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BRIEF COMMUNICATION

Double Barrel Sign: A Pointer for Prenatal Detection of Bifid Nose

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Abstract Congenital anomalies of the nose can range from subtle forms to overt deformities. Bifid nose is a relatively rare anomaly. Here, the authors report a case of bifid nose detected in utero and suggest the double barrel sign as a pointer for the prenatal detection of bifid nose.

Keywords Double barrel sign · Bifid nose · Prenatal diagnosis

Introduction

Nose is a prominent feature of human face. Congenital nasal deformities can occur in a diverse spectrum of severity and can range from overt to subtle deformities. Bifid nose is a relatively rare anomaly and there are very few reports of bifid nose in the fetus [1, 2]. Here, the authors report a case of bifid nose detected in the prenatal period.

Report of Case

A 28-year-old $G_2P_1L_0$ came at 19 weeks of gestation for evaluation of fetal anomaly—ventriculomegaly with suspicion of cleft-lip palate. It was a spontaneous conception, and there was no history of consanguinity. The first baby was a male child and had multiple anomalies—unilateral cleft-lip palate, low-set ears, and bilateral talipes. The baby expired on the second neonatal day. No investigations were done.

Ultrasonography showed a fetus with parameters corresponding to 19 weeks of gestation. There was hypertelorism and ventriculomegaly. The lateral ventricle measured 12 mm. In the coronal view of the face, the nose was bifid—it appeared broad with a cleavage between the nostrils resembling the mouth of a double barrel gun (Fig. 1). The lips were intact. A diagnosis of frontonasal dysplasia was suspected in view of the bifid nose, hypertelorism, and cranial anomaly. Amniocentesis was done. The couple, when counseled regarding the anomalies, opted for termination of pregnancy.

Post-termination the antenatal findings were confirmed. The fetus was found to have bifid nose, broad nasal root, and hypertelorism (Fig. 2). The fetal karyotype was normal.

Discussion

Formation of human face involves multiple tissue swellings—the frontonasal, medial and lateral nasal, and maxillary and mandibular prominences derived from the neural crest. The nose is formed from the frontonasal, medial, and lateral nasal prominences. Bifid nose is a relatively rare entity. A bifid nose results when the medial nasal prominences do not merge completely [3].

Bifid nose can be seen in frontonasal malformation (FNM) which is also known as median facial cleft syndrome or frontonasal dysplasia. FNM is a developmental defect caused by an abnormal development of the

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frontonasal eminence. It consists of mid-facial anomalies that involve the eyes, nose, forehead, and occasionally the brain. The common features are orbital hypertelorism and abnormal nose (bifid or cleft). Other associated anomalies seen are widow's peak, microphthalmia, strabismus, accessory nasal tags, cleft-lip/-palate, bifid uvula, low-set ears, conductive deafness, intellectual disability, congenital heart disease (tetralogy of Fallot, atrial septal defects), abnormalities of the corpus callosum (lipoma, agenesis, dysgenesis), encephalocele, and limb anomalies [4–6]. The present case had bifid nose, broad nasal root, hypertelorism, and ventriculomegaly and clinically fits into the spectrum of frontonasal dysplasia.

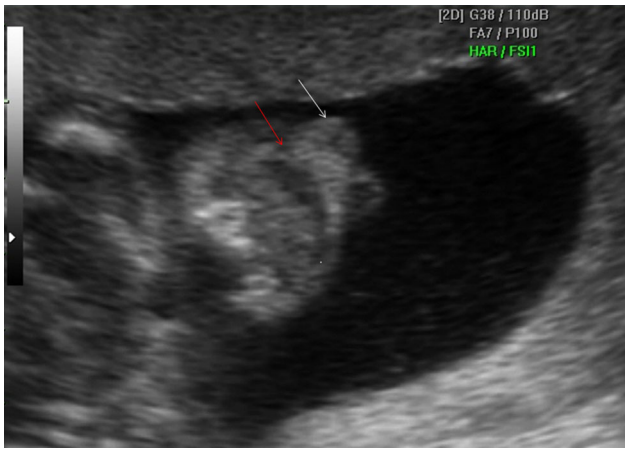

Fig. 1 Image showing bifid nose (*white arrow*) and upper lip (*red arrow*)

In this case, a 3D evaluation of the fetal face would have been very useful. In such cases, a 3D surface rendering will expose the full extent of the anomaly which will help the emotionally-disturbed parents take a decision regarding continuation/termination of pregnancy.

To conclude, bifid nose is a rare congenital anomaly which can be detected in the antenatal period and the authors propose the "double barrel sign" as a pointer for the prenatal diagnosis of bifid nose.

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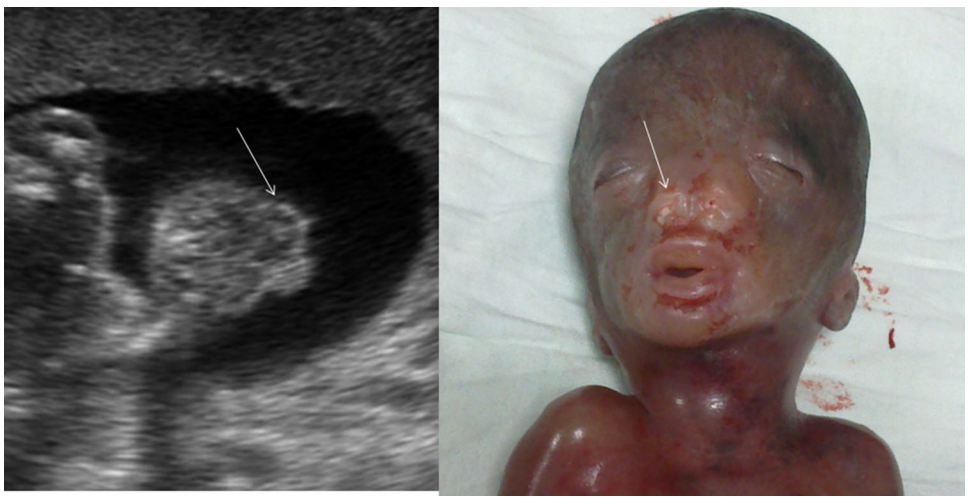

Fig. 2 Prenatal and postabortal image of bifid nose: Ultrasound image (*left*), clinical photo (*right*)