早期治療：確實根除，並保留正常組織避免對外形的損壞
- \* 卵巢纖維瘤之手術治療：需保留卵巢組織。
  
- \* 預防主要症狀：
  - 避免過度暴露於陽光底下。
  - 不使用放射治療(如X射線)，以避免提高治療部位發生多發性基底細胞瘤的風險。



- \* 追蹤監控：
  - ~ 兒童階段需定期測量頭圍。
  - ~ 由於患者罹患髓質母細胞瘤的風險高，滿週歲以內即需開始每半年做一次發展評估及身體理學檢查。
  - ~ 滿8歲後，每12~18個月做一次牙科orthopantogram檢查，目的在於及早發現口腔角化囊腫。
  - ~ 至少每年做一次全身皮膚視診 (skin examination)。
- \* 對有罹病風險的家人進行評估：由於戈林症候群在照護上需針對併發症 (如兒童髓質母細胞瘤、口腔角化囊腫、基底細胞瘤) 進行追蹤監控，患者在生活上也需避免陽光的曝曬，因此建議有罹病風險而尚未出現症狀的家人，應進行基因檢測，以確認是否罹病。

# 醫學倫理討論





# Tom Beauchamp & James Childress

## 六大原則 - 1979

Sanctity of life (生命的神聖性)。

- 1.. **Justice(公義原則)**: 醫師在面對有限的醫療資源時，應以社會公平、正義的考量來協助合理分配此醫療資源給真正最需要它的人。
2. **Confidentiality (保密原則)**: 醫師對病人的病情負有保密的責任。
3. **Veracity(誠信原則)**: 醫師對其病人有「以誠信相對待」的義務。
4. **Nonmaleficence (不傷害原則)**: 醫師要盡其所能避免病人承受不必要的身心傷害。
5. **Autonomy (自主原則)**: 病患對其己身之診療決定的自主權必須得到醫師的尊重。
6. **Beneficence (行善原則)** : 醫師要盡其所能延長病人之生命且減輕病人之痛苦。



# 生命的神聖性

- 在《聖經》的第一篇<創世紀>中,上帝告訴以色列人說:「上帝按他自己的形象造人。」「你將是神聖的,因為我是神聖的。」「生命神聖」觀即由此衍生而得。
- 該觀點主張人的生命是無條件的,有價值及神聖的,人繼承了上帝的品質,包括一切價值的來源-內在的善 (*intrinsic goodness*),因此必須受到尊重。
- 藉此瞭解他個人生命的原真,而認知他個人存活在世上的主要工作和生活的目的,找到個人存在的意義、價值、目的與任務。



# 公義原則

## ◆ 手術的必要性？

→ Dentigerous cyst最佳的治療方式是 surgical excision，將病灶完整的清除(enucleation)才能將復發率(recurrence rate)降到最低。



# 保密原則

## 告知的對象

1. 本人為原則
2. 病人未明示反對時，亦得告知其配偶與親屬
3. 病人為未成年人時，亦須告知其法定代理人
4. 若病人意識不清或無決定能力，應須告知其法定代理人、配偶、親屬或關係人
5. 病人得以書面敘明僅向特定之人告知或對特定對象不予以告知



# 誠信原則

- ◆ 對於患者的疾病嚴重程度是否有確實地通知，盡到告知的義務？
- ◆ 是否有清楚的向病人說明清楚疾病病程、治療計畫、預後、風險？  
→皆以已告知病人後，經同意才進行手術。



# 不傷害原則

- ❖ 是否有先完整瞭解病人的病史?  
→治療前有完整蒐集病史資料，並與病患溝通後擬定進一步的治療計畫
  
- ❖ 手術過程中，是否有造成不必要的醫源性的傷害？  
→沒有不必要的醫源性傷害。



# 自主原則

- ◆ 充分說明病情及治療計畫、風險之後，是否有讓病人充分自主地選擇治療計畫？  
→ 病人及家屬選擇並同意醫師的建議。
- ◆ 在做全身麻醉以前，是否有說明完整之後再請病人自主的簽名同意？  
→ 已充分說明並與家屬溝通。



# 行善原則

- ◆ 做了Excision 後是否有減輕病人的疼痛感？或是使病人更不舒服？  
→有完整去除病灶區域並拍照記錄術後情形。並告知術後傷口會疼痛，但持續癒合後疼痛會逐漸緩解



# 醫學倫理總結

- ◆ 在病例撰寫方面(病灶描述,治療計畫,病人態度)應書寫詳盡，使治療過程有詳實的記錄及治療順利。
- ◆ 在進行治療之前,須請病人簽屬同意書
- ◆ 應在不違反醫學倫理的原則之下進行治療的行為

THANK YOU FOR  
YOUR ATTENTION!

