ORIGINAL RESEARCH PAPER

Paediatrics

CONGENITAL UNILATERAL HYPOPLASIA OF DEPRESSOR ANGULI ORIS ALTHOUGH A MINOR AND RARE ANOMALY BUT IS ASSOCIATED WITH THE MAJOR UNDERLYING DEFECTS OF VARIOUS SYSTEMS.

KEY WORDS: Congenital Unilateral Hypoplasia, Depressor Anguli Oris Muscle Daom, Anomalies

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ABSTRACT

Congenital unilateral hypoplasia as one of the cause of asymmetry of face was first described by Parmalee in 1931(1). It presents as a facial asymmetry which become prominent while child smiles or cry. (4) Diagnosis is established by clinical examination and electromyographic studies.(5) This condition is associated with various other major anomalies. A 10 month old male child reported to the tertiary care institute with urinary complaint and on examination had asymmetry of face becoming pronounced during crying and smiling, the angle of mouth of child deviated towards right side. Rest of face was normal. Renal ultrasound revealed bilateral hydronephrosis. Asymmetrical appearance of face due to congenital hypoplasia of depressor anguli oris is a rare condition but has high association with the other major anomalies. So paediatrician needs to be aware of the underlying anomalies associated with it.

Introduction

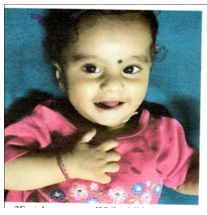
Congenital unilateral hypoplasia as one of the cause of asymmetry of face was first described by Parmalee in 1931(1). It is estimated to occur in 0.25% to 0.6% of infants. (2, 3) It presents as a facial asymmetry which become prominent when child smiles or cry. (4) Diagnosis is established by clinical examination and electromyographic studies.(5) This condition is associated with various other anomalies like circulatory, urogenital, bone, muscles, gastrointestinal, CNS and skin etc. (6) So it is important to recognize the condition and look for the underlying anomalies so that early treatment can be started. In the present case we intend to describe this case of asymmetrical face due to hypoplasia of depressor anguli oris muscle and its association with underlying anomalies.

Case report

A ten month old child reported to paediatric clinic for the evaluation of fever and hematuria. Mother noticed that child was passing red coloured urine one and half month back which subsided after two to three days and again recurred five days back. There was history of associated fever during the episode of haematuria. Antenatal period was uneventful and child was born by emergency lower section caesarean section in view of meconium stained liquor, child cried immediately after birth. The post natal period was uneventful. No history of similar asymmetry in the other family member. On examination child had high grade fever documented up to 103 0 F, anthropometric parameters were normal.

Child had symmetrical face while child was quiet as shown in figure -1 while had deviation of lower lip during smiling and crying as shown in the figure 2 and 3. Facial nerves were normal bilaterally as determined by furrowing / forehead wrinkling, nasolabial fold depth was normal. Neuro-developmental examination was normal. Echocardiographic examination did not show any abnormality.

Figure -1 Showing Symmetrical face of child during quiet state.



Fingue-2

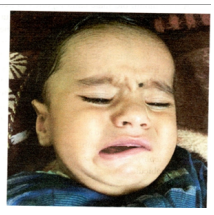


Figure -3

Figure 2 Showing the facial asymmetry during the smiling and figure 3 showing facial asymmetry during crying of child.

On abdominal examination cystic swelling was noticed in the infraumbilcal region. Ultrasound abdomen was done which showed bilateral hydronephrosis with raised cortical echogenicity, distended urinary bladder with mild wall thickening with trabeculation.

Discussion

Congenital hypoplasia of facial muscles is a rare condition. Cases of unilateral hypoplasia of depressor anguli oris have been reported, where the facial asymmetry is due to the unopposed action of opposite normal side muscle, the angle of mouth is pulled towards the normal side during crying and smiling.

Although these are minor anomalies and not detrimental to the health but are associated with the major underlying defects of various systems. [7] Our case has unilateral hypoplasia of depressor anguli oris muscle of left side and associated urogenital abnormality, the bilateral hydronephrosis, raised cortical echogenicity, distended urinary bladder with mild wall thickening and trabeculation was found on ultrasonography.

The pathogenesis of unilateral hypoplasia of depressor anguli oris muscle is not known. [8] The various causative factors have been suggested as intrauterine moulding, viral infections during the pregnancy. Due to the high incidence of affection seen in the first degree relative of the cases, the role of hereditary factors in aetiology has also been suggested.[5,9] Differential diagnosis of this condition includes facial nerve palsy and obstetric related compression or trauma. Congenital hypoplasia of depressor anguli oris affects only lower lip while other functions of facial nerve are intact [10] Diagnosis can be established by clinical picture and electromyographic studies.[5] Prognosis of congenital unilateral hypoplasia of depressor anguli oris has been poor.

Hypoplasia of DAOM is associated with anomalies of various systems like circulatory, urogenital, bone, muscles, gastrointestinal, CNS; respiratory system etc. [6] Cardiovascular and head and neck anomalies have been most common. The basis of all these associations might be due to their common embryonic origin, as all of these systems develop from the various subdivisions of embryonic mesenchyme. [11] The paediatricians need to be aware of the underlying anomalies associated with it so that children can be carefully investigated for the associated major anomalies.

Conclusion

Asymmetry of facial appearance due to the congenital hypoplasia of depressor anguli oris is a rare condition. Although these are minor anomalies and not detrimental to the health but are associated with the major underlying defects of various systems. So paediatricians need to be aware of the underlying anomalies associated with it so that children can be carefully investigated for the associated major anomalies.

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