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Dental Care for Kabuki Syndrome Patient: A Case Report

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Abstract

Index terms—

1 I. INTRODUCTION

Kabuki syndrome (KS) was described in 1981 at two Japanese centers in the Kanto area and Hokkaido [1] **??**2]. It is a rare genetic disorder (congenital distortion syndrome) [3] which characterized by multiple congenital anomalies and mental disability [4] [5]. Proportion of Kabuki Syndrome is around 1/32,000 of births [3]. The main cause of Kabuki Syndrome is unknown. However, X-linked and autosomal dominant gene have been suggested [6] [7] [8].

Whereas KMT2D (MLL2) gene mutations were observed in most patients with Kabuki Syndrome, and a few have mutation or deletion of KDM6A [3]; thus the first pathogenic gene recognized in Kabuki Syndrome patients was KMT2D according to Ng, S.B., et al., study [9], But nearly in 30% of patients with Kabuki Syndrome, the Potential genetic defects are still unknown [3].

Diagnosis of the syndrome is based on 5 main clinical features: (1) a special face (100%) which tip and long palpebral fissures with eversion of the lateral third of the lower eyelids, higharched eyebrows with sparse lateral one-third, (2) Growth deficiency (83%) with short stature, (3) moderate to severe mental disability (92%), (4) skeletal anomalies (92%) and (**??**) abnormalities of dermatoglyphic (93%) [7] [10].

The face of patients with Kabuki syndrome is similar to the makeup worn by actors of Kabuki; a Japanese traditional play. That's why it's called Kabuki syndrome [8].

Other important clinical features have been reported include: early puberty, premature breast development in girls, anal atresia, congenital heart disorder, craniofacial anomalies, gastrointestinal anomalies, fingers abnormalities (Short fifth fingers), dental anomalies, [6] [7] [4] [5] [10] and renal and vertebral anomalies [3].

The most frequent oral manifestations reported were: cleft lip/palate; bifid tongue and uvula; malocclusion (micrognathia, severe maxillary recession, mid-facial hypoplasia, high-arched palate, widely spaced teeth); delayed tooth eruption pattern; dental abnormalities (hypodontia, conical teeth, neonatal teeth, large pulp chamber); diastema and lower lip pits [11] [12] [8].

2 Intra-Oral Examination

In intraoral examination; no abnormalities were observed in lips, tongue and oral mucosa.

The patient was in primary dentition stage; with edge-to-edge bite, high-arched palate, in the upper arch the present teeth were primary central incisors, primary canines, first and second primary molars, the upper incisors as 'flat head' screwdriver-shaped, in the lower arch the present teeth were primary central and lateral incisors, primary canines, first and second primary molars.

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The primary maxillary lateral incisors were absent (hypodontia) with interdental spacing.

Carious cavities were seen in the mandibular first and second primary molars.

4 Radiographic Findings

A panoramic photo (Figure . 4) showed carious mandibular second primary molars. The primary maxillary lateral incisors (previously noted as absent), the maxillary permanent incisors incisors buds were absent, while

the maxillary permanent second and third molars and the mandibular third molars buds were not considered as absent because they need time to develop.

According to the American Association of Pediatric Dentistry (AAPD), this case is classified as Early Childhood Caries (ECC) [13].

5 Treatment

The girl's behavior was Negative (Reluctance to accept treatment, uncooperativeness, some evidence of negative attitude but not pronounced (sullen, withdrawn) according to the Frankl behavior rating scale [14].

The dental treatment was accomplished under intravenous sedation (one session) by an anesthesiologist in the oral and maxillofacial surgery hospital, Damascus University.

Whereas a medical specialist consultation for sedation had been requested, and pre-sedation dietary instructions which determined by the American Academy of Pediatric Dentistry had been given: (1). Clear liquids: up to 2 hours before the procedure. (2). Breast milk up to 4 hours before the procedure. (3). Infant formula, nonhuman milk or a light meal up to 6 hours before the procedure [15]. And the medication used in IV sedation was (Midazolam 1.

6 IV. DISCUSSION

Kabuki syndrome is considered as a rare condition; although dentists may find difficult to understand the case, but they may contribute to the diagnosis, so it is important to know the facial and oral clinical manifestations accompanying syndrome to request further examinations when noticing any changes in the normal state

The etiology of the Syndrome is unclear, and diagnosis is clinically and mainly based on facial features in addition to other clinical features: Growth deficiency, mental disability, skeletal anomalies, abnormalities of dermatoglyphic [4].

Typical facial features can be identified from an early age to help in clinical diagnosis. However; clinical identification of the syndrome in the neonate is difficult, maybe the phenotype is developed by the time [12].

The patient in this case has a short stature, craniosynostosis with Microcephaly, short finger, special facies consisting of narrow Front, high-arched eyebrows with sparse lateral onethird, elongated palpebral fissures, eyes with eversion of the lateral one-third of the lower eyelids, Small eyeball, prominent ears, broad depressed nasal root with flat nasal tip. These manifestations also reported by **??**etzold et [16], [12], [17], [18], [10], [19].

Dental abnormalities have been reported in over 60% of patients with Kabuki Syndrome [11] [17] [20]. The most common dental finding was the hypodontia [8], in this case Maxillary primary lateral incisors, the maxillary permanent lateral incisors buds, mandibular permanent central incisors buds, and the right mandibular permanent lateral incisors buds were absent, these findings about missing teeth are in agreement with the literature reported by **??**hanni et [21], [11], [16], [12], [17], [8], [20]. The finding of absent premolars or molars as described by Mhanni et al., Tuna et al., and do Prado Sobral et al., were not observed [21], [20], [4].

Space between maxillary teeth that was found in this case is associated with hypodontia, and this characteristic was reported previously by Petzold et al., [16], in addition to the high-arched palate which also observed by Matsune et al., and do Prado Sobral et al., [11], [4].

The upper incisors were as 'flat head' screwdriver-shaped, this finding was reported by **??**hanni et [21], [16], [12], [4], [18], whereas Dental shape abnormalities were not observed in Teixeira et al., who study dental examination and panoramic radiography of nine patients [8]. There are no lips, oral mucosa or tongue abnormalities observed in this present case.

Pediatric dentists should choose the best behavioral management technique that fit the patient status as well as the procedure nature which needs to be accomplished, and they have often found that anxiety and behavioral assessment to be helpful in determining the behavioral management technique to be chosen for each child [15].

Sedation requires an accurate medical history accomplishment to determine whether the patient a good candidate to it or not. The American Society of Anesthesiologists (ASA) guidelines are considered as the most accurate method when taking patients medical history [15], and case in this report is classified as ASA class II that is frequently considered appropriate candidate for minimal, moderate, or deep sedation. However, counsel with an anesthesiologist is often desired [15].

While IV Sedation can be a suitable alternative to general anesthesia for children with ECC and the equipment to provide general anesthesia is far more expensive than what is required for IV sedation [22], in addition to the girl's behavior in this case was negative according to the Frankl behavior rating scale [14] as well as the treatments were required enough time to be accomplished, therefore dental treatment in this case was done under intravenous sedation.

7 V. CONCLUSIONS

The dental manifestations observed in this case were hypodontia, abnormal teeth morphology and high-arched palate.

These dental abnormalities in addition to another clinical features may help in the clinical diagnosis of the syndrome, so It is important that dentist be aware of this syndrome and its facial manifestations and oral/dental findings to recognize children who may be affected by this disorder.

8 REFERENCES

Figure 1: Figure 1 : 7 Dental

[1] [2] [3] [4]

[1] © 2023 Great] Britain Journals Press Volume 23 | Issue 7 | Compilation 1.0
[2] © 2023 Great] Britain Journals Press Volume 23 | Issue | Compilation 1.0
[3] Dental Care for Kabuki Syndrome Patient: A Case Report
[4] © 2023 Great] Britain Journals Press Volume 23 | Issue 7 | Compilation 1.0

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Figure 2: Figure 2 :

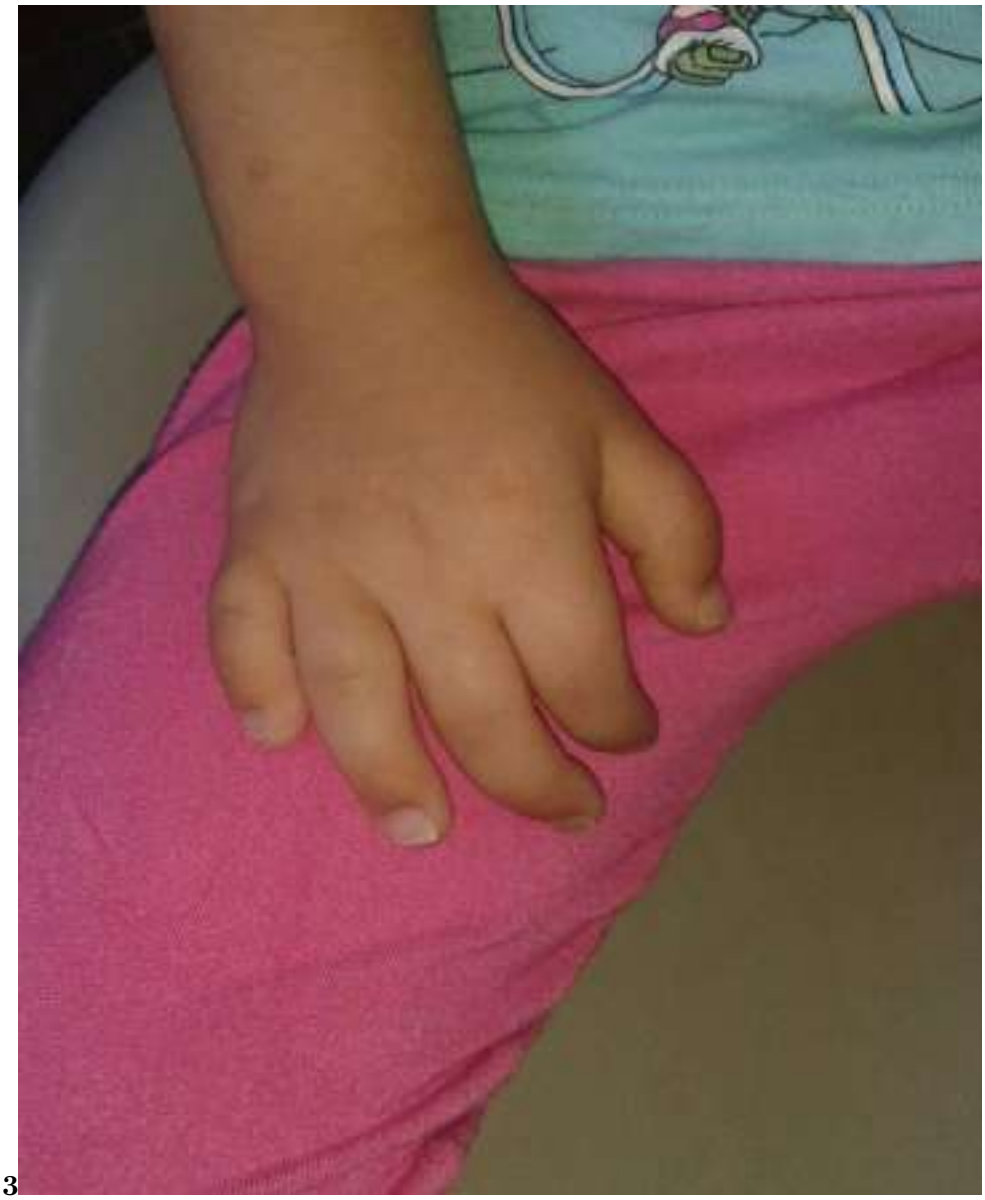
3 

Figure 3: Figure 3 :

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Figure 4: Figure 4 :

Figure 5: 7 DentalFigure 5 :Figure 6 :

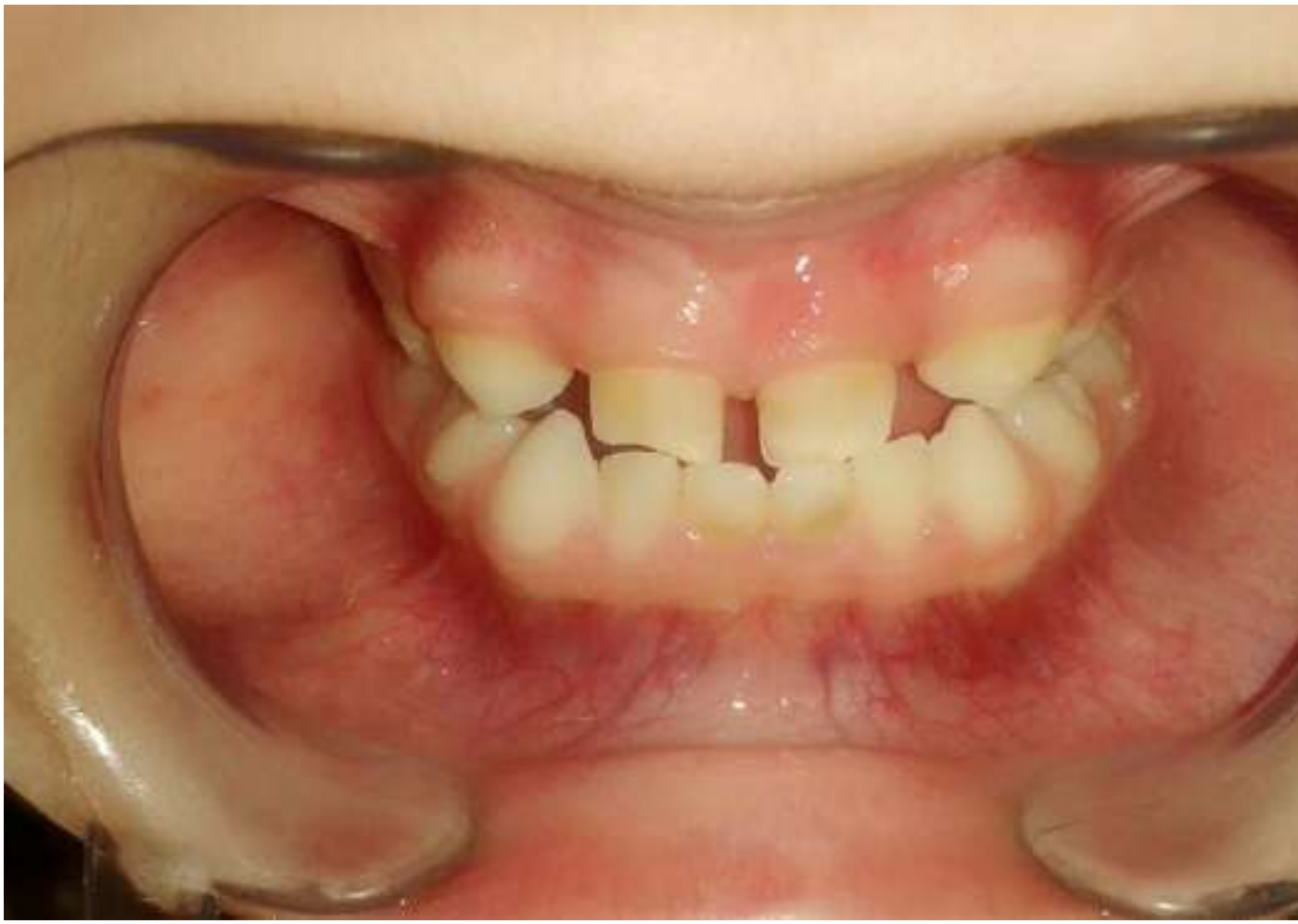

Figure 6:

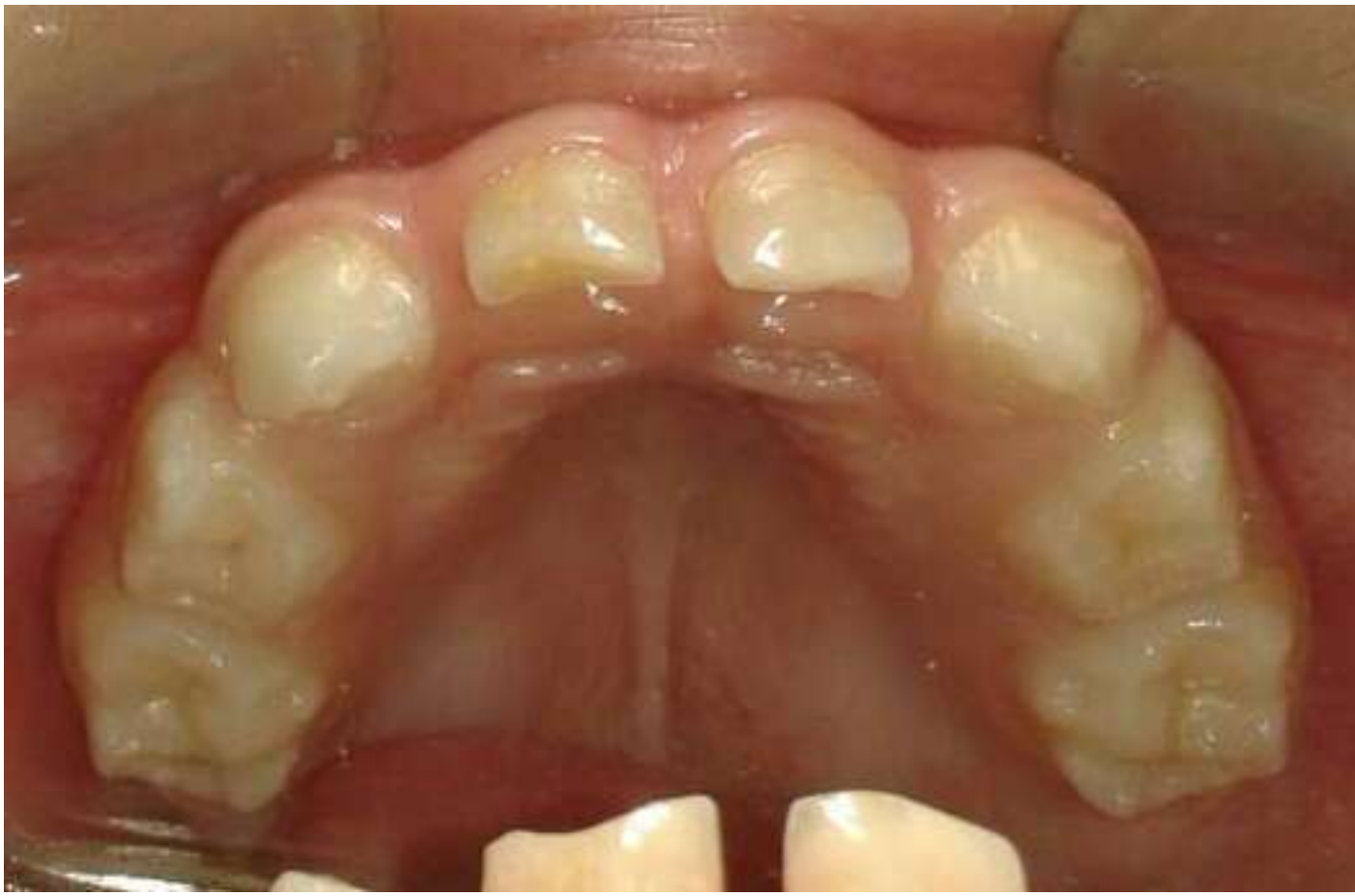

Figure 7:

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Figure 8:

Figure 9:

Figure 10:

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Figure 11:

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