Aplasia of the columella and cartilaginous nasal septum associated with choanal atresia

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ABSTRACT

An uncommon case of aplasia of the columella and cartilaginous nasal septum associated with choanal atresia is reported. Immediate simple management has been described. This appears to be the first report of such a congenital abnormality in the English literature.

Keywords: Aplasia, columella, nasal septum, choanal atresia.


Despite the complex embryological development of the nose and the surrounding structures, developmental nasal anomalies are rare, occurring once in every 20,000 - 40,000 births (Hughes et al 1980). Aplasia of the columella and cartilaginous nasal septum are extremely rare. Our search revealed that only one case has been previously reported in Literature by Jacobs, 1984. We, therefore, present this as the second case ever reported.

Case Report. G.B. was a full term baby girl delivered spontaneously and vaginally to a second degree cousins Pakistani couple on 20th December 1993. She had Apgar scores of 3 and 6 at one and five minutes respectively. The mother was twenty three years; Para 1, Gravida 2 and one abortion. She aborted the first pregnancy spontaneously at fourteen weeks of gestation in Pakistan. The cause was not known. Father was twenty six years old with no other wives. There was no relevant family antecedence nor maternal drug ingestion at any stage of the pregnancy. Apart from Rubella titre being positive at 1:32 in the mother, other TORCHES (toxoplasmosis, rubella, cytomegalovirus, herpes simplex and syphilis) screening results were negative.

At birth, G.B. was noticed to be symmetrically intra-uterine growth retarded: Birth weight: 1.89kgm., Occipito-frontal circumference: 28cms.; Chest circumference: 28cms.; Crown-hell length: 49cms. All data were below fiftieth centile for her gestation and sex.

There was absence of columella and nasal septum though the external nasal configuration was normal. The palate was marginally high arched; there was neither a cleft lip nor a cleft palate. There was no associated cardiac nor ocular defects. Attempts at passing a naso-gastric tube failed as it got arrested about 2 cm from the nasal outlet. Choanal atresia was suspected; this was later confirmed by the dye test Fig. 1.

G.B. did not require an emergency tracheostomy because immediately after the delivery she tolerated oro-tracheal intubation and respiratory support with a continuous positive airway pressure (CPAP) with positive end expiratory pressure (PEEP) of 3cms of

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water and fractional inspiratory oxygen FiO₂ of 40%. She was pink and maintained satisfactory arterial blood gases for five days during which period she was weaned off from the ventilator. Then after, she established respiration through an oro-pharyngeal tube. She remained pink and had satisfactory blood gases till she was fifty nine days old. She then established successful breathing through the mouth with a face mask oxygen of 0.5 litre per minute.

While in the incubator, GB from time to time experienced occasional distress at sucking, regurgitated milk quite often and failed to thrive. Therefore, GB’s immediate problems were: firstly, to establish independent breathing through the nasal airways; and secondly to coordinate the acts of sucking, swallowing and at the same time with that of breathing. At the age of two months, the first author carried out a transnasal approach to open up the choanal atresia. A size 4.0mm. soft portex tube stent was left in situ for eight weeks and the child was covered with broad spectrum antibiotics for entire stenting period (Singh 1990). She made an uneventful progress post-operatively.

Discussion. The facial structures - nose, mandible, maxilla and frontal prominence - developed around a shallow ectodermal depression known as stomodeum. By the time the embryo is four and a half weeks old, mandibular swellings can be distinguished caudally to the stomodeum, the maxillary swelling laterally and the frontal prominence, a slightly rounded elevation, cranially. During the fifth week, the lateral and the medial nasal swellings surround the nasal placode which then forms the nasal pits. The nasal pits then become closed inferiorly and form the nasal cavities. The lateral nasal swellings will form the alae of the nose and the medial swellings will give rise to the middle portion of the nose, the upper lip, and the maxilla as well as to the entire primary palate. The nasal septum develops from a midline ridge from the posterior edge of the frontonasal process and fuses with the palatine process thereby forming a partition between the primitive nasal cavities.

The majority of nasal anomalies present as a nasal mass with or without nasal obstruction (Hughes et al 1980). The most common nasal anomalies reported in literature are in form of dermoids, gliomata, fibroma and hairy polyps. Fifty patients with congenital nasal lesions (26 males and 24 females) were treated at the Hospital for sick Children in London, between 1978 and 1988; Morgan et al 1990. Their study included all developmental nasal anomalies over the period apart from choanal atresia. In these series covering a period of ten years, no single case of aplasia of the columella and cartilaginous nasal septum was found.

Genetic factor, maternal drug ingestion such as steroids, phenytoin, reserpine etc. (Richard L.J. Dyball et al 1992) and maternal infections especially in the first ten weeks of pregnancy (syphilis and measles) have been incriminated as possible causes of congenital facial anomalies (Mark G. Martins 1990). However, in our case all these factors were negative. In the screening for congenital viral infections, G.B. had negative IgM titre for Cytomegalo-virus and Herpes simples. Toxoplasmosis and V.D.R.L. screenings were also negative. Rubella specific IgM was positive at 1:32 in the mother. However, it has not been thought that rubella can lead to this degree of facial anomaly without any other added defects especially at such a low dilution. We, therefore, consider the etiology of these anomalies to be idiopathic.

A thorough search of the literature reveals that the first case of aplasia of the nasal columella and cartilaginous septum was reported in a German literature (Jacobs K.F., 1984). Unlike our own case, the case reported by Jacobs was not associated with choanal atresia so the patient was able to live till adolescent age before he was treated. Jacobs described the reconstruction of the nasal septum and further correction of the nose by means of a tube pedicle from the upper arm.

In this case reported, in order to ameliorate the problems of G.B. from getting distressed at sucking and choking on each attempt at feeding which is a contributing factor to her failure to thrive, we carried out a transnasal approach to create a nasopharyngeal airway in the first instance. Having established a good airway, definitive operation to construct the septum was planned for a much later date.

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References
