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Case Report

Syngnathia: a jaw dropping case of fused jaws in a neonate

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ABSTRACT

Syngnathia is an extremely rare condition with a congenital fusion of the maxilla and mandible described as an occlusion of the oral cavity opening. No case of complete bony synostosis, identified in the new-born period have survived beyond few months of age, hence making this case a novel addition to literature. Here we present a case of a term neonate with fused maxillo-mandibular jaws and synechial closure of the mouth with subtle dysmorphic features. Respiratory distress and feeding issues remained the most important challenges. Investigations with baseline radiography, a 3D CT reconstruction of the head, face and neck anatomy elucidated bilateral bony fusion of maxillomandibular jaws, a midline cleft in the soft palate and micro-glossia. After initial stabilisation, surgical intervention was done in two graded phases. First, an orifice was established to enable an airway route, in an anticipation of difficult airway in an event of distress followed by major synechiae and bony excision with placement of an oral distractor and exteriorisation of the small tongue. With multi-disciplinary team effort and physio-motor stimulation, she was established on direct breast feeding by 37 days of life. She was developmentally normal at 7 months of age with decent growth.

Keywords: Syngnathia, Maxillo-mandibular fusion, Synechiae, Synostosis, Neonate, NICU

INTRODUCTION

Congenital craniofacial disorders represent approximately 20% of all birth defects.[1] Syngnathia, a congenital fusion of the jaw, is an extremely rare condition described as an occlusion of the oral cavity opening. An extensive search of literature done from 1936 to 2018 showed 118 cases with 62 cases of bony fusion (syngnathia), 48 cases of fibrous fusion and 8 cases of combined fusion.[2]

To emphasize, no case of complete bony synostosis identified in the newborn period have survived beyond few months of age, hence making this multidisciplinary case management a novel addition to literature.

CASE REPORT

A term female neonate of birthweight 2.7 kg was born at a referring hospital to a gravida 2 woman who had a 2nd degree consanguineous marriage, via elective caesarean section in view of gestational diabetes mellitus. There was no history of any medication during pregnancy other than iron and folic acid. She had a normal transition after birth and was noted to have no oral cavity opening and hence referred to our hospital at 2 hours of life for further management. At admission, she had fused upper and lower gums along with absence of oral aperture and other minor dysmorphic features like broad forehead with mid-facial hypoplasia, protruding lips, small nasal columella, low-set ears, retrognathia and dynamic bilateral varus

deformity of lower limbs. She was haemodynamically stable, however feeding and probable difficult airway remained the major issues. Initially Intravenous fluids were given which was later transitioned to nasogastric feeds. Her baseline biochemical, haematological, cardiac (echocardiogram) and neurologic (neurosonogram) were unremarkable. Her clinical features did not fit into any major known chromosomal disorder/sequence.

Our multidisciplinary team of paediatric surgeons, plastic surgeons, oral & maxillofacial surgeons and paediatric anaesthesiologists were consulted for a better understanding of the anatomy and feasible surgical corrective options were discussed in a multidisciplinary meeting. CT face and neck showed bony fusion of maxillomandibular complex with hypoplastic mandible and normal temporomandibular joint, suggestive of syngnathia with synostosis and cleft palate (Figure 1).

Based on CT scan, a 3D acrylic bone model was reconstructed using which the defects were precisely located for ease of access during surgery (Figure 2). On day 3 of life, in order to establish per-oral access, synechiae release was done under local anaesthesia with bilateral infra-orbital and mental nerve block by the oro-maxillofacial surgery and paediatric surgery team. Using unipolar cautery, mucosal synechiae release was done and using Fergusson's mouth gag, an opening was created, and the 3 mm ET tube was kept in situ to act as an oro-pharyngeal airway (Figures 3-5). The infant was continued on nasogastric feeding with close monitoring of nutrition and respiratory status. Physiotherapy and occupational therapy were involved for early initiation of physio-motor stimulation in anticipation of disuse atrophy of masticatory muscles.

On day 16 of life, she was taken for definitive surgery under general anaesthesia. Naso-tracheal intubation using fibreoptic scope was done. Bilateral bony synostosis was excised using a piezo crystal cutter and cautery. On visualisation of oral cavity, an extremely small tongue with posterior cleft palate was noticed. In view of anticipated difficult airway, stay sutures were inserted into the tongue musculature and were left outside to act as an anchor and prevent tongue fall-back (Figure 1).

Vaseline-soaked gauze were placed bilaterally between the jaws to prevent granulation tissue and re-synechiae formation. Post-operative inspection showed minimal bleed after which the gauze packs were removed and a silicone oral mould of the baby's jaw, prepared with the help of 3D reconstruction was placed in the oral cavity to prevent re- synechial fusion. After continuing nil by mouth for 48 hrs, tube feeds were re-started. The infant was extubated onto non-invasive respiratory support after 5 days of ventilation assistance.

Constant pooling of secretions due to primitive swallowing reflex and small tongue posed major concerns of aspiration. Close assessment of nutrition and growth were done along with optimal fortification of feeds. Gradual suck and swallow coordination were ensured with the help of oral pacifiers and continued stimulatory exercises. Surgical review ruled out refusion of the mandible and maxillary joints and she had satisfactory oral opening. She was transitioned onto room air by day 35 and was able to take paladai and direct breast feeds by day 37 of life and discharged home.

Birth dose immunisations were given, screening for inborn errors of metabolism report was normal and hearing screening was satisfactory. Parents were counselled by genetics team regarding whole exome sequencing which couldn't be done due to the financial reasons.

Figure 1: CT head and neck showed bony fusion of maxillomandibular complex with hypoplastic mandible suggestive of syngnathia.

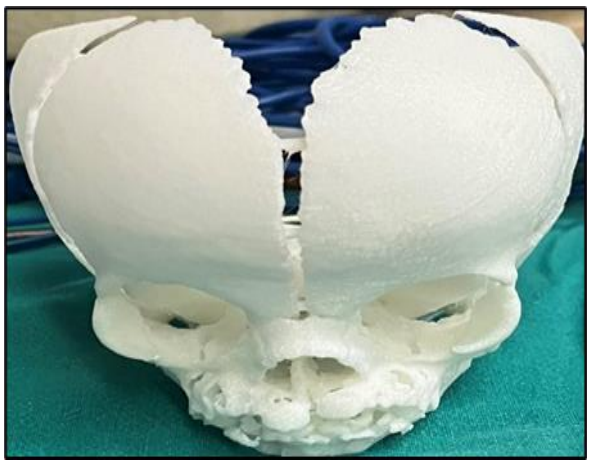

Figure 2: Acrylic 3D reconstruction of the neonatal head to visually identify the defect better, prior to surgery.

Figure 3: Intra-op picture of the neonate showing absent oral aperture.

Figure 4: Post synechiae release (first surgery) on day 3 of life.

Figure 5: Post synechiae release a 3 mm endotracheal tube kept *in situ* to act as an oropharyngeal airway.

Figure 6: Post-operative picture after the 2nd surgery with excision of the bilateral bony synostosis with stay sutures inserted into the tongue musculature and left outside to act as anchor and prevent tongue fall-back.

Follow up

Parents were taught basic life support skills, correct feeding techniques to prevent aspiration, physiotherapy and oro-motor exercises to be continued at home. She was followed up regularly along with oro-motor rehabilitation and occupational therapy assistance. At 7 months of age, she has achieved a weight of 6 kgs, head circumference of 48 cm. Her development is normal in major domains, her oro-motor movements remain to be vigorous and coordinated. She is on breastfeeding and complementary feeds. Palatal defect closure and reconstruction has been planned by plastic surgery team.

DISCUSSION

Syngnathia, being an extremely rare condition with fused jaws results from fusion between the maxilla or zygoma and the mandible.[3] This may occur as fusion of simple mucosal bands (synechiae) to complete bony fusion (synostosis) or combined fusion i.e., fibrous on one site and bony on another side. Though the exact pathogenesis is unclear, defective stapedial artery or loss of neural crest cells has been suggested as one of the reasons.[4] It can occur in the midline, laterally, or can be uni/bilateral with unilateral fibrous fusion being commonest variety.

Till date, 60 cases with the same degree of severity as ours, have been documented in the literature.[5] Co-existing head and neck abnormalities include: cleft lip, cleft palate, clefts of mandible, oblique facial clefts, tongue anomalies (bifid, small or absent), glosso-palatine ankylosis, mandibular hypoplasia, coloboma, hypophyseal duplication and frontonasal malformations. Commonly associated syndromic conditions include Van der Woude, popliteal pterygium, Dobrow syndrome and craniofacial microsomia. In our case, the infant had a

complete bilateral bony fusion of the mandible to the zygomatic complex, a hypoplastic temporomandibular joint and a posterior cleft palate.

The primary management of syngnathia warrants a specific multidisciplinary approach with surgery being the mainstay to decrease mortality, disability, and to improve the functional and aesthetic outcomes. As the condition is extremely rare, there are no standardized management protocols.[4] The major concern is securing the airway and early tracheostomy or intubation using fibreoptic technique have been recommended by different groups. Another challenging issue is the high recurrence rate of ankylosis and refusion. The engagement of an appropriate maxillary-mandibular distractor and active physiotherapy for continual mouth opening is key to prevent recurrence. In our case, airway was relatively well maintained and needed fibreoptic nasotracheal intubation just prior to surgery.

If intervention is not timely administered, it could lead to several maladaptive consequences like hypoplastic teeth, lower face growth restriction, 'bird face' appearance due to restricted mandibular growth. abnormal teeth eruption, caries and subsequent retrognathia. In a similar case reported by Daniels et al the bony fusion affected the growth of the infant's lower face resulting in hypoplastic maxilla. The child did not survive beyond 2 months from the surgery due recurrent infections secondary to chronic malnutrition.[6] In another case of congenital bilateral zygomatico-mandibular fusion with mandibular hypoplasia, a male neonate underwent tracheotomy for severe upper airway obstruction and limitation of mouth opening.[7] He had difficulty in opening his mouth and masticate. He underwent several surgical procedures including tracheostomy, osteotomy to release maxilla-mandibular bony fusion, skin grafting to the floor of the mouth to release and reposition the tongue and finally reconstruction of temporomandibular joint and lengthening of the ramus with a costochondral graft at 1.5 years of age. Despite intensive care, he died about 24 hours after the operation. Synostoses hence remain a challenging area in neonatal critical care with reports in the literature highlighting the initial challenge with treatment, survival and long-term follow up.[8,9] However, in our case, the infant has been thriving reasonably until 7 months of age and is being managed by multiple teams involved in her care which make this case unique as getting multidisciplinary care is often a hurdle in settings like ours. Long term neurodevelopmental follow up is planned under high risk follow up clinic as per protocol.

Care of such complicated neonates involves multidisciplinary team planning from the start and targeting short- and long-term goals in the best interest of the neonate. Having repeated family meetings with parents and concerned team members provides the parents a realistic approach, detailed understanding of the goals of management and strengthens their confidence during these difficult times. The role of a neonatologist in co-ordinating these multiple teams and holistic management of the neonate is highly pertinent.

Lessons for the clinician

The key issue in syngnathia is difficult airway, and hence in anticipation of respiratory distress, emergency surgery for synechiae release may be needed to secure an access for an emergency oropharyngeal conduit. Maintaining airway is first priority in all these cases due to the absent oral aperture. In such cases, nasopharyngeal intubation can be considered as it is less invasive and technically easier. A complex, rare congenital structural anomaly such as this case requires a high degree of co- ordination amongst multiple specialist departments to ensure both surgical, medical and rehabilitative success in terms of intervention and prevention of complications/ relapses. Due to varied severity of the condition, timing and planning of the surgery must be modified to situation and overall state of patient. At present, there is not enough long-term outcome data available of these patients.

CONCLUSION

Syngnathia presents a significant challenge primarily due to the difficulty in managing the airway, making it crucial to be prepared for potential respiratory distress and emergency surgery to release the synechiae and establish access for an emergency oropharyngeal conduit. Given the absence of an oral aperture, maintaining the airway becomes the top priority, with nasopharyngeal intubation being a viable option due to its less invasive nature and technical simplicity. This rare and complex congenital anomaly necessitates extensive coordination among various specialist departments to achieve success in surgical, medical, and rehabilitative interventions, as well as to prevent complications and relapses. The timing and planning of surgery must be adapted to the severity of the condition and the patient's overall health, but currently, there is lack of long-term outcome data for these patients.

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