

MEDIAN CLEFT OF THE UPPER LIP IN ASSOCIATION WITH FRONTAL NASAL MENINGOCELE, LEFT ANIRIDIA AND LENTICULAR OPACITY

Pages with reference to book, From 43 To 45

Abdul Rashid Khawja, Abdul Rashid Chaudhry, Nasir Saeed, Ijaz Ahmed (Department of Surgery, Rawalpindi Medical College, Rawalpindi.)

Abstract

Two cases are reported. One child was male and other a female. Anomaly is rare. This anomaly was found in 2 out of 500 cases (0.4%) in the present series of cleft lip, (1979-1984). Repair of lip is easy as compared with other cleft lip reconstructions(JPMA36.43 1998).

CASE 1

I.N. female full term, 10 Lbs at birth, child of non consanguinous parents. Baby was first seen on the third day after birth, No family history of cleft lip or other anomalies was present. Mother had not taken any drug, had not suffered illness or radiation and had not been vaccinated, during pregnancy. The child had a median cleft of upper lip. There was also a cleft of alveolus and a cleft of hard and soft palate Figures 1,2).



Figure 1. Median cleft of upper lip.



Figure 2. Cleft of hard and soft palate.

An anterior meningocele, arising from cribriform plate, presented itself in Thasopharynx through the cleft palate. X-Ray of the base of skull showed a linear defect in the median plane of the anterior cranial fossa. There was no evidence of holoprosencephally. In this case we found complete median assymetry, starting from the lip to base of skull. Since no facilities for CAT scan and transillumination

were available, these were not carried out.

Lip cleft was repaired at the age of 12 weeks. Palate repair was postponed till the correction of meningocele.

CASE 2

U.K. was an 11 month old, male, healthy child of consanguinous parents, product full term and normal delivery. No history of medication, radiation or illness of the mother during pregnancy was present.

Median cleft (Figure 3)

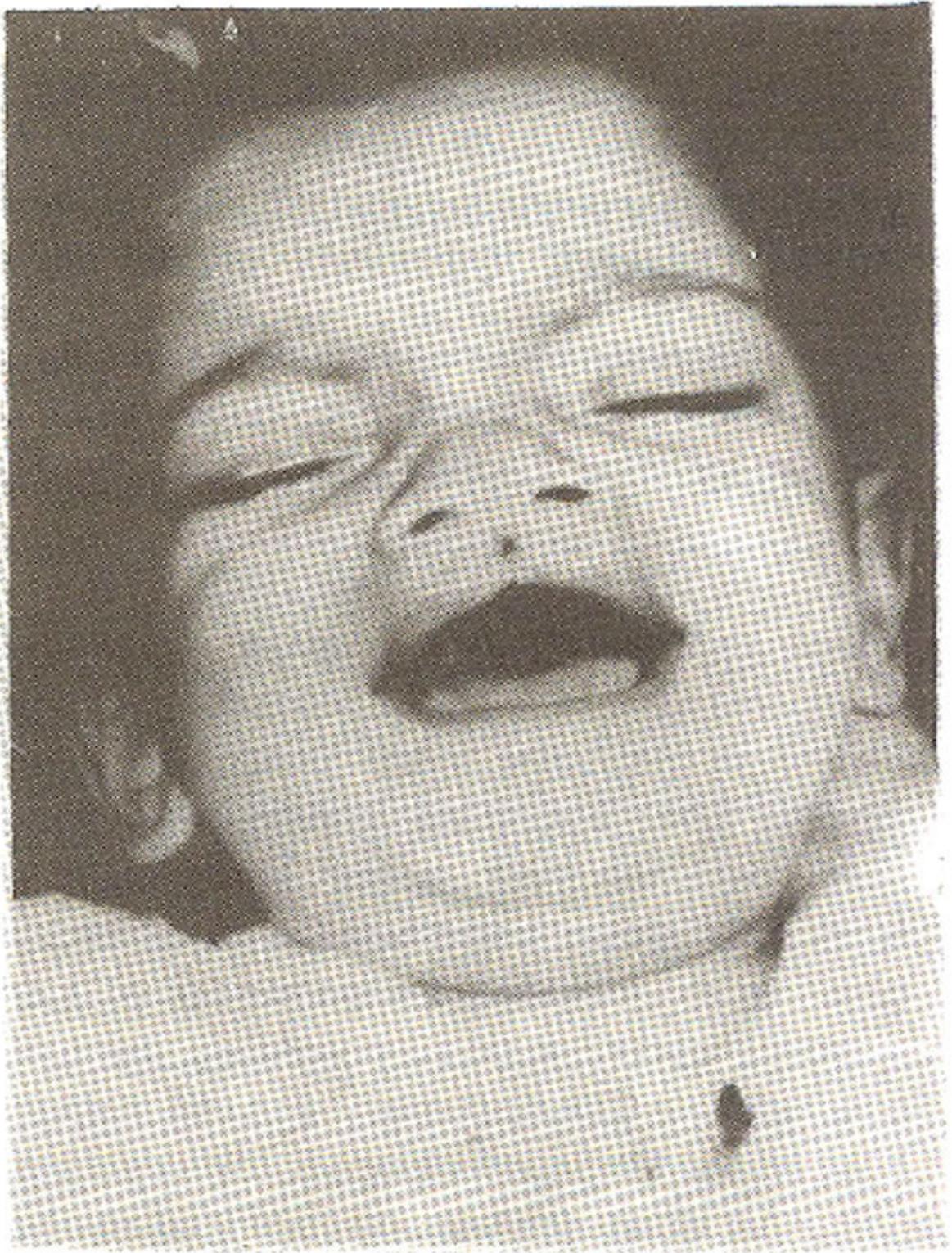


Figure 3. Median cleft with partial cleft of the alveolus.

was noticed by mother at birth, and her physician had referred the child to us for correction at the age of 11 months. Mother had noticed an opacity in the left eye, and the ophthalmologist was treating the case as aniridia and lenticular opacity. Right eye was normal. There was partial cleft of alveolus, no other abnormality was detected. Cleft lip and alveolus were

corrected with good result.

DISCUSSION

The first case of median cleft associated with other abnormalities was reported in 1909.¹ Median cleft lip is a very rare condition and is due to failure of downward growth of mesoderm in the median nasal process.² We have the above 2 cases in our series of 500 cases treated in past 6 years. Five cases have been reported only previously of median cleft associated, with polydactyly, syndactyly and toe anomalies.³

The presentation is unique in the sense that both cases presented with associated abnormalities of anterior process of the face and of anterior cranial fossa. No other abnormalities were detected in these children. Both these children were mentally alert. Milestones of growth and development were normal. There was no sign of mental deficiency in these children.

In view of the clinical findings of frontal asymmetry and associated holoprosencephaly, we intended to confirm it with CAT scan and transillumination tests, but these facilities were not available.⁴

Radiographs of base of skull showed a linear defect in the linear plane of anterior cranial fossa.⁵

Repair of median cleft is easy, we used a bilateral median longitudinal incisions, with bilateral lateral advancement 3mm above the vermilion border. Results were good.

REFERENCES

1. Thurston, E.D. A case of median hare-lip associated with other malformations. *Lancet*, 1909; 2:996.
2. Christie, A.B. and Mustarde, J.C. *Surgery in infancy and childhood*. 3rd ed. Edinburgh, Churchill Livingstone, 1974, p. 428.
3. Christophorou, M.N. and Nicolaidou, P. Median cleft lip, polydactyly, syndactyly and toe anomalies in a non-Indian infant. *Br. J. Plastic Surg.*, 1983; 36:447.
4. Burian, F. *Syndromes of the head and neck*. Edited by Gorlin, R.J. and Pindborg, J.J. 2nd ed. New York, Mc Graw-Hill, 1976.
5. Khoo, C.T.K. and Sand, M.N. Median cleft of the upper lip in association with bilateral hexadactyly and accessory toes. *Br. J. Plastic Surg.*, 1980; 33:407.