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Simultaneous Presentation of Duane Retraction Syndrome and Coats' Disease: A Case Report

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ABSTRACT

Introduction: Duane retraction syndrome and Coats' disease are two relatively rare ocular conditions that occur in congenital and acquired forms in children. We present a 12-year-old boy with the chief complaint of eye deviation who was diagnosed later on to have Duane retraction syndrome in one eye and Coats' disease in the other. After a comprehensive review of literature, we assume that this is the first case of simultaneous presentation of these two disorders ever to be published. However, we do believe that these are two separated entities and their simultaneous presentation in this patient is pure coincident.

Case: A twelve-year-old Asian male was presented with about 25 prism diopters of exotropia in primary position, limited abduction/adduction, and narrowing of palpebral fissure of the right eye since childhood. The left eye showed lipid deposition, macular edema, and peripheral retinal telangiectasia.

Conclusion: The occurrence of two different congenital and acquired ocular diseases is rare. This is the first simultaneous presentation of Duane syndrome and Coats' disease ever to be reported in a young patient.

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Introduction

Duane syndrome is a rather rare ocular motility disorder (1%-5% of patients with strabismus) with abnormal muscle innervations and impaired horizontal eye movements (co-contraction of medial and lateral rectus muscles), globe retraction, palpebral fissure narrowing, and esotropia or exotropia in primary position. This syndrome has been reported in association with several others. Most cases are sporadic. Females are the main victims, and left eye involvement is predominant (1).

Coats' disease is a condition with retinal vascular dilatation (retinal telangiectasia) most often associated with retinal detachment and retinal capillary nonperfusion. This disease is best diagnosed by angiography. Serum and blood components can leak from these abnormal vessels and accumulate under the retina. This condition is not hereditary and usually affects just one eye. Most patients are male (85%) (2).

It is believed that the process which ultimately results in Duane retraction syndrome occurs during the 4th to 10th week of embryogenesis (1). That is why accompanying congenital malformations are 10 to 20 times more prevalent than the general population; malformations of the skeletal, auricular, ocular, and neural systems are the most common associated abnormalities (3). The syndrome has some associations with different disorders and genetic abnormalities, for instance Goldenhar or Wildervanck syndrome and chromosomal anomalies such as 12q12 deletion (4).

Some retinal abnormalities have been seen in cases with Duane retraction syndrome, for instance optic nerve hypoplasia, morning glory disc anomaly, and retinitis pigmentosa (5,6,7). As mentioned before, the syndrome is sporadic in most cases but can also show autosomal dominant inheritance.

The following associations have been reported in autosomal dominant cases of the syndrome: mutations in the CHN1 gene or abnormalities in sex chromosomes (8,9). Coats' disease is an acquired, isolated disorder which is most frequently seen in young males.

This disorder presents with peripheral retinal telangiectasia accompanied with massive macular exudation (10).

Differential diagnoses of coats' syndrome are retinoblastoma, persistent hyperplastic primary vitreous and retinal dysplasia (2).

Case

The reported patient was a 12-year-old male who referred to Hospital of Mashhad University of Medical

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Sciences with the chief complaint of right eye deviation. The problem had been noticed from early childhood. No history of previous ocular surgery was found. The family history was negative for strabismus.

The corrected visual acuity was 20/20 in the right eye and 20/400 in the left eye. The left eye visual acuity was told to be this low since early childhood. The alternate prism and cover test with accommodative target revealed a 25 Prism Diopter (PD) exotropia in the primary position.

The ocular movement was limited in abduction and adduction along with the narrowing of palpebral fissure and retraction of the globe in attempted adduction of right eye (Figure 1).

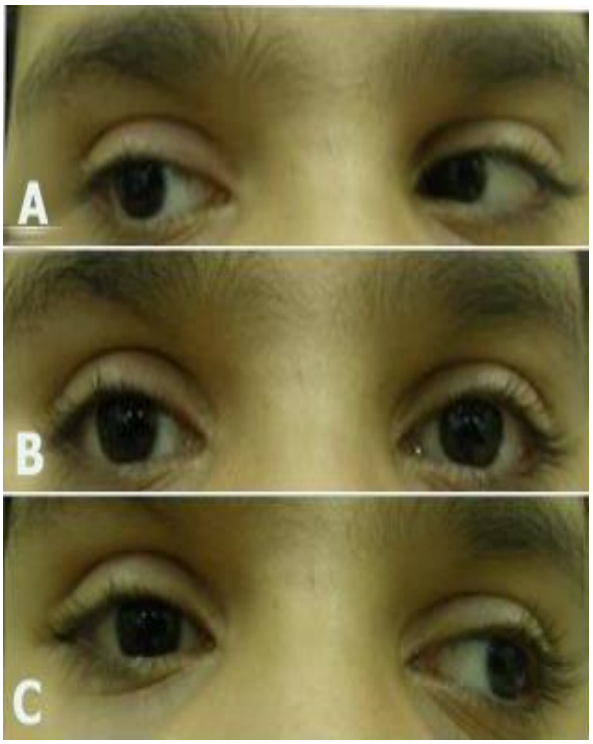

Figure 1: A) This picture shows the abduction deficiency of the right eye in lateral gaze.
B) The primary position of gaze which shows the exotropia of the right eye.
C) The adduction deficiency of the right eye which is accompanied with great palpebral fissure narrowing and globe retraction.

The slit lamp biomicroscopic evaluation was within normal limits in the anterior segment. Intraocular pressure was 15 mmHg in both eyes.

Retinal examination was within normal limits in the right eye, but the left eye retina showed lipid deposition within posterior pole and macular region.

Vessels in the temporal region of the left eye retina showed typical vascular telangiectasia, which is characteristic of Coats' disease (figure 2).

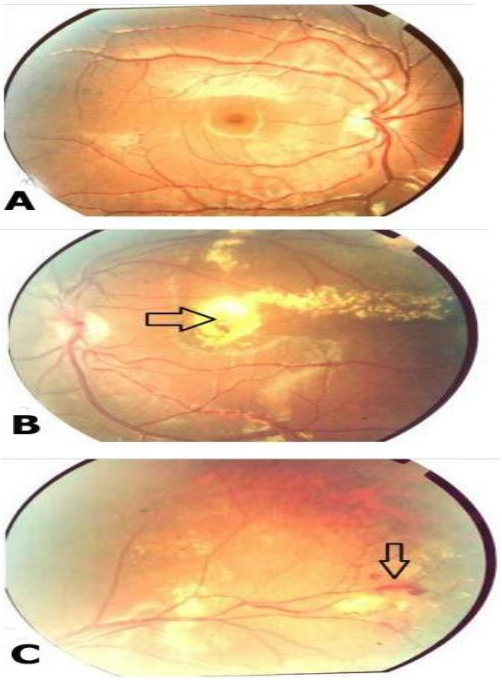

Figure2: A) Fundus photo of the right eye's retina which is a normal fundus.
B) Fundus photo of the left eye which shows great hard exudates within the posterior pole (horizontal arrow).
C) Temporal fundus region of the left eye with typical telangiectasia which is characteristic of Coats' disease (vertical arrow).

The patient was diagnosed with Duane retraction syndrome in the right eye and the Coats' disease in the left eye.

Discussion and Conclusion

After literature (Pub Med, Med line) using "Duane retraction syndrome" and "Coats' disease", as keywords we did not find any associations between these two entities. The negative family history of our patient makes the possibility of genetic disorders very unlikely.

As far as the literature review showed, our case is the first patient with concomitant occurrence of Duane syndrome and Coats' disease in different eyes. Same patient: an original case report of interest to Strabismus and retina entity. Written informed consent was obtained from the patient's legal guardians for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal. Authors declare that they have no competing interest.

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