Crocodile Tears and Dandy-Walker Syndrome in Cervico-oculo-acoustic Syndrome

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INTRODUCTION

A 13-year-old girl with crocodile tears and Dandy-Walker syndrome in addition to Duane’s retraction syndrome, Klippel-Feil syndrome, and deafness is presented. To our knowledge, association of Dandy-Walker syndrome and crocodile tears with this rare syndrome has not been reported previously.

Cervico-oculo-acoustic syndrome (COAS) was first described by Willervanck in 1952. The syndrome is characterized by the triad of Klippel-Feil syndrome, Duane’s retraction syndrome, and congenital hearing loss.1,2

Klippel-Feil syndrome, a congenital malformation, is a decreased number of and partial or complete fusion of the cervical and, occasionally, upper thoracic vertebrae. The classic clinical triad is composed of an extremely short neck with limited movement and a low posterior hair line.3

Dandy-Walker syndrome includes malformations of the fourth ventricle and cerebellum. Various cerebral anomalies, such as enlargement of the fourth ventricle, hypoplasia or agenesis of the posterior vermis, rudimentary cerebral hemispheres may accompany Dandy-Walker syndrome.4 The association of Dandy-Walker syndrome and COAS has not been reported.

Duane’s retraction syndrome in its classic form is characterized by severe limitation of abduction, slight limitation of adduction, globe retraction, and narrowing of the palpebral fissure on abduction. Variations of this special form have been described.5

The present report describes a case with multiple congenital anomalies in addition to complete triad of COAS.

CASE REPORT

A 13-year-old girl admitted to the Department of Ophthalmology, Çukurova University with a complaint of lacrimation while eating and squint since birth. She was born spontaneously at term with low birth weight after an uncomplicated pregnancy. Her personal history revealed developmental delay: she walked at 36 months and talked at 48 months. There was no history of familial anomalies.

Ophthalmologic examination revealed a visual acuity of 20/20 in both eyes. Slit-lamp and fundus examinations were within normal limits. Ocular motility examination revealed bilateral Duane’s syndrome with limited abduction, globe retraction, and narrowing of the palpebral fissure on adduction (Figures 1, 2). We also observed tearing while chewing.

On physical examination, she was of short stature (1.35 m) with a head circumference of 55 cm (Figure 3). Her neck was extremely short with severe restriction of neck movements. Posterior hair-line was low; thoracic scoliosis was present. There was a 6×5-cm mass in the neck (Figure 4). We observed mirror movements (synkinesis) of the upper extremities.

Audiometric examination revealed mixed hearing loss in the left ear. Assessment of her intellectual function revealed moderate mental retardation. Examination of cardiac, gastrointestinal, and urogen-
ital systems did not reveal any pathologic finding.

Plain radiography revealed complete fusion of C-1, C-2, and C-3 vertebrae including dense, partial fusion at the corpus of C-4 and C-5 vertebrae (Figure 5). We also observed thoracic scoliosis and sacral spina bifida. Partial fusion was found in the thoracic vertebrae. We observed dense hypoplasia, an enlarged foramen magnum, spina bifida of C-1 and C-2, encephalocele, lateral thoracic meningocele, and bilateral loss of mastoid aeration in computed tomographic examination (Figure 6). Cervical and cerebral magnetic resonance imaging revealed Dandy-Walker syndrome, cerebellar hypoplasia, vermian agenesis, corpus callosum splenium dysgenesis, cranium bifidum, asymmetry of the cranium especially at the posterior fossa, partial platybasia, an encephalocele at the proximal cervical region, hydromyelia, and a defect in the posterior proximal part of the medulla spinalis (Figures 7,8).

**DISCUSSION**

Sporadic occurrence of unknown etiology is common. A careful evaluation of family members is indicated, as autosomal dominant inheritance with complete penetrance and variable expressivity has been suggested. There is a tenfold female to male
Our female patient's family had no history of any congenital malformations. Cases with an incomplete classical triad of COAS have been described in literature. The patient presented here had multiple congenital anomalies in addition to the complete triad of COAS.

Wildervanck stated that deafness should be sensorineural in type and tomographic examination demonstrated an abnormality of the vestibular labyrinth in most of these cases. However, cases with conductive or mixed losses have been reported. Thus, in our case, hearing loss was mixed in type and tomographic examination did not reveal a pathological finding.

Dandy-Walker syndrome consists of cystic transformation of the fourth ventricle, rudimentary cerebellar hemispheres, and hypoplasia or absence of posterior vermis. In addition to these findings, our case had dysgenesis of the corpus callosum splenium, hydromyelia, and encephalocele.

Wildervanck's syndrome is known to be the most common multiple congenital abnormality found in association with Duane's syndrome. The lateral thoracic meningocele is the protrusion of the spinal meninx from the defect of vertebral columns or foramina. The collaboration of this rare spinal malformation with neurofibromatosis is common.

Crocodile tears is a unilateral or bilateral secretion anomaly of the lacrimal gland. Anticipation or taste of food provokes excessive tearing. This gustatory lacrimal reflex is explained by misdirection of regenerating salivary axons. On rare occasions, this autonomic dyskinesia is congenital. No previous report about the association of bilateral crocodile tears and lateral thoracic meningocele with COAS has been described.

To our knowledge, there are only 38 cases with Wildervanck's syndrome in literature. This patient is the 39th case with bilateral crocodile tears, lateral thoracic meningocele, and Dandy-Walker syndrome in addition to COAS. The existence of these anomalies in our case may be coincidental as it has not been reported previously, or may be an extended part of the syndrome.

**REFERENCES**