Tessier number 30 cleft: report of an antenatally diagnosed case

Abstract: Tessier no. 30 cleft, also known as median mandibular cleft or lower midline facial cleft, is extremely rare. Less than 100 cases are reported in the literature, which show great variation in severity and associated abnormalities. We herein report a median mandibular cleft extending into the neck. Its importance comes from the fact that it is one of the two cases diagnosed antenatally and it is the first one diagnosed as early as 17 weeks of gestation. Early diagnosis allowed the patient to deliver in a more specialized clinic that was able to deal with immediate postnatal complications. The family was psychologically prepared for the delivery of a baby with a major anomaly.

Keywords: Antenatal diagnosis; facial cleft; Tessier no. 30.

Introduction

Facial clefts are among the most frequently occurring congenital malformations, with an incidence of about 1:1000 births [6]. Tessier [5] classified craniofacial clefts in 1976 and numbered the lower midline facial cleft as 30. Tessier no. 30 cleft, in contrast to cleft lip and cleft palate, is extremely rare. There is a broad variation in the severity of this deformity, ranging from a single notch in the mucosa to complete cleavage of the inferior part of the face and neck. No hereditary factor has been identified. No sex and racial predilection is observed.

We herein report a case with Tessier no. 30 defect diagnosed early in the second trimester and discuss its management. Timing of the surgery and staged repair of the cleft are the major keystones for optimal functional and esthetic results in the treatment of Tessier no. 30 cleft.

Presentation of the case

A 35-year-old gravida 6 para 5 female was referred to our hospital from a rural area of our region due to suspicion of facial anomaly at 17 weeks of pregnancy. She did not have any chronic disease, and her obstetric history was unremarkable. A kinship was present between the parents as her husband was her first cousin. There was no history of exposure to teratogenic substances or X-ray during pregnancy. There was Rh incompatibility, but the indirect Coombs' test result was negative. She did not have regular follow-up, and facial anomaly was suspected in her first prenatal visit at the 17th week of pregnancy.

An ultrasound examination was performed using a Logiq 200 PRO system (GE Medical Systems, Milwaukee, USA). This examination demonstrated a midline defect of the mandibular bone accompanied by soft tissue defect (Figure 1). Ultrasound examination revealed no associated abnormalities of the fetus. Amniofluid index was normal. The family was informed about the purpose of genetic amniocentesis. The family rejected amniocentesis and did not accept any other diagnostic method or follow-up either.

The patient delivered a 3100-g male infant vaginally at 39 weeks of pregnancy. The Apgar scores were 9 and 10 at 1 and 5 min, respectively. He was transferred to the pediatric intensive care unit of the same hospital for further evaluation and to recognize any early postnatal complications. Respiration of the baby was normal, and feeding was achieved by using a nasogastric tube (Figure 2).

He was referred to the Plastic and Reconstructive Surgery Clinic on the second day after delivery. On detailed examination, it was reported that the cleft extended from the lower lip to the sternal notch, but the larynx, trachea, thyroid gland and other structures of the neck were not affected. Examination of the oral cavity revealed that the tongue was bifid with ankyloglossia. Palatal structures and upper lip were intact. Examination of the neonate revealed no associated cardiac or gastrointestinal anomaly. “Tessier no. 30 defect” was the diagnosis.
Surgery was performed in a stepwise manner as described in the literature. Lip and soft tissue repair was performed with multiple Z-plasties at the second month. The second stage was delayed to minimize damage to the developing tooth buds.

**Discussion**

Median mandibular cleft was first described by Couranne [3] in 1819, and less than a hundred cases have been recorded worldwide until now. It varies greatly in presentation, ranging from minor clefting of the lower lip to complete cleavage of the mandible [2]. Median mandibular cleft is thought to occur due to the failure of the midline union of the first brachial arch.

It can extend to inferior neck structures, and separation, hypoplasia or absence of hyoid bone, thyroid cartilages or manubrium sterni may be present [1]. Tongue involvement is typical and is also variable in expression, ranging from a bifid anterior tip with ankyloglossia to marked lingual hypoplasia. Severe cardiac malformations such as single ventricle and transposition of great vessels and chromosomal anomalies can also accompany the condition [4].

Advances in prenatal diagnosis allowed obstetricians to diagnose facial clefts in utero. In the literature, there is only one case of labiomandibular cleft diagnosed prenatally at 32 weeks of pregnancy [8]. Our manuscript is also important in this respect as the case was diagnosed at 17 weeks of gestation.

The importance of antenatal diagnosis arises from its association with chromosomal abnormalities and associated anomalies, which can worsen the neonatal prognosis. Knowledge of the associated anomalies may provide the chance of delivery in a more equipped center with attendance of pediatric specialists. Also, identification of an anomaly before birth gives parents the chance to prepare psychologically.

The goal of treatment is to restore the tissues both functionally and esthetically. The psychological well-being of the child is also important. So, pediatric plastic surgeons, maxillofacial surgeons, orthodontists, speech therapists, physiotherapists and psychologists must work as a team in the management [7].

Surgery is usually performed in a stepwise manner. Closure of the lower lip and release of the tongue are mostly performed in the early stages of life to restore sucking, swallowing and phonation. The timing of the treatment of mandibular cleft remains controversial, but defect closure is mostly delayed until after 10 years of age in order to prevent damage to tooth buds during mandibular surgery. Z-plasty is often used if the cleft extends into the neck [1].

In conclusion, with the advances or advancement in prenatal diagnosis, malformations can be diagnosed at an earlier time. Examination of the face can give us the chance to diagnose many syndromes, chromosomal defects and isolated anomalies. In this way, early complications after delivery can be minimized and treatment can be planned even before delivery.

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References


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