Oral Manifestations of Keratosis Ichthyosis Deafness (KID) Syndrome

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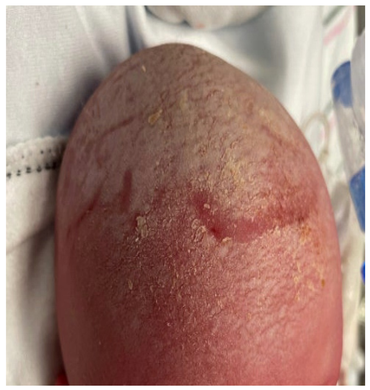
BACKGROUND

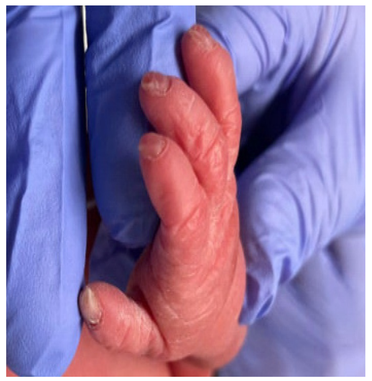
Keratosis Ichthyosis Deafness (KID) Syndrome is a rare genetic disorder present at birth that is caused by a gap junction protein beta 2 (GJB2) gene mutation. The impairment of the gap junction function results in cell miscommunication within the skin and other tissues. Clinically, KID is consistent with corneal defects (keratitis), abnormal epidermal cell function (ichthyosis) and sensorineural deafness. Other symptoms including alopecia, recurrent infections, abnormalities in dentition and impaired sweat gland function are often observed. Additionally, individuals with this disease have an increased risk of developing squamous cell carcinoma of mucus membranes and skin due to the increased cell turnover.

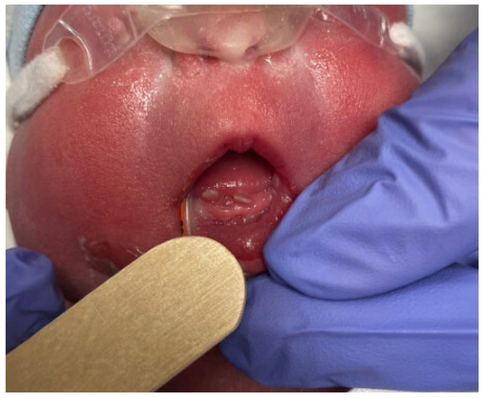
CLINICAL PRESENTATION

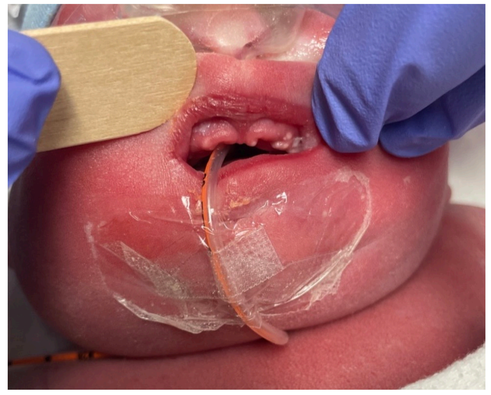
A 2 day old (former 32 week + 1 day old) female patient with KID syndrome was transferred overnight to Riley Children's Hospital due to metabolic acidosis. The patient was admitted into the Riley Level 3 NICU where she was receiving ongoing respiratory, nutritional and thermoregulatory support. Additionally, genetics and dermatology were consulted due to the presence of dysmorphic features. Pediatric dentistry was emergently consulted for oral evaluation and concern of loose mandibular natal teeth posing a high risk of aspiration. Clinical examination revealed multiple small (< 3 mm) well circumscribed white nodules present on the entirety of the maxillary alveolar ridge and the posterior mandibular alveolar ridge bilaterally. Two natal teeth were present in the mandibular anterior (#O/P) region embedded in soft tissue with Class III mobility. Radiographs were not able to be obtained due to the patient's medical status, intubation, and significant mobility of the natal teeth. This report will include clinical findings associated with KID Syndrome, differential diagnoses, and treatment provided.

CLINICAL PHOTOGRAPHS









DIFFERENTIAL DIAGNOSES

- **Maxilla**
  - Palatal Cysts of the Newborn
    - Bohn's Nodules: minor salivary gland remnants on the buccal/lingual alveolar ridge
    - Epstein Pearls:
  - Gingival Cysts of the Newborn
    - Dental Lamina Cyst: dental lamina remnants located on the crest of the alveolar ridge
- **Mandible**
  - Natal teeth- present at birth

TREATMENT/MANAGEMENT

Due to the significant risk of aspiration, it was determined the mandibular natal teeth should be promptly extracted. The patient was hemodynamically stable enough for natal tooth extraction. The patient remained in isolette due to NICU precautions and their intubation status. No anesthetic was used. 2 x 2 gauze was used as a throat shield. Routine dental extraction of the natal primary mandibular central incisors was performed without complication. Pressure was applied and hemostasis obtained. The patient's genetic results were consistent with KID syndrome. The parents and medical providers were instructed to monitor the nodules present as they were benign and self-limiting in nature and to periodically reassess for further eruption of natal/neonatal teeth.

FOLLOW UP

At the time dental treatment was rendered, the genetic workup for this patient was incomplete. The genetic results were determined in the days following dental consultation and were consistent with KID syndrome. Additionally, the parents and medical providers were instructed to periodically reassess for further eruption of neonatal teeth (within first 30 days). Due to patient's medical complexity, the patient's prognosis was poor and the life expectancy for this patient at that time was limited.

REFERENCES

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