

Discussion: JXG is a rare and typically benign disease that presents in children. Small isolated forms are under-diagnosed.³ Our case typically belongs to the 2nd variant and can be a cause psychosocial disability. The aetiology is unknown – Non neoplastic process characterized by an abnormal response to a non-specific injury e.g trauma or viral infection. Common systemic associations are Juvenile chronic myelogenous leukemia, Neurofibromatosis, urticaria pigmentosa, Insulin-dependent DM, Aquagenic pruritus, CMV infection. Serious complications can arise when systemic juvenile xanthogranuloma is involved. Simple excision biopsy yields good results in uncomplicated cases.

Conclusion: Bilateral eyelid swelling could result from Juvenile Xanthogranuloma. Histo-pathologic diagnosis and careful examination to rule out systemic association is required. Complete resolution can be achieved after excision biopsy.

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Goldenhar Syndrome: A Case Report

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Introduction: Goldenhar Syndrome, (GS) also called oculoauriculovertebral dysplasia is a rare congenital disease arising from abnormal development of the first and second branchial arches¹⁻³ Incidence is 1 in 3500-5600^{4,5}, male to female ratio of 3:2¹, 85% unilateral presentation^{1,5}. Multifactorial risks factors implicated are genetic^{1,6}; deletion of 5p, 14q23 duplication, Chr18,22 anomaly and 14q23 duplications are implicated. Others are gestational diabetes, second trimester bleeding, multiple gestation and maternal ingestion of vasoactive drugs: aspirin, pseudoephedrine, accutane, ibuprofen⁶⁻⁸ and alcohol.⁹ Viral⁵, environmental and other unknown factors are suspected as well. Clinical features include hemifacial microsomia, facial asymmetry maxillary hypoplasia, malar flattening⁴. Ears may show, accessory tragi, tags, meatal atresia, low set ears, microtia and ⁶ laryngomalacia^{4,6,10}. Spine may have scoliosis and hemivertebrae¹⁰. Cardiac defects include ventricular septal defects (VSD) and transposition of great vessels^{4,6,10}. Tracheoesophageal fistula and gastrointestinal (GIT) defects are associated^{4,6,10}. Central nervous system (CNS) defects include hydrocephalus, hypoplastic corpus callosum^{4,6,10}. Ophthalmic features occur in 60%¹¹ and include epibulbar dermoid, usually located infero-temporally. Others are upper lid coloboma, unequal palpebral fissures, subconjunctival dermoid, uveal coloboma, retinal coloboma, cataract, strabismus, microphthalmia and anophthalmia^{4,6}. There is an association with glaucoma⁴. Diagnosis is usually by phenotypic appearance and radiological investigations (Computed Tomography, Magnetic Resonance Imaging)^{3,4}. Genetic tests help to support the diagnosis and Ultrasound scans aid prenatal diagnosis¹. Treatment is multidisciplinary with interventions depending on age, severity and extent¹. Early intervention may be required in airway problems such as obstructive sleep apnoea,

eyelid defects, jaw problems affecting nutrition, and hearing problems¹¹. Ophthalmic intervention is to prevent potential visual loss, amblyopia and enhance aesthetics¹². Corneal protection,

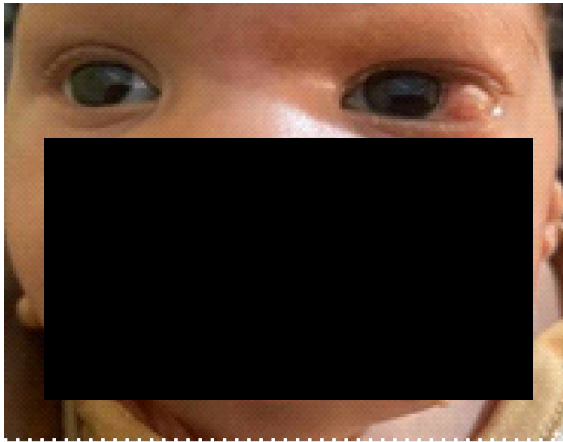


Figure 1: Left epibulbar growth and face tag



Figure 2: Left low-set, mis-shapen pinna and ear

therapeutic contact lens use and lubricants are used conservatively¹. Surgical lid repair of coloboma depends on its size, location and general health of the patient¹³. Repair may be varied techniques of direct apposition, to sliding flaps and more extensive reconstructions¹. Limbal choristomas excisions, with a consideration for their adhesions to cornea with lamella keratoplasty or amniotic membrane grafts are done^{1,12,14}. Careful excision of lipodermoids to prevent conjunctival shortening is indicated¹⁵. Social support is necessary, due to the psychosocial effects on the patient⁷.

Case Report: A 3week old female neonate is presented, with abnormal growths noted on the

face, left eye and misshapen ears since birth. She was the fifth child in a nonconsanguineous union with unremarkable ante- and perinatal history and no maternal vasoactive medications, fever or diabetes. Both eyes followed light well, with normal corneal reflexes. Eyelids were normal. The left eye showed temporal epibulbar dermoid (Figure 1). Anterior and posterior segments were normal in both eyes, with facial skin tags, left microtia with ear tags both sides (Figure 2). Further paediatric evaluation showed umbilical hernia, cardiomegaly, ventricular septal defect, lower GIT Hirschsprung defect. There were no skeletal defects. Plastic repair of ear, audiological intervention and later repair of epibulbar growths were scheduled.

Discussion: The spectrum of GS ranges from mild to severe¹⁶. Additional testing to detect systemic complications is vital. Individualized approach is adapted by age and severity. Anatomic and functional maintenance/restoration of ocular surface, vision and ocular motility is key. Craniofacial, vertebral airway and cardiac effects may pose anaesthetic risks at reconstruction¹⁷ and require multidisciplinary input.

Conclusion: Correct identification and anticipation of potential associated complications, with multidisciplinary staged management is crucial for future quality of life enhancement in these patients.

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Anophthalmic Socket: Clinical Presentation, Complications, Risk Factors, Management and Challenges in Lagos, Nigeria

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Background: An anophthalmic socket is devoid of an eyeball, oftentimes following non - salvageable ocular injuries, severe ocular infections, and intraocular malignancies. It could be associated with unacceptable functional, aesthetic deficits and impaired quality of life. It also constitutes management challenges to the oculoplastic surgeon. Literature is sparse on anophthalmic socket in Lagos, Nigeria. This study aims at assessing the clinical presentation, complications, risk factors, management, and challenges of acquired anophthalmic socket with the goal of providing a data base and suggest appropriate management guidelines

Methods: A prospective cross -sectional hospital - based study was conducted between January 2018 and April 2022. All patients presenting with acquired anophthalmic socket