Goldenhar Syndrome: A rare case report
Hamillour Bochra, MD1; A.H Khadri, MD2; A.ABensebiti, MD; Z Atrih MD
1University of Constantine 3 . Algeria, Department of Pediatric surgery.

ABSTRACT

Goldenhar syndrome or oculo-auriculo-vertebral dysplasia is a complex syndrome characterized by a combination of maxillomandibular hypoplasia, ear deformity, auricular dermoids and vertebral anomalies.

INTRODUCTION

Goldenhar's syndrome, or oculo-auriculo-vertebral syndrome, is a poly-malformation syndrome associated most often an epibulbar dermoid, pretragial diverticula and vertebro-ocular anomalies. We report a rare observation of Goldenhar syndrome and vertebral anomalies

OBSERVATION

2 years-old patient reported to the Department of pediatric surgery, Constantine Algeria, with a transverse cleft present in the left angle of mouth, with incompetent lips epibulbar dermoid cyst protuding from the right eyelid. the patient was referred to the ophthalmic department, where she underwent a simple excision of limbic tumors (Fig.5.6.7.8).

RESULTS

The postoperative evolution was very favorable from an esthetic point of view, without complication ;a retreat of a year later. The intervention under general anesthesia was done, in which the repaired orbicularis oris muscle was reconstructed by reconstruction of the angle of the mouth and the lateral cleft lip, by demarcation of the orbicularis, preauricular adnexal appendage and pre-auricular diverticula. No family history or other symptoms were recorded. The baby, had a normal auditory meatus without any evidence of skin and bone abnormalities stained, radiograph of the spine, echodoppler did not show any abnormal vascularization.

DISCUSSION

Goldenhar's syndrome is a rare malformation syndrome related with an anomaly of development of the first gill arches. Although most cases of GS were sporadic, the modulation of autosomal recessive inheritance and the dominant have been described. Also referred to as oculo-auriculo-dysplasia, it is most often unilateral affecting the soft tissues and the bone component. It usually include cleft lip and palate of different stages, as well as cardiac and genitourinary malformations, cleft lip or palate of different stages, delicate care.

CONCLUSIONS

Goldenhar syndrome is a rare malformation disease. Clinically stands as a combination of several abnormalities such as limbal tumors, malformation of the ears of different degrees, as well as cardiac and genitourinary malformations, cleft lip or palate of different stages. delicate care.