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"Multidisciplinary Management Of A Complex Case Of Goldenhar Syndrome With Craniofacial Anomalies And Airway Complications"

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Abstract:
Goldenhar syndrome, or Oculo-Auriculo-Vertebral Spectrum, is a rare congenital disorder characterized by craniofacial anomalies, ear deformities, ocular defects, and vertebral malformations. This report presents a complex case of a newborn diagnosed with Goldenhar syndrome, exhibiting multiple craniofacial abnormalities, including ear tags, a displaced lower jaw, and a mass in the right eye. Laboratory findings revealed low hematological parameters. At three years old, the patient underwent bilateral mandibular distraction to correct the jaw deformity and improve airway function. Postoperatively, airway complications led to the need for a tracheostomy. The patient was discharged with a multidisciplinary care plan, including pediatric surgery, ophthalmology, and otolaryngology. Medications included antibiotics, multivitamins, gastroprotective agents, and nebulized bronchodilators. This case highlights the importance of coordinated, long-term management for individuals with Goldenhar syndrome, addressing both immediate and progressive challenges associated with the condition.

Key-words: Goldenhar syndrome, Craniofacial anomalies, Mandibular distraction osteogenesis, Multidisciplinary management

Introduction:
Goldenhar syndrome (Oculo-Auriculo-Vertebral Spectrum) is a rare congenital condition. The syndrome involves malformations primarily affecting structures derived from the first and second branchial arches, leading to facial asymmetry, ear anomalies, ocular defects, and vertebral malformations. The etiology of Goldenhar syndrome remains uncertain, although genetic and environmental factors are thought to contribute to its development.
This report details a complex case of Goldenhar syndrome in an infant presenting with multiple craniofacial anomalies, low hematological parameters, and airway complications, managed through a multidisciplinary approach involving pediatric surgery, ophthalmology, and otolaryngology.

Case History:
A newborn was delivered with severe craniofacial anomalies, including multiple facial ear tags, a mass growth in the right eye, and facial deformity(fig1,2). The patient had a displaced lower jaw, abnormal ear development, and a mass growth in the right eye, according to a general physical examination. Low levels of hemoglobin, total cell volume (TCV), red blood cell (RBC), eosinophils, and absolute lymphocyte count were found during laboratory testing. The patient was diagnosed with Goldenhar syndrome (oculo-auriculo-vertebral spectrum) based on a clinical evaluation. A pediatric surgeon was suggested for follow-up care by the doctor.

A cornea specialist suggested patching the left eye (LE) for two hours a day to treat amblyopia brought on by the right eye tumor after a one-year follow-up. Given the patient's condition at three years old, the surgeon underwent a general anesthetic procedure called bilateral mandibular distraction in order to address the lower jaw deformity and enhance airway patency. The procedure went well, and there was no immediate complications throughout the recovery phase. But after being taken out of the air, the patient became unwell and needed a tracheostomy. A bronchoscopy identified an obstruction of the upper airway. With orders for further care and a tracheostomy, the patient was released from the hospital.

The following medications were prescribed upon discharge:
· Syrup Taxim-O Forte (Cefixime) 100 mg/5 ml, to be taken for 5 days
· Syrup A to Z (multivitamin), to be taken as per instructions
· Tablet Junior Lanzol (Lansoprazole), for 5 days
· Nebulization with Budecort (Budesonide) 0.5 mg, and Duolin (Ipratropium bromide and Salbutamol) as needed, for 5 days

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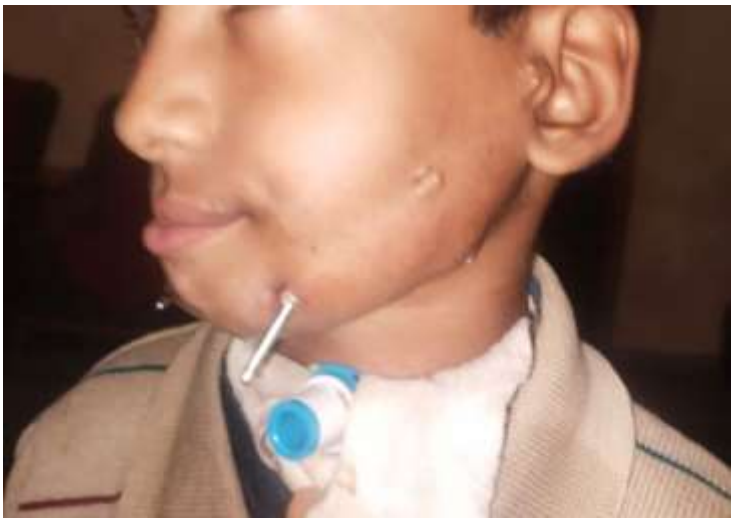
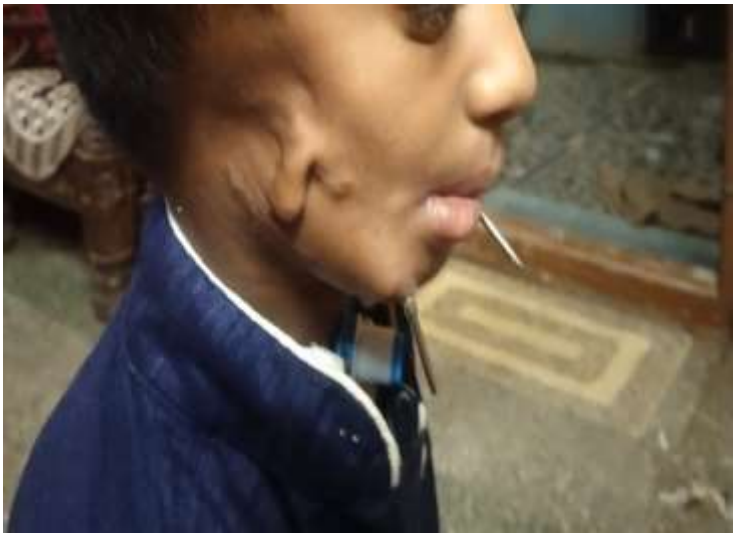
FIGURES:





Discussion:
As demonstrated in this instance, the eyes, ears, spine, and facial bones can all be affected by the vast spectrum of clinical signs of goldenhar syndrome. The baby had characteristic symptoms such as mandibular hypoplasia, ear tags, a mass development in the right eye, and facial deformity. An underlying hematological involvement—which is infrequently described in syndromic cases—was suggested by the low counts of hemoglobin, RBC, TCV, eosinophils, and absolute lymphocytes 5*.

Because of the development in mass in the right eye, the first management included frequent follow-up and patching therapy to prevent amblyopia2*. In order to address mandibular hypoplasia and enhance airway patency, a bilateral distractor insertion procedure was carried out under general anesthesia on a three-year-old . A tracheostomy was necessary due to post-operative respiratory distress issues, indicating the possibility of airway difficulties in these individuals 4*.

A comprehensive approach is often necessary for the successful care of Goldenhar syndrome3*. In order to properly handle difficult patients, this example emphasizes the value of early intervention and collaboration among pediatric surgeons, ophthalmologists, otolaryngologists, and other specialists. Following tracheostomy, the post-operative period was uneventful with appropriate airway control. The patient was discharged with the prescribed medicine and instructions for aftercare.

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