Primary Hiperplastic Persistant Vitreus

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ABSTRACT

Persistent hyperplastic primary vitreous (PHPV) is a disorder of ocular development which occurs due to incomplete regression of the embryonic vitreous and hyaloid vasculature. Most cases of PHPV are sporadic and unilateral. We report a case of PHPV in a 5-year-old male who presented with complaint of decreasing eye size. Grey-scale ultrasonography (US) evaluation revealed an hypoechogenic band in the posterior segment of the left globe extending from the posterior surface of the lens capsule to the optic disc. Also magnetic resonance imaging (MRI) findings suggested the diagnosis of persistent hyperplastic primary vitreous.

Key words: *Persistent hyperplastic primary vitreous, imaging, MRI, ultrasound*

Persistan hiperplastik primer vitreus

ÖZET

Persistan hiperplastik primer vitreus (PHPV) embriyonik vitreusun inkomplet regresyonuna bağlı olarak ortaya çıkan gelişimsel oküler bir hastalıktır. PHPV vakalarının birçoğu sporadiktir ve tek taraflı görülür. Bu yazıda gözde küçülme şikayeti ile başvuran 5 yaşındaki PHPV olgusu sunuldu. Ultrasonografi (USG) incelemesinde sol glob posterior segmentinde, lens posteriorundan optik diske uzanan hipoekoik bant görüldü. Aynı zamanda magnetik rezonans görüntüleme (MRG) bulguları da PHPV tanısını destekledi.

Anahtar kelimeler: *Persistan hiperplastik primer vitreus, görüntüleme, MRG, ultrasonografi*

INTRODUCTION

Persistent hyperplastic primary vitreous (PHPV) is a developmental anomaly of the eye which occurs due to incomplete regression of the embryonic vitreous and hyaloid vasculature (1). The disease is subclassified into three presentation; anterior, posterior and combination of anterior and posterior. The most commonly seen clinical presantation is the mix type (1). The most common presenting signs and symptoms are leukocoria, poor vision and small eye. PHPV is the second most common cause of leukokoria. Ophthalmoscopic and biomicroscopical examination may be sufficient in the diagnosis of PHPV. But especially in patients, that could not seen posterior segment of the eye, radiological examinations become important and suggest the diagnosis of disease. In addition, imaging modalities provide useful information on the separation from retinoblastoma (2). We report a case of PHPV with clinical and radiological findings.

CASE

5-year old male patient was admitted to eye clinic with complaints of decrease in eye size that noticed the last 2 weeks. At ultrasonography (US) examination; hypoechoic linear structure that was compatible with the hyaloid artery remnant was observed, was ranging from posterior to anterior (Figure 1a, b). Also vitreous contents on the left was more intensive than the right (Figure 1a, b). A linear band, seen hypointense in all

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sequences, extending from the posterior surface of the lens capsule to the optic disc on the left eye was also seen on magnetic resonance imaging (MRI) (Figure 2). The axial length of the left globe was 21 mm and the axial length of the right globe was 25 mm, which was suggestive of microphthalmia of left globe on orbital MRI (Figure 3). On T1-weighted MRI image, the content of left vitreus was homogeneously hyperintense and it was thought hemorrhagic or protein content of the liquid (Figure 3). After IV injection of gadolinium, no pathological enhancement was seen. The right glob was normal. The case was diagnosed as persistent hyperplastic primary vitreous with these findings.

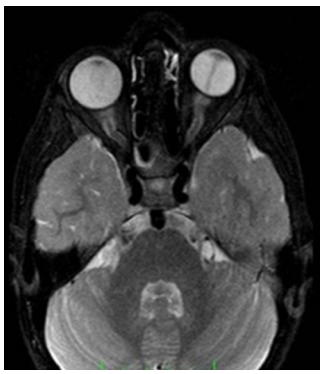

***Figure 2**. T2-weighted axial MRI image showed a linear hypointense band extending from the posterior surface of the lens capsule to the optic disc in the left eye.*

DISCUSSION

Persistent hyperplastic primary vitreous is a group of complex ocular malformations caused by the failure of regression of the primary vitreous (3). The primary vitreous is formed during the first month of development and contains branches of the hyaloid artery. This hyaloid artery begins to regress during formation of the avascular secondary vitreous at 9 weeks. By the third month, the secondary vitreous, which ultimately forms the adult established vitreous, fills most of the developing vitreous cavity. The primary vitreous becomes condensed into a narrow band (Cloquets canal), running from the optic disc to the posterior aspect of the lens (4).

The disorder is one of the most common congenital malformation syndromes of the eye and is usually unilateral, but a rare bilateral condition has also been described. Bilateral lesions are more associated with systemic or syndromic conditions. Most cases of PHPV are sporadic. However, it can be inherited as an autosomal dominant or recessive trait (2, 3). The disease primarily affects the premature infants. The most common presenting signs and symptoms are leukocoria, poor vision and microphthalmia. Gloucoma, cataracts, strabismus can also be seen.

PHPV shares many of its clinical features with other ocular pathologies, such as retinoblastoma, retinal dysplasia, conjenital cataract, retinopathy of prematurity, uveitis and Coat's disease. PHPV usually presents with

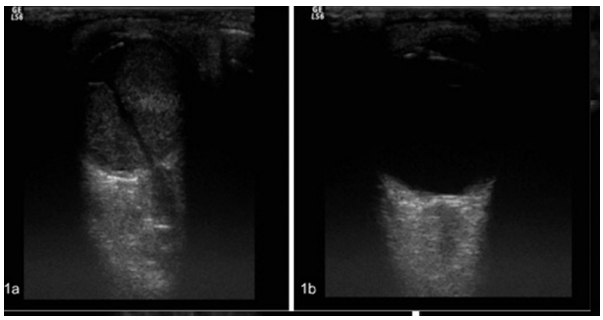

***Figure 1**. US image of left eye showed lineer hpyoecogenic band extending from posterior to anterior that was compatible with the hyaloid artery remnant (1a). US image of bilateral eye showed the more intensive content of left vitreus than the right (1a, b).*

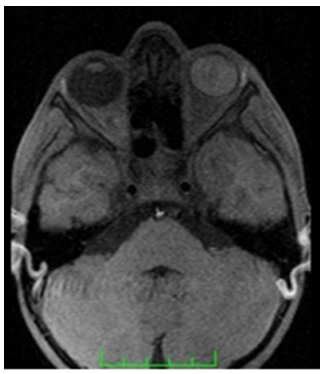

***Figure 3**. T1-weighted axial MRI image showed the microphthalmia and increased signal intensity (due to hemorrhage or protein content) on left eye.*

unilateral leukokoria. In all cases of leukokoria it is important to exclude retinoblastoma, which can be either unilateral or bilateral. Retinoblastoma is life threatening and requires aggressive treatment. Persistent hyperplastic primary vitreous has a typical imaging appearance which allows reliable differentiation from retinoblastoma (2).

In PHPV, grey-scale ultrasound shows an echogenic band extending from the posterior surface of the lens capsule to the optic disc. Colour Doppler may show flow within this band representing a persistent hyaloid artery (2, 5). Computed tomography (CT) findings of PHPV are absence of calcification, increased density of the entire vitreous, tubular intravitreal density (Cloquet's canal or nonattached retina), decubitus positioning showing a gravitational effect on fluid-fluid level, micro-ophthalmia, enhancement of abnormal intravitreal tissue, and small or irregular lens (6). MRI findings of PHPV consist of a tubular structure, representing the hyaloid vessel; a funnel-shaped retinal detachment, with the subretinal fluid hyperintense on both T1- and T2-weighted images; fluid-fluid level due to the presence of hemorrhage in the subretinal space; a retrolental mass; micro-ophthalmia, and vitreous hemorrhage (7).

In conclusion, especially in the diagnosis of PHPV radiological imaging methods offers useful information in the differatiation of the disorders that cause microphthalmia and leukocoria. PHPV can be differentiated from retinoblastoma by the absence of a calcified mass, artery running through Cloquet's canal, and typical signal characteristics of retinoblastoma on MRI. Also US and CT findings provide useful additional informations. Demonstration of tubular, triangular or round-shaped abnormal hyaloid tissue, that extending from posterior lens to the retina, is the most important radiologic finding.

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