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"A CASE OF MICROGNATHIA, MICRODONTIA MICROSTOMIA AND CROWDING IN BILATERAL TEMPOROMANDIBULAR JOINT ANKYLOSIS"

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ABSTRACT

Aim: *The objective of present study is to report a case of micrognathia with microdontia and crowding of teeth in a 19 years old adolescent male.* ***Place of Study:*** *This case was studied in outpatient department of oral medicine, Dental College, Mahatma Gandhi Medical College Campus, Puducherry.* ***Period of Study:*** *This case was studied during May - June 2012.* ***Case Report :*** *A 19 years old adolescent male born to a non-consanguineous couple came with the history of difficulty in swallowing and unable to chew properly. He had difficulty in articulation of speech. On examination, he had microstomia, crowding of teeth in the lower jaw and microdontia associated with small mandible with receding chin. His movement of the mandible was very much restricted.* ***Discussion:*** *This case is well compared and correlated with available literature.* ***Conclusion :*** *The present study on microdontia and micrognathia has developmental and dental significance. Hence it was studied and reported.*

Keywords : *Hypoplasia of mandible, small mandible, microdontia, single gene disorders.*

INTRODUCTION

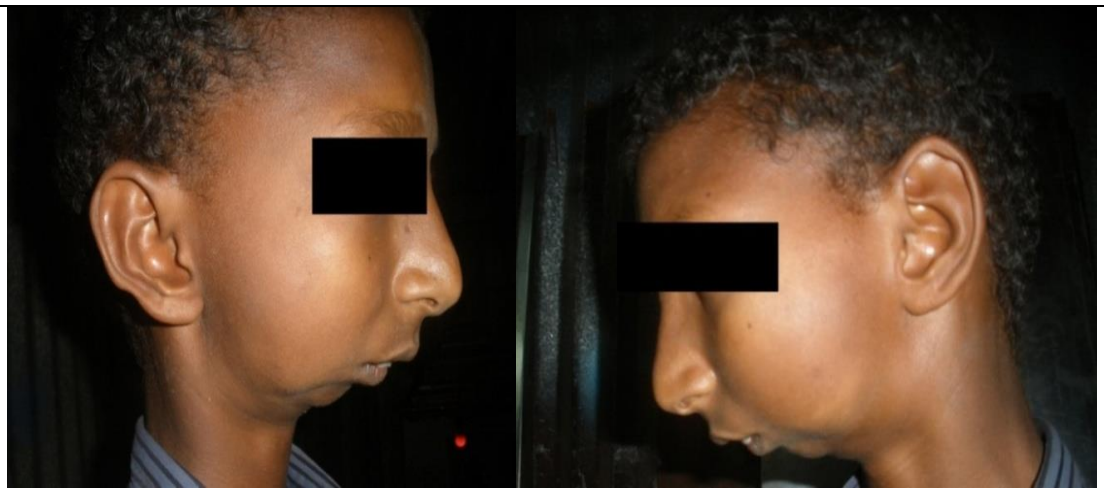
Micrognathia is malformation of face characterized by hypoplasia of mandible and a small, receding chin that fails to maintain the tongue in a forward position [1]. This malformation is

associated with single-gene disorders, chromosomal abnormalities, neuromuscular abnormalities, and other syndromes. The prognosis of fetal micrognathia is poor [2]. In isolated cases of micrognathia, it is component of Pierre-Robin syndrome (PRS) [3]. Growth of mandible is due to oral motility, which begins in early part of intrauterine life [4]. Lack of mandibular movements and respiratory movements (which are required for development of lung) results in development of micrognathia. It has been suggested that prenatal and neonatal brainstem dysfunction as a neuroembryological hypothesis for the explanation of onset of Pierre-Robin syndrome (PRS) by the same investigators [5,6].

Case Report

A nineteen years old adolescent male came with history of difficulty in talking, difficulty in swallowing, difficulty and painful chewing to outpatient department of oral medicine present since birth from Pilliyarkuppam, Puducherry. He also complained of small vestibule of mouth. He was examined thoroughly and investigated and found to have small mandible and receding chin, associated with small teeth overcrowded anteriorly in the mandible. He was under built and of short stature (145cm). He had bird like face, receding neck-chin angle, crowding of dentures with Andy gump appearance and bilateral temporomandibular joint ankylosis. There were no microcephaly, hydrocephalous, dysmorphic features, hand/foot anomalies, chromosomal abnormalities, pulmonary hypoplasia, neuromuscular abnormalities, single-gene disorders. All his systems, including central nervous systems were normal. He was of normal intelligence and working in a furniture factory. There were no localizing and lateralizing signs and symptoms but he had pain while chewing food. His bulk and nutrition of muscles were normal, reflexes were sluggish in the limbs. He had no dysphagia. He was the first child of his non consanguineous parents and there was no family history of any anomalies. There was no eruption of last molar tooth (wisdom tooth). No other anomalies were present.

Facial profile demonstrating rounded face, microstomia, crowding of dentition and restricted mouth opening



Lateral profile demonstrating bird like facies, retrognathic mandible, acute neck chin angle

DISCUSSION

The first case reported on micrognothia and cleft palate in medical literature was done by Fairbairn in 1846 [7]. The infants born with bird like face associated with hypoplasia of mandible and cleft palate is known to mankind and has already been recorded for thousands of years. The earliest recorded oro-facial deformities was found in the ancient Babylon dating back to 1700 BC. Many prophecies were linked with congenital anomalies. There were many references of congenital anomalies in Hippocratic writings [8]. In Splenogonadal fusion, there are two known types. They are continuous type and discontinuous types. In the continuous type, spleen is connected to gonads associated with defective limbs,micrognathia, cardiovascular anomalies like ventricular septal defects, skeletal anomalies, spina bifida,

gastrointestinal system anomalies like anal atresia, microgastria, spina bifida, craniosynostosis, deficient coccyx and bifid spine C6-T3, thoracopagus, diaphragmatic hernia and anomalies of lung like hypoplastic lung and abnormal lung fissures. The discontinuous type is usually not associated with congenital defects but there will be fusion of gonad with accessory spleen but not attached to native spleen. Etiology is unknown [9]. Singh et al [2012] has reported a rare case of giant encephalocele associated with micrognathia and microcephaly in a 5 months old, male second child from a non-consanguineous couple delivered by caesarean section. There was no other abnormality. The association of meningo-encepahlocoele with micrognathia and microcephaly has been reported by Hukum Singh et al [2012]. It is very rare and due to partial deletion of chromosome 13q. The presence of micrognathia faces the problem during intubation and surgical intervention. Giant meninigoencepahlocoele gives rise to cerebrospinal fluid loss causing hemodynamic alteration. The prior knowledge of presence of micrognathia helps to handle the case carefully to avoid further complications [10]. Morokuma S et al reported a case of micrognathia in 27 years old Japanese primigravida of 33 weeks of intrauterine life who had polyhydramnios and threatened preterm labour. Ultrasound studies showed clinical polyhydramnios (amniotic fluid index: 28 cm) and micrognathia was on midsagittal and 3D scan. The child had hypoplasia of manbdible and glossoptosis. These findings in ultrasound is suggestive of brain dysfunction [1]. Lannelongue and Menard [11] and Shukowsky (1911) [12] reported cleft palate as associated anomaly. Pierre Robin in 1923 also described a syndrome characterized by cleft palate, micrognathia, and glossoptosis [13].

Present Study

Ninteen years old adolescent male from Puducherry had micrognathia microstomia microdontia and crowding of teeth from birth. He had bird like face, receding neck-chin angle, crowding of dentures, andy gump appearance and bilateral temporomandibuar joint ankylosis due to his small mandible and receding chin with small vestibule of the mouth. He had difficulty in articulation of speech and difficulty in chewing. Due to fusion of condyles to mandibular fossae multiplanar growth of mandible is affected. There may be high frenulum lingular attachment and as a result there may be restriction of movements of mandible. There were no microcephaly, hydrocephalous, or hand/foot anomalies, chromosomal abnormalities, pulmonary hypoplasia, neuromuscular abnormalities.

Take Home Message

This combination of micrognathia, microstomia and microdontia seen in 19 years old male is unusual. So any patient with such anomalies has to be evaluated for any associated systemic anomalies.

CONCLUSION

This is a rare case of micrognathia with microdontia in a 19 years old male who had difficulty in chewing and speech. Micrognathia may be congenital or acquired. Congenital micrognathia may be due to single gene disorder. Acquired micrognathia may be due to bilateral temporomandibular joint ankylosis

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