INTRODUCTION

Interruption of lid development during normal embryogenesis may cause a wide spectrum of congenital anomalies, however congenital eversion of eyelids are rarely seen. Its etiology is unknown, however it is frequently associated with Down syndrome. In this report we present bilateral eyelid eversion in a baby with Down syndrome to discuss its course and management.

CASE REPORT

A 3- days- old female infant, born after an un- complicated pregnancy and delivery presented to our clinic with bilateral chemosis protruding from the everted, upper eyelids which obstructs the visual axis bilaterally (Figure 1).

Physical examination of the baby revealed normal findings except simian crease and drop ear deformity which warrants a cytogenetic study to investigate Down syndrome. Cytogenetic findings with G banding method demonstrated a regular type Down syndrome (47 XX, +21). MR Angiography showed bilateral well circumscribed lesions which demonstrate hyperintensity on T2 weighed images and homogeneous enhancement after administration of contrast agent. Conservative treatment with topical lubricants and ointments in addition to eyepatching after repositioning the eyelids were applied for 2 weeks. Unfortunately no improvement was observed. As visual axis was occluded totally, we decided to perform en bloc excision to prevent deprivation amblyopia.

In her examination, under general anesthesia, before the operation, both eyes were normal in size.
and no abnormality was noted in the anterior segment structures. Intraocular pressures measured with Perkins tonometer were within normal limits and both fundi were unremarkable. Chemotic conjunctiva was excised and sent to the laboratory for pathological evaluation. The operation has been completed following the lid reconstruction with a wedge resection at the lateral canthus for both eyes (Figure 2).

Unfortunately the baby developed cardiac arrest a few hours after the operation which answered to resuscitation initially. During his observation in the newborn unit cardiac insufficiency and lung infection developed within the following days, necessitating mechanical ventilation and respiratory support. Despite all efforts the patients condition deteriorated resulting in exitus in the 35th day after birth.

Although parents of the baby signed the consent form for publication, they did not allow a postmortem autopsy. The histopathological analysis of the excision material revealed dilation of capillaries and hemorrhage. This finding may be explained by congestion of the conjunctiva due to eversion of the eyelids.

**DISCUSSION**

Down syndrome, known as the most common chromosomal anomaly, encompasses numerous ocular abnormalities like epicanthal fold, hypertelorism, epiblepharon, ectropion etc. Among these eyelid deformities constitute a wide spectrum like upward slanting of the palpebral fissure, hypertelorism, epicanthus, epiblepharon, ectropion, and upper lid eversion.\(^1\)\(^3\) Congenital eversion of the upper eyelids was first described by Adams in 1896 who called it as “double congenital ectropion”\(^4\). This acute ectropion is reported most frequently in Down syndrome, black babies and difficult deliveries.\(^5\) Abnormalities like orbicularis hypotonia, birth trauma, vertical shortening of the anterior lamella or vertical elongation of the posterior lamella of the eyelid and failure of the orbital septum to fuse with the levator aponeurosis, absence of effective, lateral canthal ligament and lateral elongation of the eyelid have all been implicated as possible pathophysiological factors.\(^6\)

This bilateral condition becomes usually evident intermittently when the child cries. Venous stasis
during delivery may cause marked chemosis and prolapse of the conjunctiva which may obscure the globe. This may recover spontaneously. Surgery is recommended only if initial attempts of pressure patching or repositioning of the lids and taping fails.

In our case no remarkable change was noticed during the conservative treatment period. Surgical excision was undergone to avoid obstruction of the visual axis. In the literature other surgical treatment options like temporary tarsorraphy, subconjunctival injection of hyaluronic acid, fornix sutures and full thickness skin graft to the upper eyelid are reported. We preferred to excise the excess conjunctiva to confirm the diagnosis.

Upper eyelid eversion is a rare abnormality and may threat the vision if not treated early. As it is to our knowledge synaptic connections are established permanently during the critical period for visual development and a retarded treatment may be less effective for restoring vision. However risks of general anesthesia should be also kept in mind when deciding the for surgical treatment.

REFERENCES: