CASE REPORT

Cleft palate lateral synechia syndrome: Inhereditary or not?

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1. Introduction

Cleft lip and palate is one of the common congenital abnormalities in the head and neck1,2 constituting 65% of head and neck anomalies in isolated or syndromic forms.3 Each year about 150,000 infants are born with cleft palate in the world.4 In 1972,5 described a novel syndrome consisting of cleft palate (CP) and lateral synechia (LS) between the palate and the mouth floor. So far, this constellation of malformations has been denoted as cleft-palate lateral synechia syndrome (CPLS). Although cleft palate is a congenital anomaly with a relatively high incidence, there are few cases in which it is accompanied by oral synechia due to cord-like adhesions. Other symptoms include cleft palate, mouth adhesions, and inability to fully open the mouth. Release of the synechia was performed without complications. Although some researchers believe CPLS is inherited in an autosomal recessive pattern, our case indicated that a new mutation may cause this situation. Further research is recommended to clearly determine its inhereditied and phenotypic variabilities.

2. Case report

A 12-week-old female infant was presented in the Tabriz Pediatrics Hospital, Tabriz, Iran for evaluation of a cleft palate. She had a wide unilateral left cleft palate and a lateral synechia. The mucosa-lined, fibromuscular tissue bands were connected from the lateral free border of the cleft palate to the mouth floor in the left side of the tongue, so she was unable...
to fully open her mouth (Figs. 1 and 2). Except for the shortened superior labial frenulum, no other anomalies in craniofacial, digital, genitalia or limbs were observed. Her twin brother did not show any similar anomalies.

The case was preterm (34 weeks) and was hospitalized for 11 days after birth due to respiratory distress. Her birth weight was 1200 g. No history of cleft palate or other anomalies was reported in her family. Her parents were not attributed, too. Her mother had no history of smoking, however she had to take atenolol for her gestational hypertension during her pregnancy. Release of the synechia and her short frenulum was performed by cauterization in the operation room without any complications. As a result, the patient could fully open her mouth with no difficulty. She was discharged on antibiotics and anticipation for the proper time to undergo an operation on cleft palate.

3. Discussion

Interalveolar adhesions are usually associated with such congenital syndromes as the Van der Woude syndrome, Popliteal Pterygium syndrome, Orofacial Digital syndromes, and cleft palate lateral synechia (CPLS) syndrome. As a rare condition, in some studies, CPLS supports the suspected pattern of autosomal dominant inheritance with variable expressivity. Guion-Almeida and Nakat examined three Brazilian children affected with LSC. One patient was an isolated case while the other had an equally affected brother. Genetic aspects and phenotypic manifestation were compared with those observed in the patients reported with oral synechia. They were diagnosed for autosomal recessive inheritance of this syndrome due to the recurrence in siblings. Congenital interalveolar synechia frequently shows syndromic associations despite the occurrence of isolated instances. In a report by Tanrikulu et al. (2005), the patient was a 10-month-old female infant with isolated congenital alveolar synechia without any other anomalies.

Since cleft palate with oral synechia is among the rarest congenital deformities, the midline synechia is less common than lateral ones. Several case reports were published on the midline oral synechia with epithelial cyst in neonate and the cleft palates. In a study, the researchers reported on one case of oral synechia by a single medial cord-like adhesion accompanied by the cleft palate, micrognathia, ankyloglossia, and shortening of the superior labial frenulum. They tentatively named this rare condition as “cleft palate medial synechia syndrome”. In another study, it was suggested that CPLS might represent a mild phenotypic expression of the Fryns syndrome influenced by environmental factors like temperature, viral infection or stochastic events.

Fryns syndrome should be considered in the diagnosis of all patients with the combination of alveolar synechia, cleft palate, and distal digital hypoplasia. Clinicians are recommended to expect the phenotypic variability in counseling and caring for the families of the children with CPLS.

In this article, a 12-week old female infant was presented with CPLS and lateral synechia syndrome with no other anomalies. She had a healthy twin. No histories of cleft palates existed in her family. The chances were that she had a new mutation. On the other hand, since she had a healthy twin, the effects of environmental factors on this manifestation are under question. Further research is highly recommended to study the origin of the lateral synechia syndrome in this case.

4. Conclusion

As discussed earlier, some researchers believe that CPLS is inherited in an autosomal recessive pattern, but our case indicated that a new mutation may lead to this situation. Further research is recommended to determine inheridity and phenotype variability in similar cases.

Conflict of interest

We have no conflict of interest to declare.

References


Figure 1 Wide cleft palate is shown in the figure.

Figure 2 A fibromuscular band is shown from the left cleft edge to the floor of the mouth.


