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Pierre Robin Syndrome: A Case Report

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ABSTRACT

Pierre-Robin syndrome (PRS) is characterized by micrognathia, ptosis, and palatal malformations. Infants often have immature mandibles and difficulty breathing. A small lower jaw pushes the tongue backward, leading to PRS. A wide, U-shaped cleft palate is also commonly associated with this abnormality. PRS is not just a syndrome, it is a range of disorders, one abnormality leading to another. We report the case of a 10-day- old neonate who complained of malnutrition and dyspnea and was later diagnosed with Pierre-Robin syndrome (PRS).

KEYWORDS: *Cleft palate, Micrognathia, Glossoptosis, Pierre Robin Syndrome*

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INTRODUCTION

Pierre Robin Syndrome (PRS) is characterised with the aid of using triad of micrognathia, glossoptosis and cleft palate. Pierre Robin Sequence is taken into consideration to be a nonspecific anomalad which can also additionally arise both as an remoted disorder or as a broader organization of malformations. In 1923 French medical doctor Pierre Robin brought the term `glossoptosis` in affiliation with micrognathia. In 1934 he suggested an affiliation with cleft palate and this constellation of findings changedinto later termed as syndrome. The Pierre-Robin Syndrome (PRS) is a unprecedented malformating pathology and its anticipated frequency is about 1/30000. Some familial instances have additionally been suggested, which can also additionally suggest that a few instances have an inherited basis.

Case Description

We report the case of a 10-day-old neonate of AGA (of childbearing age) of35 ± 2 weeks' gestation/(2.35 kg) LBW (low birth weight), born to a 27-year-old mother. The patient presented to the Department of Pediatrics, Saveetha Medical College and Hospital, Chennai, Tamil Nadu, with symptoms of feeding and breathing difficulties.

About collecting exhaustive and detailed histories, It turned out that the mother had complained of hyperemesis gravidarum early in her pregnancy. Her prenatal ultrasound revealed oligohydramnios with an amniotic fluid index (AFI) of 6. Upon careful examination of the patient, micrognathia, a "U"-shaped cleft palate, and ptosis were observed (Figure 1, Figure 2, Figure 3). With a head circumference of 32 cm, a length of44 cm, and a chest circumference of 29 cm, she had a new Ballard score of 32.

Respiratory examination observed mild intercoastal and substernal recession. On examination of the central nervous system, the newborn's reflexes were blunted. No murmurs were detected on cardiovascular examination. Sepsis screening was normal.

Two-dimensional echocardiography was found to be normal. Ophthalmologic examination was normal. Breastfeeding in a prone or lateral position provides comfort to the patient. In our case, noartificial airway was required. No family history was reported and was the first birth orderwithout siblings.

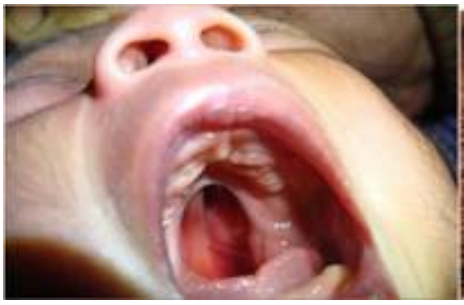

Figure 1: Showing U shaped claft palate Figure 2: Showing micrognathia Figure 3: Showing glossoptosis

Discussion

Pierre Robin syndrome, also known as Pierre Robin chain, or Robin anomaly, is characterized by varying degrees of severity of micrognathia, bullous disease, and palatal deformity. In this malformation, the size of the tongue remains normal but bilateral asymmetry due to microbiology, increased opacity is often observed.

The word "sequence" suggests that one abnormality leads to the next, and micrognathia is considered to be the cause abnormality in PRS patients. Airway obstruction and feeding difficulties occur and the severity of problems varies.

PRS can occur alone or in combination with other syndromes such as Stickler syndrome, myocardial syndrome. Therefore, a geneticist should be consulted to find out the likelihood of this occurring in future children. In about 30% of cases, PRS may be a single event, while in the next 30%, it is associated with other malformations and in thelast third of cases it is part of a complex syndrome. more complex (usually Stickler syndrome). This diversity of manifestations is the result of a mixed genetic origin: in 40% of cases, PRS is genetically isolated, otherwise it is autosomal recessive or autosomal dominant. In our case, no association with other abnormalities was observed.

Whatever the cause, infants and infants with Pierre Robin sequence can have varying degrees of airway obstruction and feeding difficulty. PRS can be life-threatening in the neonatal period with the onset of airway obstruction, which can occur any time soon after birth. If left untreated, persistent airway obstruction can lead to acute or chronic hypoxia, cyanosis, apnea, aspiration, respiratory infection, difficulty feeding, malnutrition, and growth retardation. . Later complications of chronic hypoxia are chronic carbon dioxide retention, increased pulmonary vascular resistance, arrhythmias, right-sided heart failure, and cerebral hypoxia.

Most patients with PRS are in remission with conservative measures, but those with marked micrognathia, growth retardation, prolonged nasopharyngeal use, or prolonged intubation require surgical correction. As in our case, conservative measures were sufficient to control the infant's symptoms.

Conclusion

All cases of PRS should be thoroughly investigated to diagnose association with other syndromes and to determine further treatment strategies. Each individual case of PRS is unique and should be evaluated individually. It is our responsibility as physicians to recognize this disorder in a timely manner and to provide closefollow-up and appropriate treatment and advice.

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