Case Report

A CASE OF FACIAL CLEFT ASSOCIATED WITH MAXILLARY SINUS HYPOPLASIA

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ABSTRACT

Observation of an attendant of a patient, a 55 year old woman, exposed a case of anatomical curiosity, an uncommon variant of a developmental anomaly, an atypical facial and palatal cleft, and a rare association of a hypoplastic maxillary sinus. The woman gave history of effectively managing the deformity with certain habituated maneuvers of the tongue, without undergoing any surgical, cosmetic or functional correction. Such cases need to be documented to create awareness as these developmental pathologies with sinus opacity may be misdiagnosed as sinusitis or neoplasm.

Key Words : Facial cleft, Frontonasal process, Medial and Lateral nasal processes, Maxillary sinus hypoplasia.

CASE REPORT

A fifty five year old lady, an attendant of a patient in ENT department, Vinayaka Mission's Kirupananda Variyar Medical College & Hospital, Salem, was observed to have a defect at the junction of the ala of nose and the cheek on the left side of the face. On persuasion the woman agreed for a thorough examination.

On examination an oblique facial cleft was found at the junction of the ala of the nose, upper lip and the cheek (Fig.1).The subject was habituated to hide this gap under the ala with little upward movement of the upper lip. There was no clefting of the lip. Through the facial cleft, the tongue movements were visible. The malar prominence on the left side was normal.

Oral examination showed an oronasal cleft with the absence of the palatine process of the maxilla (Fig.2). Corresponding alveolar process of the maxilla was also missing. The horizontal process of the palatine bone was normal with the soft palate attached to its posterior border.

Nasal examination revealed rudimentary turbinates on the left side of the nasal cavity. The uncinate process was found to be hypoplastic. The premaxilla on the left side was partially developed.

Radiographic findings confirmed these defects and also the hypoplastic left maxillary sinus (Fig.3, Fig.4). Because the facial defect was hidden by the ala of the nose, not much of cosmetic problem was faced by the woman and she was habituated to certain tongue movements to cover the palatal gap to overcome the functional difficulties of hypernasal voice and nasal regurgitation. She refused corrective surgery.

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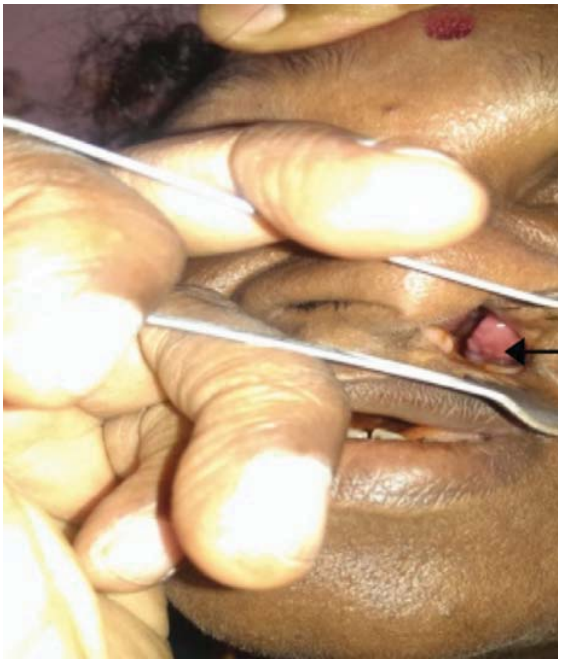

Figure-1. Oblique facial cleft.

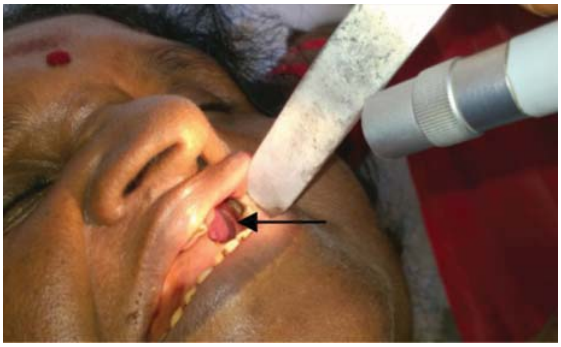

Figure-2. Defective palate.

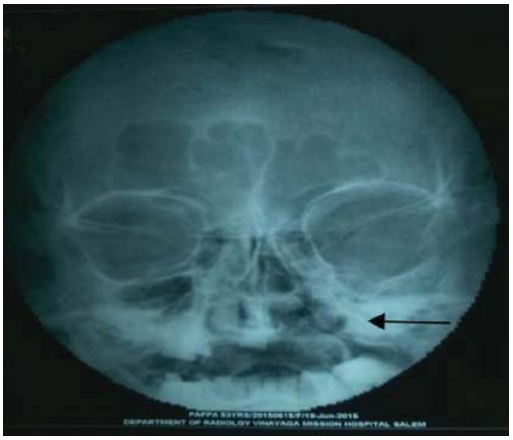

Figure-3. X ray Skull, PNS View. Absence of maxillary air sinus.

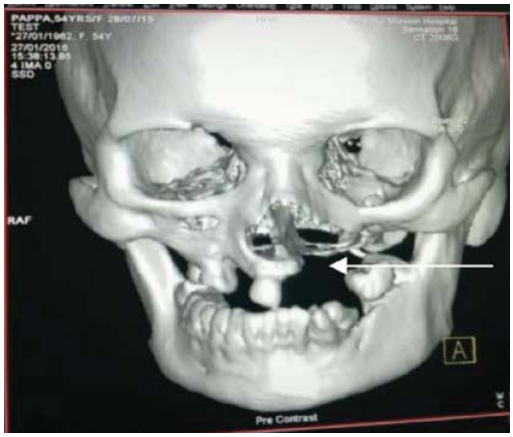

Figure-4. 3D CT View. Absence of palatine & alveolar processes.

DISCUSSION

Craniofacial development is a complex series of events that occur between the third and eighth weeks of gestation.[1,2] At the end of 4th week of development, five facial swellings surround the stomodeum, one central frontonasal process and a pair each of maxillary and mandibular processes on the sides. The maxillary and mandibular processes are subdivisions of first branchial arch. The frontonasal process is the downward proliferation of the mesoderm over the forebrain. The medial and lateral nasal processes develop from the frontonasal process.

During the 6th and 7th weeks the maxillary processes on either side increase in size and grow medially.[3] On medial migration, the maxillary process first fuses with the lateral nasal process and then with the medial nasal process, thus completing the anterior choana. The two medial nasal processes fuse in the midline to form the nasal septum, intermaxillary process (primary palate), the philtrum and central part of upper lip. The developmental aberration noted in our subject was failure of fusion of the maxillary process and lateral

nasal process on the left side. The floor of the nasal cavity which is the hard palate is formed during the 8th week. The two palatine processes of the maxilla develop from the medial aspect of the maxillary processes and grow downwards initially.[4] At the end of 9th week they rotate upwards to a horizontal position and fuse with each other in the midline and also with the primary palate anteriorly, to form the secondary palate. In our subject this part of the palatine process of the maxilla on the left side was not developed.

The lateral nasal processes also grow backwards to form the lateral nasal walls. In our subject this was defective, leading to the hypoplasia of the uncinate process. Hypoplasia of uncinate process can be associated with maxillary sinus hypoplasia.[5] Three severity levels of hypoplasia are described in literature. Type I is mild to moderate hypoplasia, with normally developed uncinate process and well developed infundibular passage. Type II is with a hypoplastic uncinate process with ill-defined or absent infundibular passage.[6] Type III is characterized by an absent or very hypoplastic uncinate process and profound sinus hypoplasia or aplasia. Our subject belongs to the third category.

More than one theory exists regarding the embryologic pathogenesis of facial clefts. Multiple genetic and environmental factors may be involved.[7] The environmental risk factors include antenatal exposure to radiation, viral infections, metabolic abnormalities and teratogenic compounds. [8]

CONCLUSION

It is important to be aware of association of maxillary sinus hypoplasia with facial clefts. The opaque maxillary sinus on radiological investigations can lead to misdiagnoses as sinusitis or neoplasia. Diagnosing maxillary sinus hypoplasia pre-operatively may prevent complications such as orbital injury, while performing uncinectomy during endoscopic sinus surgery.

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