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A rare case report of congenital maxillomandibular complex fusion: A Review of Literature on Congenital Syngnathia

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ABSTRACT: congenital maxilla mandibular fusion is a rare deformities seen in very few cases in literature. More often these cases appear to be syndromic and often involve maxilla and mandibular deformities along with musculoskeletal growth disturbance. The aim of this paper is to report a case of bilateral maxilla mandibular complex fusion of jaws, with altered growth retardation and functional disability. pre anaesthetic evaluation and physical fitness prior to surgery was a great challenge.Considering the limited mouth opening and poor body mass index awake fiberoptic intubation was done. The surgical approach was mainly to releasing of maxilla mandiblular bony ankyosis along with bucco gingival soft tissue release. Temporal muscular flap was rotated to recreate TMJ. Mouth opening was achieved from 5mm to 32 mm postoperatively where she was able to feed herself. The aim of this paper is to review on syngnatia case literature and give an idea for early identification of sure deformitesand multi stage approch both surgical and post operative challenges to over come.

Key words; syngnathia ,maxillomandubular complex fusion, blind fiberoptic intubation ,Laster classfication.

I. INTRODUCTION:

Congenital bony fusion of the maxilla and mandiblar component is a complex and rare isolated condition. Only 7 cases of Fusion involving both maxilla and mandibule with zygomatic complex along with alveolar ridges(daniels, 2004)[1]are accounted. Itoften appears to be syndromic along with accompanied additional congenital defects, such as microglossia, micrognathia, temporomandibular joint anomalies, cleft lip, and frequently cleft palate. [2] Very few cases of isolated bilateral syngnathia were reported in the past literature.

Soft tissue defects or synechiae have been reported and reviewed by Gartlan et al 1993. [3]These authors proposed a classification of soft- tissue fusions based on their presumed origins, separating them either as buccopharyngeal-membrane remnants or as ectopic membranes. It was Burket who first reported a case report on congenital temporomandibular joint(TMJ) ankylosis and facial hemiatropy(1936) . [4]Petterson reported a case of aglossia with bony fusion of the mandible of the jaws(1961,Strernberg N et al reported congenital fusion of the gums with bilateral fusion of temporomandibular joint(TMJ) 1983. True bony fusion is a very rare anomaly, and only a handful of case reports exist in the literature with bilateral maxillomandibular fusion. Syngnathia with any associated systemic or intraoral anomaly, as in our case report, is extremely rare entity, and only very few cases have been previously reported.

The purpose of this paper is mainly to put forward the rare condition of syngnathia which affects the patients over all physical development and systemic nutritional deficiency on and growth and balance. The idea behind the surgery was to release the ankysosis and mobilize the jaws so that patient was able to feed herself. We have also reviewed the literature on earlier surgically operated cases of syngnathia with various other

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treatment modalities and post-surgical complication.

A Case report

A female child patient with age of 12 years reported to our oral and maxillofacial surgery unit with a chief complaint of inability to open mouth from childhood. On examination extra orally feature showed bird face deformity, she had inability to open mouth, reduced vertical facial height. Intra oral examination was very difficult since patient mouth opening was nil. She had severe trismus and most the facial musculature was poorly developed which was suggestive of defect existing from a long span. (figure 1 & 2)

A detailed history revealed that she had the condition from birth and she had no family history or any other relevant history parents reported mother had normal delivery with no complication at the time of birth. She was known to have nutritional deficient and her blood picture with hemoglobin showed a baseline valve of 9 gm% just below the normal hemoglobin levels. Her guardians main concern was about her mouth opening&mobilize the jaws as early as possible.

The main concern for us was her weight when she first reported to our outpatient department. She was 19 kilos with height of 125 cms where her body mass index showed disproportion and below the normal levels. Her general physical examination showed malnourished body muscular tone and facial Skelton. The patient was referred to both dietitian and pediatric surgeon for further review. The aim was to bring her hemoglobin level to above base line and body mass index to standard levels. A diet chart was maintained pre-surgically and hordes of balance protein diet was given to her.

Examination reveals underdeveloped musculoskeletal structure. Visual acuity was 6/6 vision on both left and right eye. All her cranial nerves were examined and were within normal parameters. Speech was normal and patient was able to respond to all activities. Intra oral examination revels unerupted right anterior, her mouth opening was completely zero, where she was able to take only liquid diet from anterior space (fig 1).

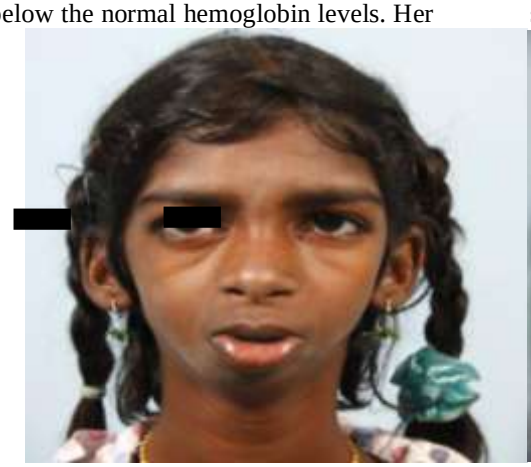

Figure 1 extra oral facial picture

figure 2- restricted mouth opening

Radiographic findings:

A set of conventional radiographs were made, which included an orthopantomogram (OPG) and lateral oblique skull views. And 3D CT scan showed complete bony fusion of maxillomandibularcomonnent with zygomatic complex along alveolus. Condyles on both sides were normal and palpable but fused to the glenod fossa. The coronoids were fused to maxillary and zygomatic complex anteriorly. There was no defined anatomical bony structure particularly in zygomatic area (Figure 3). Superior cranial base separation was ill-defined. There was no abnormality detected in cranium. Brain appears normal and with proper developed Dural space and ventricle spaces were within normal limits.

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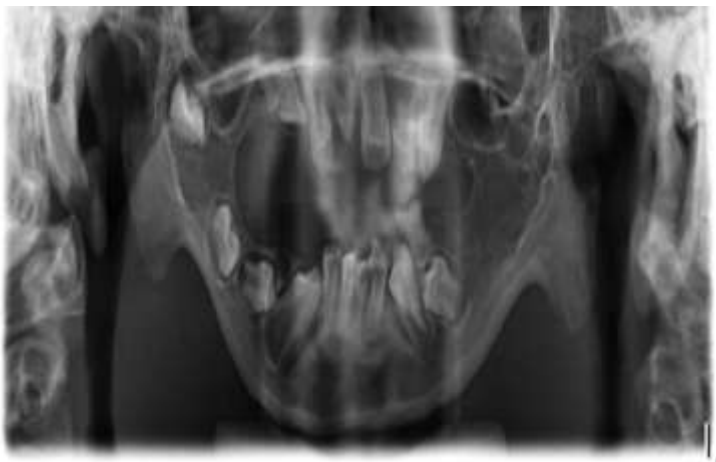

Figure 3 opg

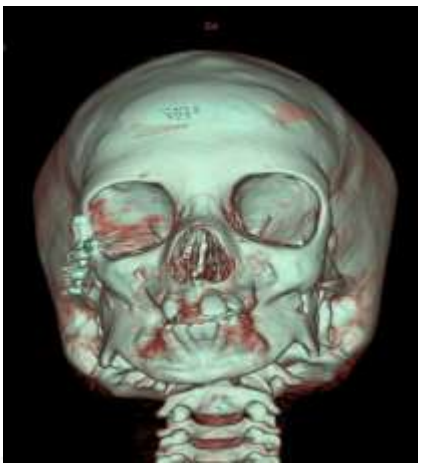

figure 4 frontal 3D CT

Figure 5 -right lateral 3D CT

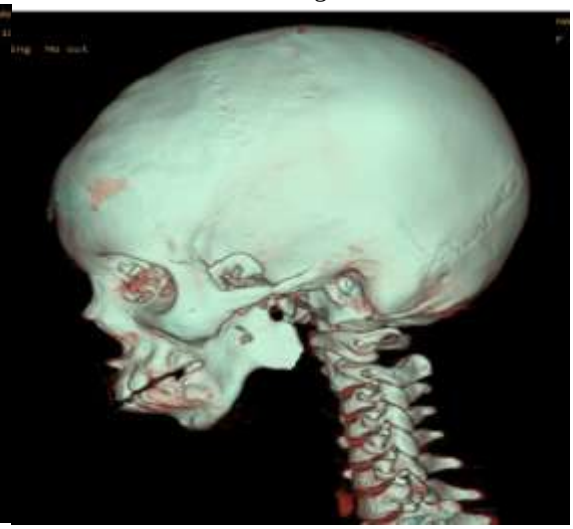

figure 6- left lateral 3DCT

Opg and 3D CT reconstruction revealed an extensive fusion of the mandible with themaxillazygoma.Maxillofacial CT with revealed the mandibular ramus to becompletely fused to the maxillary tuberosities and zygoma, withankylosis of the temporomandibular joint in figure(3, 4,5 &6) We classifed the deformity as a Type 2b according to the Laster classifcation.in Table 1.

II. MATERIALS AND METHOD:

Considering the limited mouth opening and poor BMI body mass indexan awakefiberoptic intubation was planned. Induction was done with glycopyrolate, midazolam, Fentanyl and dexmeditomidine. Lignocaine 4% spray to nose and oral cavity, 6.0 flex metal tube was inserted nasally and airway maintained by controlled ventilation. PropofolAtracurium and sevoflurane was used for maintainence of anaesthesia. Intraoperatively vitals were monitored and IV fluids were given.

Since she was a female patient Intra oral approach was planned. First leftside mucoperiosteal flap was reflected to identify bony ankylosis and with the use of pezo surgical saw the ankylotic bone was cut and excised. The cut extended from premaxilla towards posterior alveolar region extending posterosuperior region until the ramus and subsigmoid level sparing the inferior alveolar nerve pedicle. A clear safe margin of 5 mm bone was left intact at skull base area. The similar cuts were planned on right side after reflection of mucoperioteal flap. Once anterior ankylotic bone was removed by saw posterior piezo saw and osteotomies were used for posterior zygomatic region and coronoid separation. Once the separation was done a wedge shape ankylotic bone was removed both side using bone nibbler and rounger. Muscular fibers were free by using digital pressure around the mandible and to free the tensed musclofibre adhesion a favorable amount of 35 mm mouth opening was achieved intra

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operatively. (Figure 7 & 8) Interposition of muscular fibers in between the newly created joint and mucoperiosteal flap was closed. Once posterior region was closed anteriorly ribbon gauze packing was done to allow secondary healing. Post-operative recalls were planned with regular dressing with use of Bismuth Sub nitrate, Iodoform and Paraffin gel was used for dressing. Early mobilization of joint was done with mouth opening exercise and series of follow up radiographs were taken to assess the amount of space maintained.

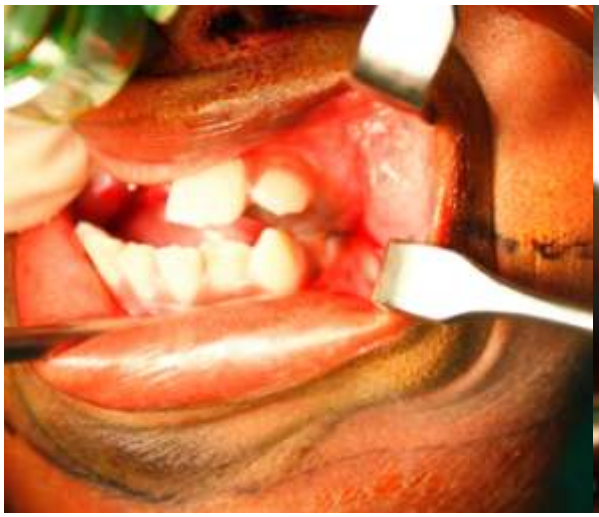

Figure7

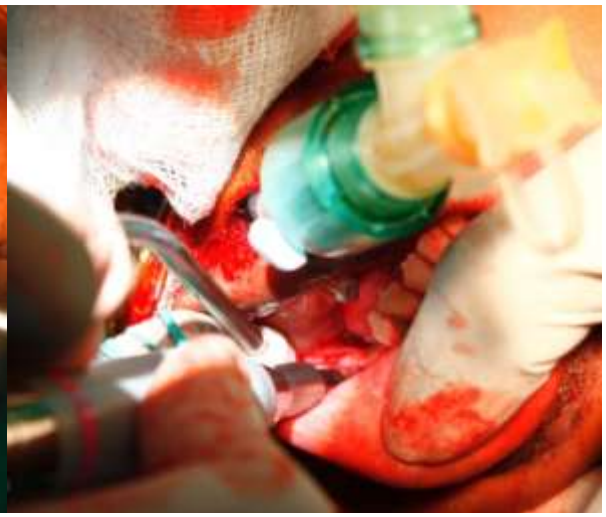

figure 8

Intra operative picture of surgeryfigure 7 & figure 8

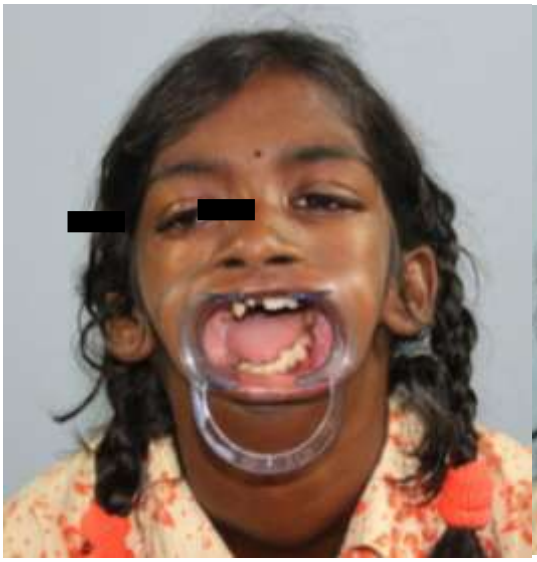

Figure 9

figure 10

Post-operative picture of 32 mm mouth opening and post 6-month physiotherapy

III. DISCUSSION:

Maxillomandiblar bilateral fusion defects of the jaws make up a rare group of anomalies. Congenital fusion of the jaw especially gums and entire maxillo mandibular complex, or isolated intraoral bands fusion, intra-alveolar synechiae are various entities. The fusion of the upper and lower jaws syngnathia can be a bony fusion of the maxilla and mandible which is called as synostosis. Soft tissue fusion synechiae are characterized by mucosal bands of tissue connecting various parts of the lower and upper jaws. Synechiae has been reported extensively in literatures, and its treatment is simple rather than complicated.Synechiae is most common reported in new born.

The first case was reported by Burket(1936) which had bony ankylosis of both maxillomandibular complex fusion along with Temporomandibular joint involvement.In our case both bilateral involvement of temporo mandibular joint (TMJ) was involved along with complex fusion.The etiopathology of syngnathia remains unknown, and various hypothesis have been proposed such as depressed fetal swallowing reflexes by Humprey(6), failure of tongue protrusion, especially in a small jaw by Walker and Fraser (7) in our patient they had history since birth where the patient had severe psychological trauma till her childhood, both the side TMJ was involved along with maxilla mandibular complex fusion.Uncontrolled proliferation of ectoderm theory was proposed bySnijmann(8). Other theories have highlighted the role of an abnormality of the stapedius artery by Poswillo(9), new born when they have bony fusion we usually classified it as acomplex type which is been classified by Laster. (10) (table 1) in our patient the history has been given as fusion of upper and lower jaw with bony fusion which categorised as complex type 2 b form according to lasterclassfication.

Till date a total out of 62 bony cases of congenital syngnathia, 24 were unilateral, 21 bilateral and 6 were midline syngnathia have been reported. Nine cases showed complete fusion, while 3 cases had bony fusion to hard palate. Temporal skull bone syngnathia and zygomaticomaxillary fusion were observed in two cases. Fifteen cases revealed TMJ involvement, while 45 cases reported normal TMJ. In our patient there was total fusion of both maxillo mandibular complex with complete involvement of TMJ.

In our patient bilateral bonyankylosis was removed with temporalis muscle repositioning was done to recreateTMJ and patient was kept on early and aggressive physiotherapy figure, follow-up period was done for 6months, post-operative adequate mouth opening was achieved. patient was able to take orally. figure (9&10)

Management protocol available for congenital syngnathia mainly includes early recognition of the deformity and surgical correction and release of the ankylosis. The relative overall presentation of this condition makes its difficult in itsmanagement& ismainly dependent on its type and severity of the condition. Assessment of thesepatients is usually done with the consideration of the current systemic state and is always multi-disciplinary approach. If there is any good credibility of survival without emergency surgical procedures, surgery should be delayed to prevent undue associated risks. Anesthetic complications like cardiac arrest secondary to perioperative laryngospasm, difficult intubation, pulmonary aspiration are few risk factors should be considered.

IV. CONCLUSION:

Congenital syngnathia is a disorder present at birth which requires early management and surgical correction of the deformities. When you see the Literature these maxillomandibular defect affects overall musculature both the functional and aesthetic stigma of the patients. Malnourishment and poor built is one of the common factors seen in these kinds of defects. Adequate nutrition has to be provided and an intervention of both dietitian and paediatrician to be followed for feeding and growth hindrance. Difficulties in providing an adequate feeding are the main cause of malnutrition in newborn infants with syngnathia.

We recommend that early management of this condition is necessary to prevent feeding and breathing difficulties. Regular jaw-opening and aggressive physiotherapy has to be done along with long term and close follow-up toavoid recurrence. Any delay in the surgical intervention will lead to impeded and distorted growth of facial structures, malnutrition and disorderederuption of teeth causing malocclusion. More cases of congenital syngnathia need to be reported to further augment the relevant clinical base so that a more definitive management protocol may be formulated.

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Laster classfication of syngnatia

Type	Category	Clinical features
Type 1a	Simple anterior syngnathia	Bony fusion of alveolar ridge without other congenital deformity in the head and neck
Type 1b	Complex anterior	Bony fusion of alveolar ridge with other congenital deformity in the head and neck
Type 2a	Simple zygomatic mandibular syngnathia	Bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia
Type2b	Complex zygomatic mandibular syngnathia	Bony fusion of the mandible to the zygomatic complex and associated with cleft or temporomandibular joint ankylosis