Congenital midline palatomandibular bony fusion with a mandibular cleft and a bifid tongue

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SUMMARY. Congenital fusion of the maxillary and mandibular alveolar arches and a cleft of the mandible are each quite rare. A patient who had both a midline palatomandibular bony fusion as well as a mandibular cleft with a bifid tongue is presented.

Congenital fusions of the maxilla and mandible are extremely unusual. Very few such cases have been reported to date.1-13 Most of these reported cases had either a lateral interalveolar fusion or fibrous synaechieae between the alveolar margins of the jaws. Further bony fusion, especially in the midline, has been reported in few cases. One reported case had an associated mandibular cleft with a bifid tongue.13 We report a newborn with a congenital bony midline palatomandibular fusion with a cleft of the mandible and a bifid tongue.

Case report

A 9-hours-old female newborn, weighing 3 kg, was referred to our centre with a diagnosis of a congenital anomaly of the oral cavity. She was the third born child of normal parents and was a full-term normal delivery. The older siblings were normal. The mother had not taken any medications during her pregnancy. Antenatal ultrasoundography had not been performed.

The general condition of the infant was very poor. She had aspiration pneumonitis and was in severe respiratory distress. A soft, fleshy, hairy, apparently pedunculated mass was seen protruding from the mouth (Fig. 1). She had severe trismus with mouth opening of only a few millimetres. The oral cavity therefore could not be seen but, on palpation, there appeared to be a midline cleft of the mandible which had a W-configuration. The lower lip also appeared to be divided into two halves turning inwards near the midline with a central skin bridge attached to the cleft mandible. A midline bony hard vertical bar fusing the maxilla and mandible seemed to be the cause of trismus. The anterior part of the tongue appeared to be bifid on either side of this bony bar. The maxillary alveolar arch appeared intact and the bony fusion seemed to be between the hard palate and the cleft mandible. X-rays of the skull and the mandible only showed that both the temporomandibular joints were normal. The poor general condition of the baby did not allow a CT scan or MRI. Thus, the exact anatomical nature and the extent of the malformation could not be assessed preoperatively.

Since intubation was not feasible, an emergency tracheostomy was performed. After detailed discussion, it was decided to operate immediately. The baby was subsequently anaesthetised and maintained via the tracheostomy airway. The fleshy mass protruding out was first separated from the lower jaw and excised. This exposed a 14-16 mm wide vertical pillar of bony palatomandibular fusion (Fig. 2). Under anaesthesia, it was possible to palpate the posterior extent of this pillar. The posterior one-third of the tongue and the pharynx were found to be free. A Gigli saw was manoeuvred around the pillar and the bony pillar was divided. Haemostasis was achieved by packing and pressure. Division of the bony fusion allowed the mouth to be opened 1.5 cm and a chance to visualise the abnormal anatomy better. The anterior two-thirds of the tongue were bifid and situated on either side of the bony bar. A silastic block was kept in the oral cavity to maintain the mouth opening. Ventilatory support was continued following surgery but the child succumbed to severe aspiration pneumonitis on the second postoperative day. Histopathological examination of the excised fleshy mass showed hair-bearing...
Figure 2—Diagrammatic representation of a sagittal section of the mouth in a normal infant and in our patient showing the location and extent of the bony fusion.

Skin encasing a core of adipose tissue. Autopsy demonstrated an associated ventricular septal defect.

Discussion

Millard\textsuperscript{14} noted that 46 patients with varying degrees of mandibular clefts were reported between 1819 and 1971. However, none of them had an associated maxillomandibular fusion.

Congenital trismus due to maxillomandibular alveolar fusion is a very rare entity. An extensive literature review has revealed only a handful of documented cases of maxillomandibular fusion, although a comprehensive compilation is admittedly difficult in this type of condition.\textsuperscript{1-13}

Burket\textsuperscript{1} in 1936 reported the first case of a congenital bony temporomandibular ankylosis. His case also had an associated facial hemiatrophy. However, most of the documented cases of maxillomandibular fusion were of alveolar fusion, situated on the lateral side. Some cases involved fibrous interalveolar synaechiae which could be easily divided.\textsuperscript{5,6} Others involved partial bony alveolar fusion.\textsuperscript{4,5} Some of these cases were associated with other facial anomalies such as cleft palate,\textsuperscript{2,3} mandibular hypoplasia,\textsuperscript{5,6,12} small or absent tongue,\textsuperscript{7,8} and, in one case, a persistent buccopharyngeal membrane.\textsuperscript{6} Sternberg et al.,\textsuperscript{9} Simpson et al.,\textsuperscript{10} Agarwal et al.\textsuperscript{11} and, most recently, Kamata et al.\textsuperscript{12} have also reported cases of syngnathia with temporomandibular or zygomaticomandibular fusion.

We know of only one reported case of midline maxillomandibular fusion with a mandibular cleft and a bifid tongue. This was reported by Calaycay et al.\textsuperscript{13} in 1976. In this case, the most striking feature was a bulky protruding mass composed of skin and subcutaneous fat. The soft tissue mass extended from the hard and soft palates to the mandible. Bilateral bony struts traversed the oral soft tissue mass and held the jaws apart. The duplicated mandible had a W-shaped configuration, as did the maxillary arch. The eyes were hypertelorlic and the nose flat and broad with a 3 cm wide columella. The anterior half of the tongue was bifid. Cases of partial facial duplication or of duplication of the mouth and the mandible have also been reported.\textsuperscript{12-20} We have not found any other case report of palatomandibular fusion with a normal maxillary arch and a cleft of the mandible with a bifid tongue. Hoggin\textsuperscript{7} and Pettersson\textsuperscript{8} come closest to our case, as far as the location of the bony fusion is concerned. In their cases too, the solid bony masses connected the hard palate with the mandible. However, the tongue was absent and the palate cleft in both their cases.

Various experimental studies have been performed to study the embryological basis for fusion. They were summarised by Goodacre and Wallace.\textsuperscript{6} Persistence of the buccopharyngeal membrane, amniotic constriction bands in the region of the developing branchial arches, environmental insults, drugs such as meclozine and large doses of vitamin A have been some of the postulated causes. Failure of mesodermal migration into the midline structures of the mandibular portion of the first branchial arch is believed to be the embryological basis of the defect. Failure of fusion of the first arch prevents succeeding arches from uniting, as fusion proceeds from above downwards.\textsuperscript{14} This explains the bifid anterior two-thirds of the tongue. The presence of this mandibular cleft with a bifid tongue causes crowding of the contents of the oral cavity during development. As the tongue is bifid, the medial ends of the hemimandible are pushed against the palate causing adhesions and fusion.\textsuperscript{6}

The treatment of maxillomandibular fusion has conventionally been urgent surgical division.\textsuperscript{6} However, successful delayed surgical correction has also been reported recently.\textsuperscript{11} The rarity of this condition makes standardisation of treatment difficult. In our case, dissection of the bony fusion relieved the trismus but the child succumbed to aspiration pneumonitis.

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References


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