Images in Clinical Practice

Goldenhar Syndrome

A 1*1/2*-year-old boy presented with macrostomia, preauricular appendages, low set ears, epibulbar dermoids and polydactyly, all occurring bilaterally *(Figs. 1&2).* Another 15-days-old boy presented with first and second branchial arch clefts on the right side with epibulbar dermoids *(Fig. 3). A* diagnosis of Goldenhar syndrome was made in these boys.

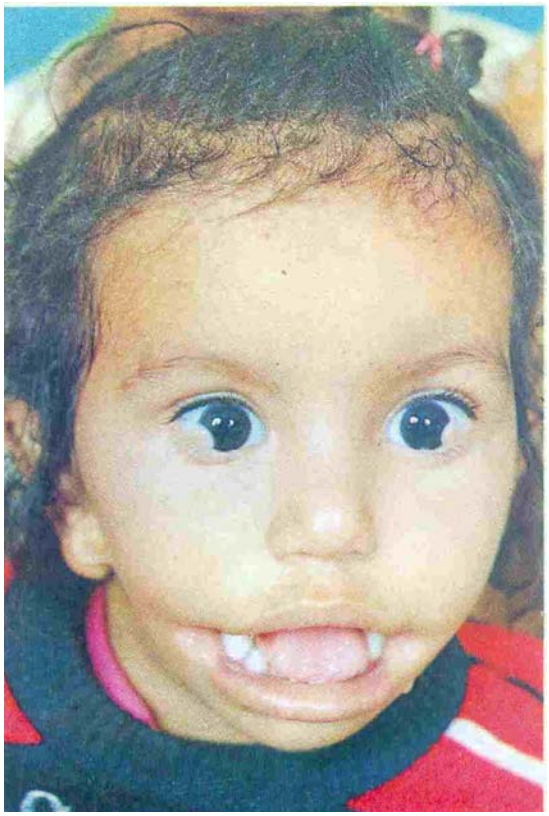

Fig 1. An 18-month-old child with macrostomia, preauricular appendages, low set ears and epibulbar dermoids, all present bilaterally.

Goldenhar syndrome (oculoauricular syndrome) forms a subset of first and second branchial arch syndrome *or* lipodermoids of the conjunctiva or cornea, and preauricular appendages usually anterior-to the tragus and associated with low set ears. Macrostomia may be present sometimes. Other vertebral, cardiac and pulmonary anomalies may occur(2). The features of Goldenhar syndrome are usually present unilaterally; although 5% to 8% may be bilateral(3). The mode of inheritance is uncertain. The syndrome may share a number of features with Treacher-Collins syndrome and, in fact, combinations of these two syndromes have been reported. The management depends on several factors including the extent of facial hypoplasia(l). Most epibulbar dermoids can be easily excised for cosmetic benefits. When the cornea is involved, lamellar keratectomy gives moderately satisfactory results(2).

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2. Brazier J. Craniofacial. abnormalities. *In:* Pediatric Ophthalmology. Ed. Taylor D. Boston, Blackwell Scientific Publication, 1990; pp 162-221.

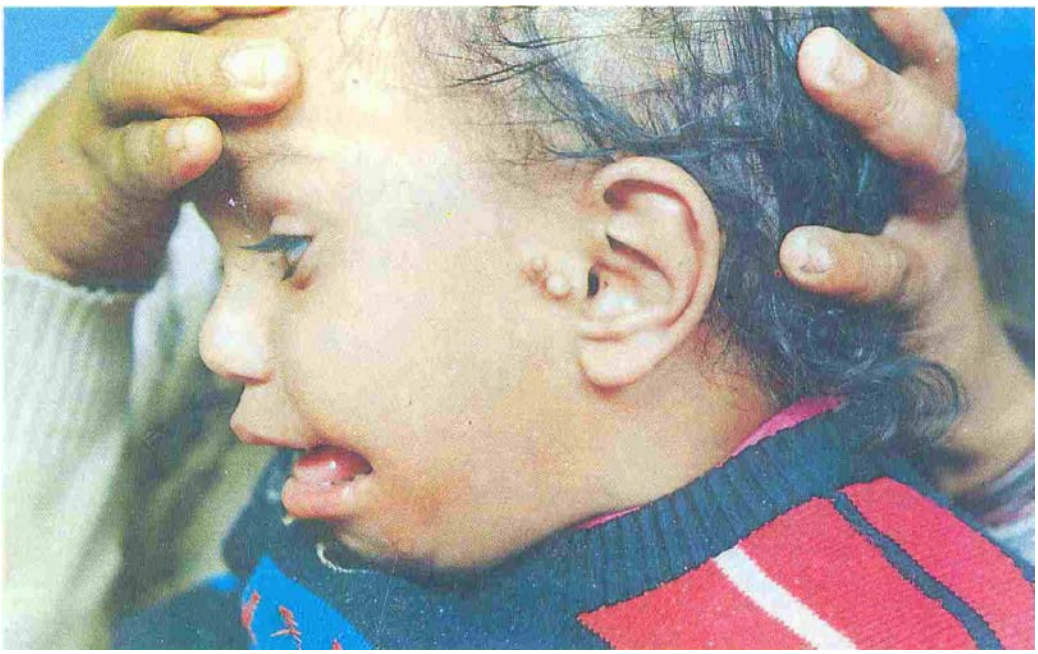

Fig. 2. *Lateral view of the same child showing preauricular appendages and low set ears.*

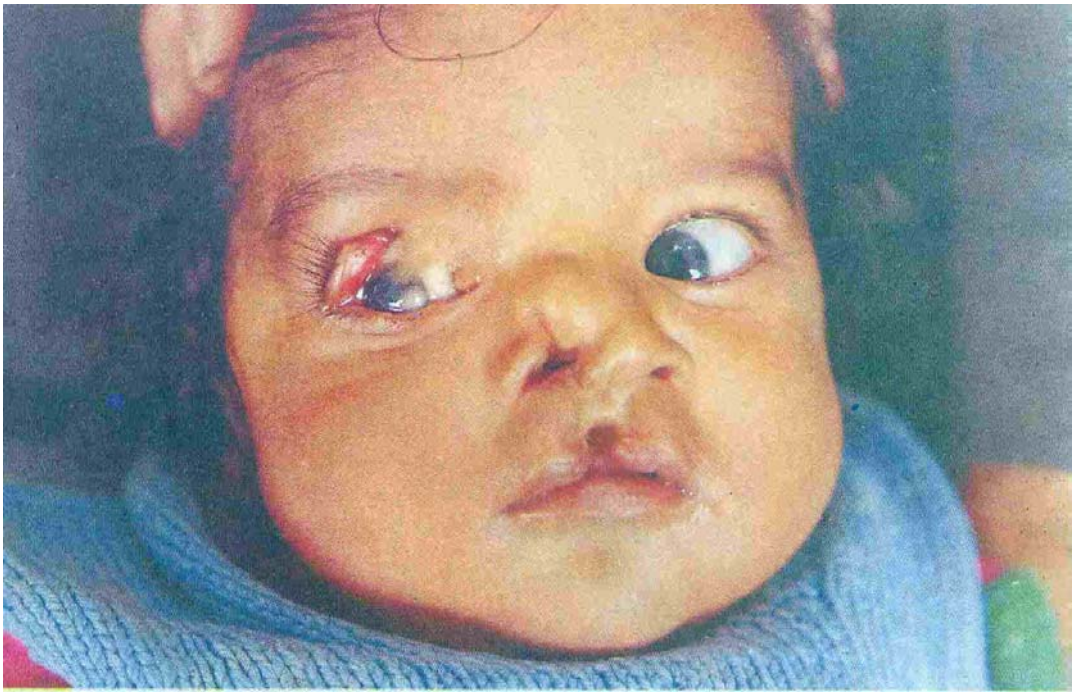

Fig. 3. *A 15-day-old child with first and second branchial arch clefts on the right side with epibulbar dermoid.*

3. Sargent RA, Ousterhout DK. Ocular manifestation of skeletal disease. In: Pediatric Ophthalomology, 2nd. edn. Ed. Harley RD, Philadelphia, W.B. Saunders Co, 1983; pp 1041-1049.