Original Article

บทบาทของโคนบีมคอมพิวเตดโทโมกราฟฟีในการวินิจฉัยใบหน้าแหว่งข้างเดียวประเภทเทสเซียร์ 5

The role of CBCT on the diagnosis of Unilateral Tessier Facial cleft 5

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บทคัดย่อ ใบหน้าแหว่งข้างเดียวประเภทเทสเซียร์ 5 เป็นลักษณะใบหน้าแหว่งที่พบได้ยากที่สุด และมีผลกระทบต่อผู้ป่วยค่อนข้างมาก การวินิจฉัยโรคใบหน้าแหว่งหากเป็นกรณีที่แสดงความผิดปกติที่เนื้อเยื่ออ่อนภายนอกสามารถวินิจฉัยได้ง่ายโดยการตรวจทางคลินิก แต่การตรวจทางภาพถ่ายรังสีจะช่วยในการประเมินตำแหน่งของรอยแหว่งในกระดูกได้ อย่างไรก็ตามเมื่อถ่ายภาพรังสีที่เป็นภาพสองมิติ อาจพบการซ้อนทับกันของกระดูกขากรรไกรและใบหน้า นำไปสู่การวินิจฉัยโรคที่ผิดพลาดได้ ในปัจจุบันการถ่ายภาพโคนบีมคอมพิวเตดโทโมกราฟฟี ซึ่งแสดงภาพแบบสามมิติ สามารถให้ข้อมูลที่ถูกต้องแม่นยำมากและมีการนำมาใช้อย่างกว้างขวาง แต่ยังไม่ค่อยได้นำมาใช้ในงานวินิจฉัยและวางแผนการรักษา

บทความนี้จึงขอนำเสนอกรณีศึกษาการใช้การถ่ายภาพโคนบีมคอมพิวเตดโทโมกราฟฟีในการวินิจฉัยและวางแผนการรักษาลักษณะใบหน้าแหว่งข้างเดียวประเภทเทสเซียร์ 5 และอภิปรายลักษณะทางภาพรังสีของโรคดังกล่าว

คำสำคัญ: โคนบีมคอมพิวเตดโทโมกราฟฟี, โรคใบหน้าแหว่ง, ใบหน้าแหว่งข้างเดียวประเภทเทสเซียร์ 5

Abstract Tessier cleft 5 is the rarest of oblique clefts and has a big impact on those who were born with it. Cranio-facial clefts (CFCs) can be easily diagnosed by clinical examination in cases with external soft tissue manifestations. Radiographic investigations were also used to evaluate the position of the cleft. However, two-dimensional radiographs may lead to

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misdiagnosis due to the superimposition of the craniofacial skeleton. Recently, cone-beam computed tomography (CBCT) provided accurate three-dimensional imaging that is widely used but not routinely in diagnosis and treatment planning.

This paper presented a rare case of Tessier cleft 5 with no extraoral soft tissue manifestations using a CBCT scan for the definite diagnosis and also the treatment planning and discussed a radiographic approach to the diagnosis of the CFCs.

Keywords: Cone-beam computed tomography; Cranio-facial cleft; Tessier cleft 5

INTRODUCTION

Cranio-facial clefts (CFCs) are malformations that affect both the face and cranium in various forms. The incidence of CFCs is approximately 1.4-6.0 per 100,000 live births[1, 2] CFCs do not only involve the lip, palate, nose, and alveolar ridge but also eyes, maxilla, and forehead. Most disfigurations related to facial anomalies are attributed to CFCs.[3] The most known cleft that occurs in the craniofacial region is cleft lip and palate.[4] There are several systems of classification and phenotype characterization of the CFCs, such as the classification systems of Morian, American Association of Cleft Palate Rehabilitation (AACPR), Boo-Chai, Karfik, and Tessier.[3-7] However, the most common and widely used is Tessier classification.

Tessier classification involves not only soft tissues but also skeletal anomalies. The numbers 0 to 14 were used to define the location of the clefts as shown in Fig. 1. The clefts are also classified about the facial midline and eye socket: 0 to 3 as middle clefts, 4 to 6 as oblique clefts, 7 and 8 as lateral or transverse clefts, and 9 to 14 as an extension of 0 to 5 respectively. When using the eye socket as a reference, those above the upper eyelids are called cerebro-cranial clefts (numbers 0 to 7) while those that are below the lower eyelids are craniofacial clefts (numbers 8 to 14). Soft tissues are often involved with the occurrence of cleft resulting in external or surface manifestations.[4, 8] Craniofacial clefts may occur as unilateral or bilateral and complete or incomplete. In case of bilateral occurrence, one side may be more prominent than the other side and usually different type of cleft occurs on each side.[4]

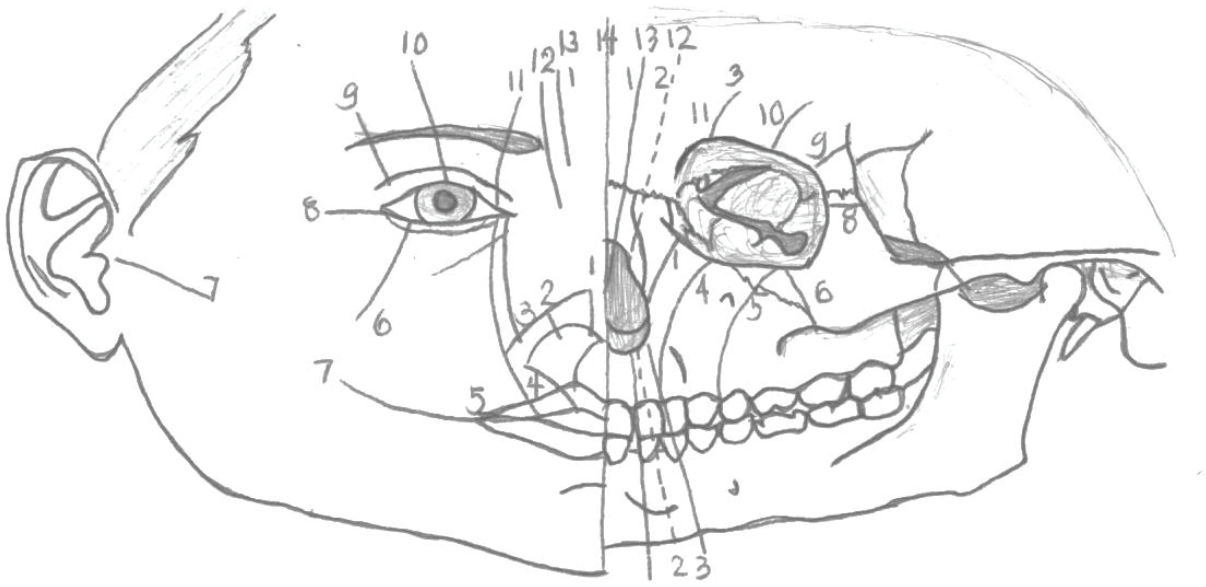

Fig. 1 Tessier classification of the craniofacial clefts; on the right (R) are the lines of the bone tissue cleft and shown on the left (L) are the lines of the soft tissue cleft.

Oblique clefts are uncommon facial clefts and may involve the palate and temporal bone. Their incidence is estimated to be 0.075-0.54% [9] which is only 0.22-0.25% of all CFCs.[3] Tessier cleft 5 is the rarest of the oblique clefts.[3, 10] In literature, there are other names for Tessier cleft 5 according to different authors and institutions, such as *"orofacial canthal cleft"* (AAPCR), *"type II oro-ocular cleft"* (Boo-Chai), *"true cleft"* (Karfik), and *"lateral maxillary dysplasia"* (van der Meulen).[4, 11] Moreover, Pereira et al.[12], Whitaker[5], and David et al.[13] described the features of Tessier cleft 5 in different characteristics.

Infants who are born with CFCs, like Tessier cleft 5, have a big impact on their lives. They are prone to nutritional deficiency due to feeding difficulties, have poor oro-motor development, and may have hearing loss.[14] In severe cases of CFCs, abnormalities relating to the eyes, such as eyesight impairment, can occur.[15] Older patients with untreated CFCs have remaining problems with speech, hearing, and eating. These problems may give an individual with CFCs low self-esteem and psychosocial effects, such as social adjustments.

Diagnosis of CFCs can be easily made by clinical examination of different anatomical areas, such as orbit, cheek, lip, alveolar bone, and soft palate. Radiographic examinations, such as panoramic and intraoral radiographs, were also performed to evaluate the position of the cleft. However, these two radiographic techniques could not cover the whole morphology of the facial skeleton and its relation to the infraorbital foramen which is a necessary anatomy for the diagnosis. Meanwhile, a cone-beam computed tomography (CBCT) scan allows an accurate 3D imaging of the complex hard tissues that aids in diagnosis and planning for the extent of surgical reconstruction of CFC malformations. In addition to the standard 3D image, the presence of serial, coronal, axial, and sagittal formats gives finer details on the skeletal distortions. Even though medical CT produces superior images, CBCT is more advantageous for maxillofacial applications especially for dental use because of its lower dose of radiation and comparable images of osseous structures at lower cost.[16]

This paper aimed to present a rare case, Tessier cleft 5, including an analysis of its characteristics which differ from the other reported cases and to introduce a radiographic approach to diagnosis. Moreover, the radiological differential diagnosis of this abnormality is also discussed.

CASE REPORT

A 17-year-old male patient came to the Dental Hospital, Faculty of Dentistry, Khon Kaen University, Thailand in 2015 with a chief complaint of having facial asymmetry since birth and an enlarged tissue at the right posterior area of the maxilla. He was born of a non-consanguineous marriage and had no remarkable antenatal history. He had good physical and mental development except for his facial deformity. Extraoral examination showed facial asymmetry with a deviation of the mandible towards the right. There was no prominent extraoral manifestation of a cleft except for the right deviation of the patient's mouth and the tissue tags anterior to the left ear (Fig. 2). Intraoral examination showed an anterior open bite with unilateral crossbite at the right posterior teeth. The alveolar bone at the area of the right posterior maxilla expanded bucco-palatally with bony hard consistency on palpation. Severe crowding and supernumerary teeth can also be observed on the upper arch especially on the right side (Fig. 3).

From the panoramic radiograph (Fig. 4), a double maxilla was found on the right side including supernumerary teeth, of which the shape like the second premolar and molars and the deciduous retained root remained between the premolar and molar. The distorted shape and enlarged size of the right zygomatic process of the maxilla were seen. Moreover, the right maxillary sinus was recessed and the right posterior hard palate was lifted. However, it is difficult to interpret accurately by the general dentist due to the superimposition of bony structures. The first interpretation led to a mixed radiolucent-radiopaque lesion or cystic lesion which could be a differential diagnosis as fibrous dysplasia, ameloblastoma, and odontoma. Therefore, the biopsy was performed to rule out the impression of the previous differential diagnosis. The histopathological report indicated that the specimen was consistent with normal compact and cancellous bone or a mature stage of benign fibro-osseous lesion. The pathologist suggested that clinical-radiologic-pathologic correlation was required.

The patient was then sent for CBCT (WhiteFox Cone Beam Computer Tomography Scanner®, Acteon Group, France) imaging. The CBCT scan demonstrated a cleft on the right premolar region of the patient's maxilla extended to the area slightly below the lateral third of the orbit and the right zygomatic arch lower than the other side. The cleft separated maxillary structures posterior of the premolar from the rest of the upper arch resulting in a double maxilla. (Fig. 5a, 5b, and 6). The cleft continued inward medially ending in the posterior area of the soft palate. The right eye socket also appeared to be smaller and the floor of the right orbit was slightly pushed superiorly. The right maxillary sinus and nasal cavity were intact but were smaller relative to the other side and were displaced superiorly showing asymmetry on both sides. There was also skeletal disturbance of the sphenoid and pterygoid plates. Right pterygoid plates seemed to have asymmetry in size and were closer to the midline compared to the pterygoid plates

on the other side. The lateral wall margin of the sphenoidal sinus was thicker on the right, which might have caused the sphenoidal sinus to be slightly compressed towards the left (Fig. 7). Bucco-palatal expansion of the alveolar bone on the right side of the posterior maxilla could also be observed. Supernumerary teeth and malocclusion of teeth could also be seen on the radiographs (Fig. 6). Both condyles were similar in shape and size. Their positions were relative to the glenoid fossa (Fig. 7C). There was no asymmetry of the cranial base and calvarium. The patient was currently undergoing pre-orthodontic surgical treatment in preparation for his orthodontic therapy. This case report was approved by the Khon Kaen University Ethics Committee for Human Research (HE612338).

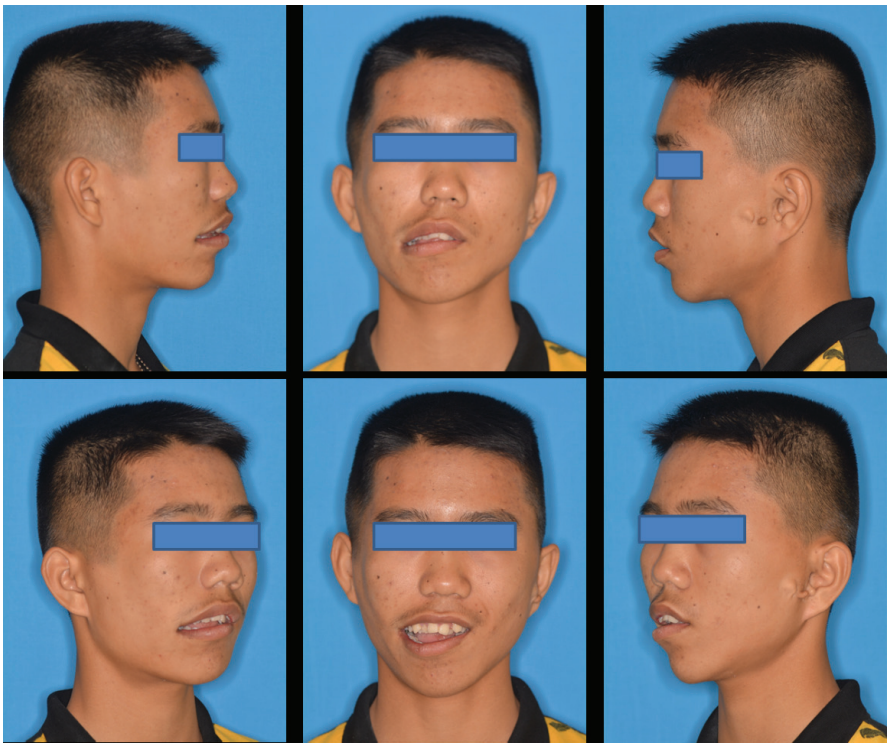

Fig. 2 Extraoral photographs of the patient showing facial asymmetry with incompetent lip and mandible deviated to the right side.

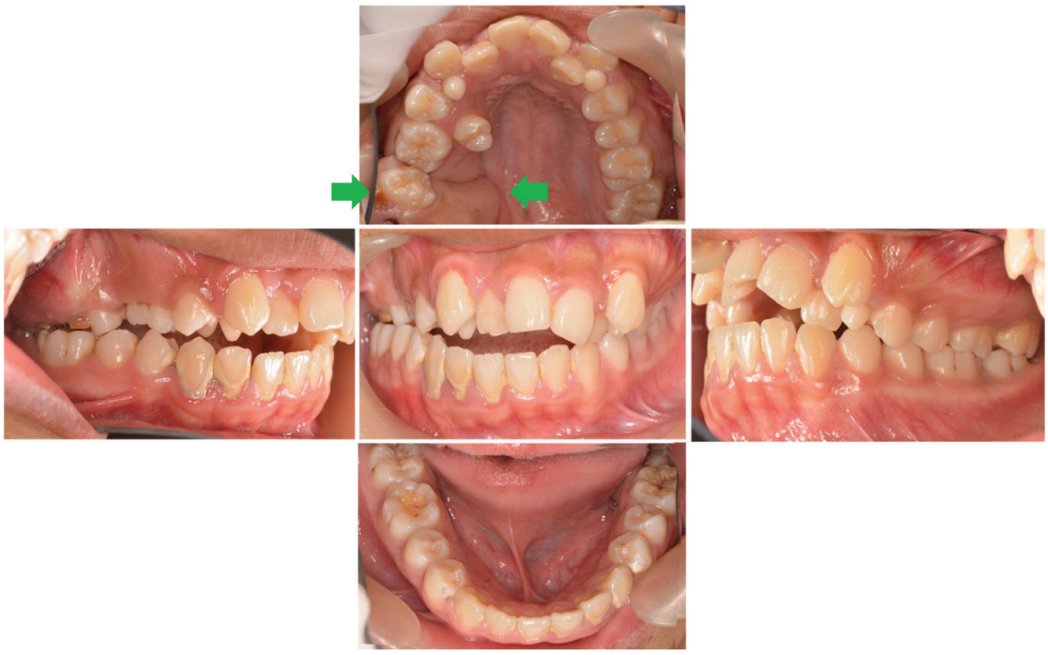

Fig. 3 Intraoral photographs of the patient showing anterior open bite and severe crowding with unilateral crossbite (right side). The alveolar bone at the area of the right posterior maxilla expanded bucco-palatally with bony hard consistency on palpation (arrow).

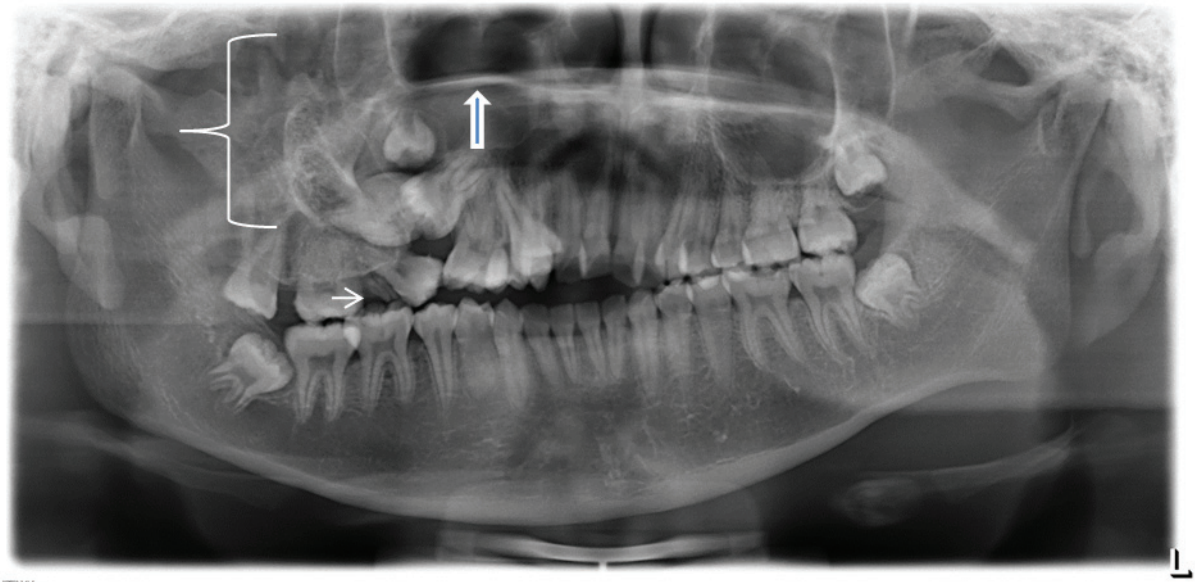

Fig. 4 Panoramic radiograph showing double maxilla on the right side including supernumerary teeth and the deciduous retained root (arrow). The zygomatic process of the right maxilla showed a distorted shape and enlarged size (parenthesis). The right maxillary sinus was recessed. The right posterior hard palate was lifted (thick arrow). The lower border of the mandible on the right side was lower than that of the left side.

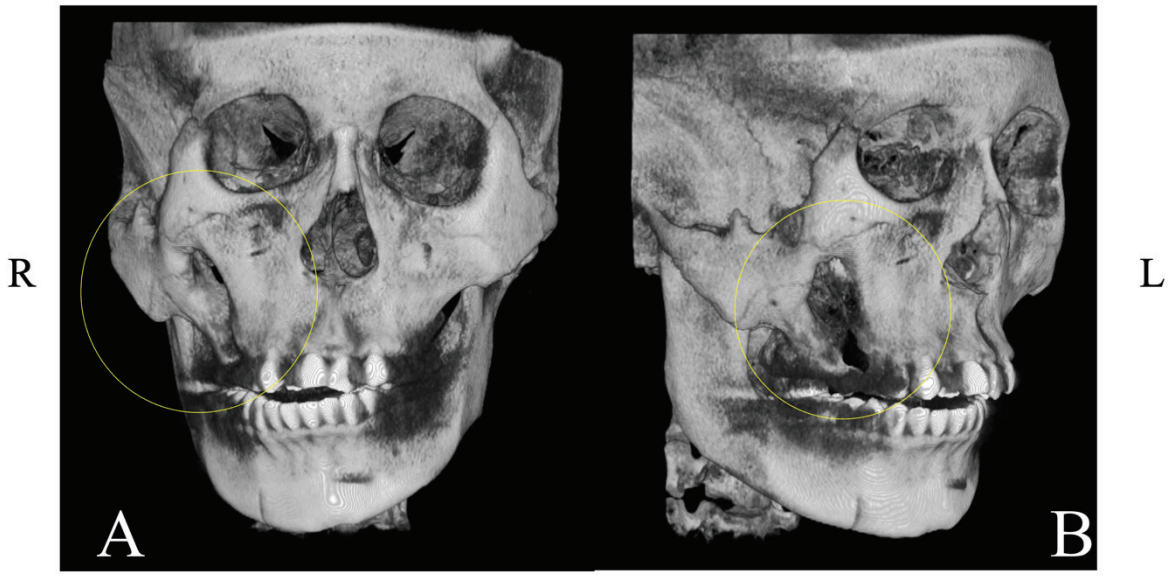

Fig. 5 Three-dimensional CBCT reconstruction in frontal (A) and oblique (B) views show the extent of Tessier cleft 5.

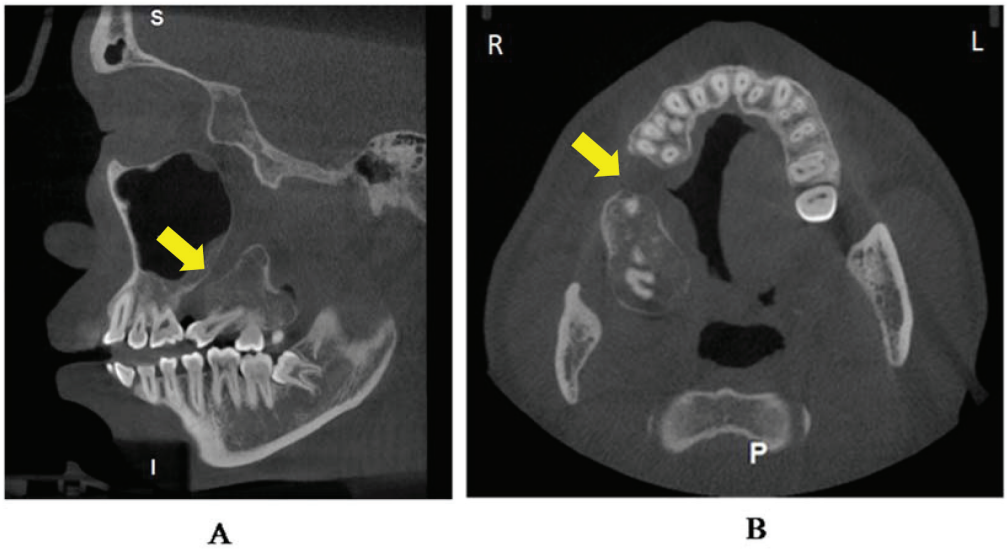

Fig. 6 Sagittal (A) and Axial (B) views of CBCT scan showing separation of the right maxilla (S = superior, I = inferior, P =posterior, R= right, L = left) .

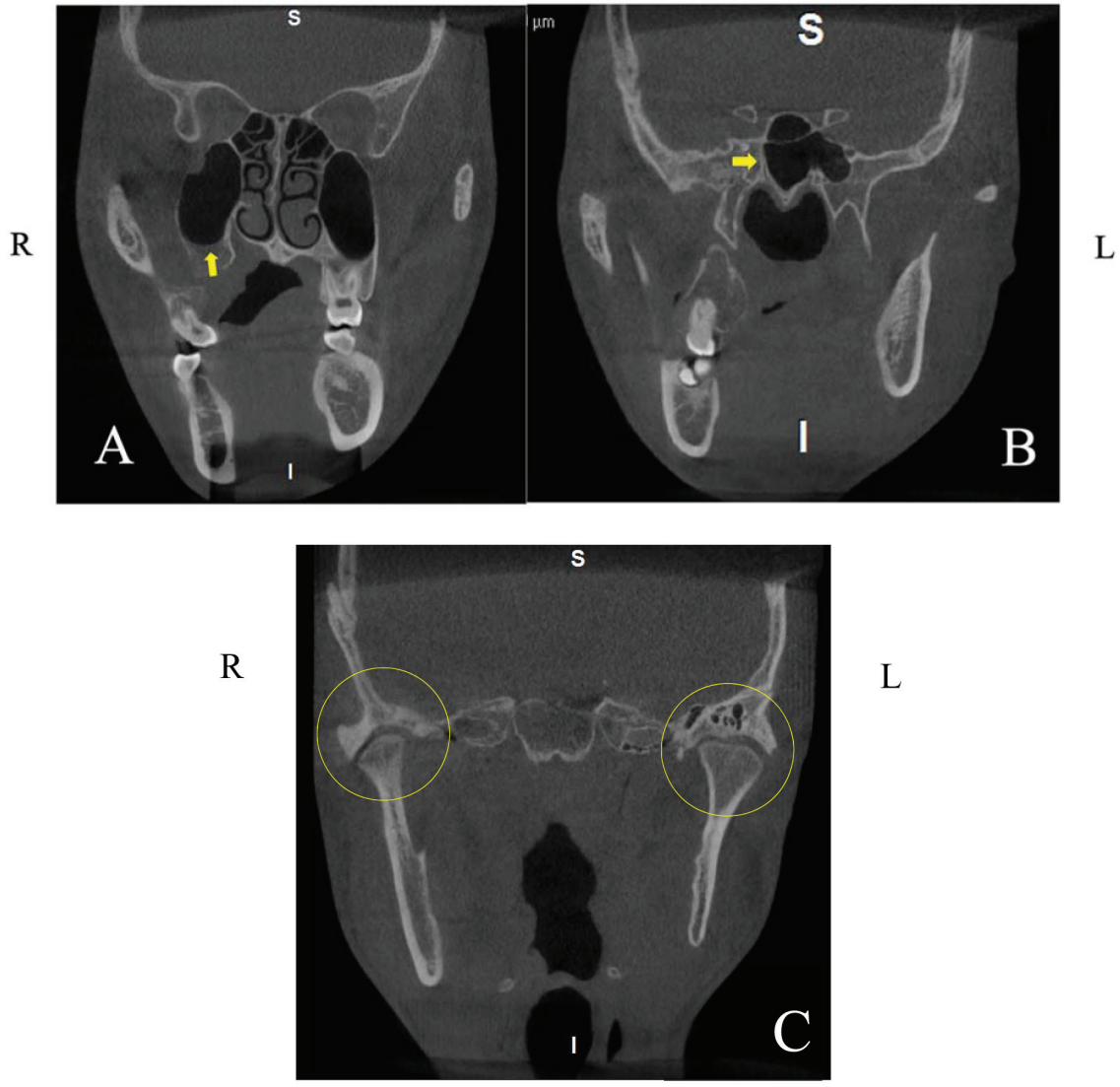

Fig. 7 Coronal view of the patient's CBCT scan showing – A. displacement of the right maxillary sinus and nasal cavity; B. compression of the sphenoidal sinus to the left; C. position of condyles in relation to the glenoid fossa.

DISCUSSION

In this report, we analyzed a Tessier cleft 5 using CBCT. Unlike most of the reported cases of Tessier cleft 5, the patient showed no significant external soft tissue manifestation of the cleft except with the presence of tissue tags anterior to the left ear and facial asymmetry with a downward oblique lip line deviating towards the right. Lack of external soft tissue manifestation combined with the rare occurrence of Tessier cleft 5 made it somehow difficult to readily diagnose this case.

Since no known embryologic grooves correspond to Tessier cleft 5, it was theorized that oblique facial clefts were due to stop in development, tissue disruption secondary to maxillary terminal branches lesions, or disturbances in the development of the maxilla. [3, 4, 17] Recent findings suggest that Tessier cleft 5 is a combination of tethered tissue migration, e.g. amniotic band, and cellular ischemia due to pressure [4], and are strongly associated with other congenital malformations such as limb constrictions. Amnion rupture syndrome exemplifies the relationship of tethered tissue migration with CFCs, which includes craniofacial clefts, amniotic bands, and visceral and extremities malformations.[3] However, Tessier cleft 5 was not linked to limb malformation and/or constrictions based on Coady.[18] Aside from genetic causes, risk factors including the use of drugs, smoking, exposure to radiation, vitamin deficiency and toxicity, and infection during pregnancy are also related to the development of CFCs.[19] The patient presented here was born with no remarkable antenatal history and malformations of extremities. The concept of tethered tissue migration cannot explain the presence of skeletal Tessier cleft 5 in this case. Furthermore, no external soft tissue manifestation in the present case can support that tethered tissue migration occurred in the patient during development in the uterus. But, we also cannot exclude the possibility that the former theories may be the reason for this Tessier cleft 5 case.

The external manifestation of Tessier cleft 5 shows on the lateral third of the eyelid and oral commissure of the lip.[8] Facial asymmetry is one of the main characteristics of Tessier cleft 5. Facial asymmetry can also be present in other conditions, such as hemifacial microsomia (HFM), which is a first branchial arch syndrome involving underdeveloped temporomandibular joint (TMJ), mandibular ramus, mastication muscles, and the ear.[20] Clinical appearance of HFM ranges from a slight facial asymmetry to severe stunted facial half, which can include orbital problems, a partially formed or total absence of ear, deviation of the facial midline to the affected side, and an upward oblique lip line towards the affected side.[20] The case presented here, although it showed a facial asymmetry that includes an oblique lip line, could not be considered as HFM because our patient had a downward oblique lip line deviating to the right contrary to the clinical appearance of the patients with HFM. Furthermore, our patient had a complete ear with normal hearing. Radiographic images also showed that our patient's affected side showed no sign of under-development of the TMJ, ramus, mastication muscles, and ear.

CFCs usually involve both bone and soft tissue deformities. Duplication of the jaw occurs more in the maxilla than the mandible which is commonly associated with cleft lip and palate. In some cases, the presence of a supernumerary alveolar process which may contain supernumerary teeth and commonly seen in the molar areas was observed.[21] Bone cleft of Tessier cleft 5 is located distal to the canine on the premolar area traversing the maxilla lateral to the infraorbital foramen through the middle third of the orbital rim to enter the orbit.[21] The case presented here had no extraoral soft tissue manifestations such as cleft lip but enlarged maxillary alveolar process and alveolar cleft resulting in a double maxilla were observed. This case different from the previously reported cases of Tessier cleft 5 that manifested external soft tissue deformities. In reported cases of Tessier cleft 5, supernumerary teeth were also observed to be included with either duplication of the jaw or supernumerary alveolar process.[21] This patient present a full set of vital permanent dentition and supernumerary teeth with normal forms on the maxilla, hence, overcrowding was still observed in addition to the size of the osseous structure of the maxilla.

Malformations due to CFCs are being managed by surgical corrections, except for loss of sight and hearing. Speech problems are being addressed through speech therapy. Achieving a high success rate depends on the time of surgical intervention. The later the intervention, the more effect it has on craniofacial development. Management of CFCs needs a multispecialty medical team to perform the multistage treatment.[8] Depending on severity, CFCs are managed in a few months after birth or after a year. Severe or extreme CFCs are recommended to be treated in the first few months. In not severe case, it can be waited for a little longer. Treatment also includes reconstruction and/or repositioning of displaced structures because of the cleft such as the skeletal malformations.[5]

The success of surgical intervention is also dependent on diagnostic aids such as radiograph investigations. In addition to the commonly used intraoral and panoramic radiographs, more sophisticated radiographic imaging techniques are already available such as CBCT which is helpful in diagnosis and treatment planning for

maxillofacial applications. CBCT image of Tessier cleft 5 can just be a narrow furrow traversing the anterior maxilla or a broad maxillary cleft lateral to the maxillary sinus and infraorbital foramen. The latter may continue to the inferolateral orbital rim and floor with no inferior orbital fissure communication at the posterior. The lateral maxillary part is collapsed with a reduction in the maxillary arch transverse dimension. It also manifests in the sphenoid as shortening and thickening of the lateral orbital walls on the area of the greater wing. Pterygoid plates are also mildly displaced relative to the midline. The cranial base and calvarium are minimally displaced.[13] These abnormalities could not be easily detected clinically, thus radiographic investigations are needed not only for diagnosis but also for treatment planning. The CBCT images of this present case conformed to the skeletal descriptions of David *et. al.*[13] on Tessier cleft 5. In comparison to the radiographic images of HFM, our case showed that the structures on our patient's affected side presented no underdevelopment.

Adequate and appropriate imaging techniques, with emphasis on 3D imaging, are very important for proper diagnosis and treatment planning of CFCs especially when there is no external soft tissue manifestation. Lack of proper radiographic imaging may result in inadequate diagnosis and treatment planning. Facial reconstruction was not done in this case because there was no significant extraoral manifestation of the cleft, moreover, the patient's cleft was found after CBCT imaging and it did not affect the patient's functions. However, pre-orthodontic procedures, such as bone grafting, were done for his current orthodontic treatment to correct the patient's malocclusion.

CONCLUSION

Tessier cleft 5 is a rare oblique cleft, which is also uncommon among craniofacial clefts, and affects patients' health. The findings of the case presented here suggest that CBCT is a good modality of choice for diagnosis and successful management of Tessier cleft 5 with no extraoral soft tissue manifestations related to the affected side. CBCT helps the medical team in planning reconstruction procedures, both soft and hard tissues. Therefore, the CBCT scan is recommended for the diagnosis and treatment planning of CFCs. Further studies are still needed to be done about CBCT analysis on the characteristics of the phenotypes of Tessier cleft 5.

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