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Case Report

Craniofacial Dysostosis Associated to Sars-Cov-2. A Case Report

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Abstract

Craniofacial dysostosis refers to the syndromic forms of craniosynostosis, characterized by a sutural abnormality with cranial vault involvement that extends to the skull base and the middle third of face structures.

Clinical manifestations reports of SARS-CoV-2 in pregnant women are limited, therefore this manuscript objective is to report a clinical case of a newborn with craniofacial dysostosis associated with SARS-CoV-2 infection during pregnancy.

Keywords: Craniofacial Dysostosis; Pregnancy; SARS-CoV-2

Introduction

Craniofacial dysostosis refers to the syndromic forms of craniosynostosis, which are characterized by a sutural involvement that, in addition to affecting the cranial vault, extends to the base of the skull and the structures of the middle third of the face [1]. Carpenter, Apert, Crouzon, Saethre-Chotzen, Muenke and Pfeiffer have described craniofacial dysostosis syndromes [2].

In addition, Severe Acute Respiratory Syndrome Coronavirus 2 (SARS-CoV-2) has represented a public health emergency worldwide that has brought with it considerable morbidity and mortality rates for the population. Among its clinical manifestations, pulmonary and extrapulmonary repercussions have been found, with signs and symptoms of damage to the multi-organ system [3].

The data reported on SARS-CoV-2 in pregnancy is limited. Some complications such as placenta previa [4], preeclampsia [5], asphyxia and fetal death have been described [6,7].

Objective of the Study

The objective of this study is to report a case of a newborn with severe craniofacial dysostosis associated with SARS-CoV-2 during pregnancy.

Case Report

A 1-month-old female patient was taken to the Cleft and Craniofacial Surgery Unit of the Coromoto Hospital of Maracaibo - Venezuela, presenting severe craniofacial dysostosis. During the anamnesis, the 31-year-old mother referred to be a primigravida, with a history of SARS-COV-2 at the fourth week of gestation, confirmed by PCR. The last ultrasound at 38 weeks plus 6 days of gestation reported severe polyhydramnios, fetal skull malformation with a bilobed "cloverleaf" appearance, with moderate dilation of the frontal horns of the lateral ventricles, as well as hypoplasia of the nasal bones and short limbs. The extraoral clinical evaluation of the infant revealed a "cloverleaf" skull, bilateral ocular proptosis and severe deficiency of the midface (Figure 1). Intraorally, a type II palatal cleft was evidenced according to Veau (Figure 2). The patient was referred to genetic studies to establish a final diagnosis. The patient died so the final diagnosis wasn't established.

Figure 1: Extraoral photographs, a "cloverleaf" skull, bilateral ocular proptosis and severe deficiency of the midface was evidenced.

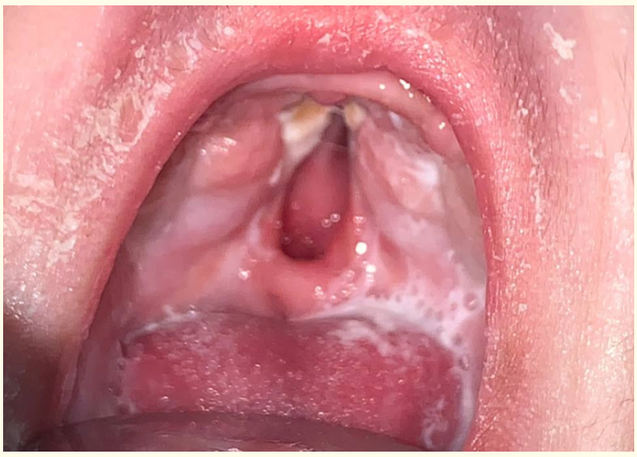

Figure 2: Intraoral photography, cleft palate was evidenced.

Discussion and Conclusion

The term "craniofacial dysostosis" is used to describe the syndromic forms of craniosynostosis. There are multiple clinical characteristics in craniofacial dysostosis syndromes. The Kleeblattschadel anomaly (cloverleaf skull) is frequently evidenced in these disorders [8,9]. Each syndrome is phenotypically different; however, all patients show varying degrees of supraorbital rim retrusion and midface hypoplasia with weak infraorbital support, resulting in shallow orbits and a high risk of exposure keratopathy that can lead to blindness. Craniofacial anomalies such as cleft palate, mandibular growth issues, stylohyoid calcification and other cranial suture affectations are common [10]. In our case, the patient presented a cloverleaf skull, severe midface deficiency, bilateral proptosis and a type II palatal cleft, which are characteristics evidenced in Crouzon syndrome, the most common disorder related to craniofacial dysostosis. In contrast to the Apert and Carpenter syndromes, the patient under discussion did not present syndactyly. In the same way, she did not present finger deformities such as those evidenced in Pfeiffer syndrome [11]. However, a genetic study is always required to determine the diagnosis.

Regarding the congenital anomalies evidenced in craniofacial dysostosis syndromes, increased intracranial pressure is the most serious problem associated with craniosynostosis. This increase in intracranial pressure can lead to papilledema and, eventually, optic nerve atrophy resulting in partial or total vision loss. Similarly, syndromic proptosis can result in abrasions or ulcerations of the eyeball due to corneal exposure. Breathing could also be affected in these patients due to midface hypoplasia, where the nasal and nasopharyngeal spaces are diminished, causing obstruction of the nasal airways. In turn, the patients become mouth breathers, which bring various morphological and functional alterations in the oral cavity. Hearing deficits are also common in patients with craniofacial dysostosis syndromes [12].

The complications of SARS-CoV-2 during pregnancy have been poorly described. This entity infects humans through the receptor for angiotensin-converting enzyme 2 (ACE2), which is highly expressed in the alveolar epithelium and is also present in various cells of the fetal tissues and the maternal-fetal junction. The expression of ACE2 in the female reproductive tract and placental tissue suggests that SARS-CoV-2 infection can compromise pregnancy outcomes and be vertically transmitted to the fetus [13]. Some authors [14,15] have reported an increased risk of preterm delivery and low birth weight, particularly when the infection occurs at 20 weeks' gestation or later.

Juan., *et al*. [16] in a review that included 324 pregnant women diagnosed with SARS-CoV-2, found 4 cases of spontaneous abortion, 4 cases of intrauterine fetal deaths, and 3 cases of neonatal death. On the other hand, Khalil., *et al*. [17] evaluated 746 deliveries from SARS-CoV-2 positive pregnant women, where there were 0.9% maternal deaths, 21.8% premature births and 1% perinatal deaths.

Regarding congenital alterations in relation to SARS-CoV-2, there is limited evidence in the literature. Luteinj., *et al*. [18] reported that viral diseases during pregnancy in early stages and some antiviral drugs are related to an increased risk of neurological congenital anomalies for the neonate, such as neural tube defects, spina bifida and brain disorders such as anencephaly, encephalocele or hydrocephalus. Khan., *et al*. [19] indicated that SARS-CoV-2, being able to enter the placenta and the nervous system, may have an impact on the pathogenesis of neural tube defects. In addition to this, the embryonic cells responsible for organogenesis may be affected and lead to developmental abnormalities.

It is extremely important to continue studying the pathophysiology of SARS-CoV-2 and its relationship with the development of craniofacial dysostosis during pregnancy.

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