Vitreous Cyst Combined With Bilateral Juvenile Retinoschisis

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ABSTRACT
Vitreous cyst, an uncommon finding, was first described by Tansley in 1899.1 Since then only a few reports dealing with congenital or acquired vitreous cyst have been reported. We report the case of a 16-year-old girl with a unilateral developmental vitreous cyst combined with bilateral retinoschisis. The pathogenesis and histopathology of these phenomena remain unclear, although a few assumptions about the common pathogenesis are discussed. According to our knowledge, this is the first description of vitreous cyst and retinoschisis in the same eye.

CASE REPORT
A 16-year-old girl was sent for a routine eye examination. Visual acuity was 6/6 and the anterior segment was normal in both eyes. On indirect ophthalmoscopy, no pathology was observed in the right vitreous; in the left vitreous a small, round, pearly cyst with spreading pigmentation was seen (Figure 1). The cyst was mobile without any strand connection to other structures of the eye, as can be seen in the B-scan ultrasonography (Figure 2). In the lower nasal periphery of both eyes, a shallow, wide retinoschisis with marked borders was found reaching the equator. Computerized visual field examination revealed bilateral upper temporal scotoma.

DISCUSSION
Most vitreous abnormalities are mesodermal in origin, resulting from the failure of the primary vitreous or its vessels to regress. The anomalies vary from tiny threadlike structures floating in the retrolental space to its almost complete replacement by fibrous tissue. Normally the central hyaloid artery starts regressing in the third month of gestation and becomes completely atrophic in the eighth month.2-3 With a swift vitreous movement and a slow movement back, Cloquet's canal and the distribution and interrelations of intravitreal opacities become more visible.4 Vitreous cysts are either developmental or acquired. Developmental vitreous cysts arise either from the hyaloid artery or Bergmeister's papilla5-7 and are not hereditary. Cysts arising from remnants of the hyaloid vessels often are attached to the optic disc. Those originating from Bergmeister's papilla, on the other hand, are usually small, round, and pearly.8 The developmental cysts usually are located in the axial region of the vitreous cavity. They are translucent, which permits viewing of the retinal details, mobile, gray, and sometimes spotted with pigment. The vitreous cavity is clear. There are no signs of vitreous inflammation or trauma.7

Developmental cysts may produce floaters and transient blurring of vision9-10 which may disappear when the cyst becomes displaced from the visual axis. Yet the origin of larger cysts, found in our patient, is obscure. Various authors claim that such cysts arise from ciliary processes and later detach and float into the vitreous.11,12 Others suggest that these are really adenomatous cysts or even an evacuated vitreous exudate.1

Because diagnosis in our patient was made 5 years ago, no change was noticed in the cyst's size, color, and consistency. No history or evidence of trauma, inflammation, or any other cause of “acquired cyst” was noticed. This confirms the diagnosis of a developmental vitreous cyst.

Juvenile retinoschisis is an X-linked recessive retinal disease usually found in males but which also may affect females.7 The disease tends to become stationary after the age of 20. Histopathologically, degenerative changes in the nerve fiber layer are shown usually without vitreo-retinal participation. Persistent hyperplastic primary vitreous or other vitreo-tapetal degeneration should be considered in

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The authors wish to thank Dr. Eduard Svetlana for his photographic assistance.
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the differential diagnosis. In 68% of juvenile retinoschisis there is macular involvement with declining visual acuity. An intimate relationship between vitreous abnormalities and retinoschisis was described by Tolentino⁶ and Morse.⁶

In retinoschisis Cloquet’s canal appears larger than normal, or congenital veins may be found in the vitreous. The combination of two rare phenomena in the same eye leads us to think of the possible linkage between them. Randall postulated that the vitreous cyst formation derives from the fetal cleft, and is freed into the vitreous.¹⁵ The fetal cleft disorders and delay in growth of the primary and secondary vitreous in this site are causes of congenital retinoschisis as well.¹⁴ Thus, a possible relation between both abnormal developments may be seen. The most common vitreous change in congenital retinoschisis is liquefaction of the vitreous above the schisis area.¹⁵ This liquefied space is surrounded by membranes and semi-liquid gel that tent to organize as a vitreous cyst. Vitreous cyst formation is one of the vitreal reactions to underlying retinal degeneration.¹⁶ We emphasize once again that the pathogenesis of vitreous cyst formation and its relation to retinoschisis is questionable and no histopathologic proof has been found. The combination of vitreous cyst with bilateral juvenile retinoschisis with sparing of the macula and visual acuity of 6/6 in a female is rare.

FIGURE 1: A fundus photograph showing the vitreous cyst.

FIGURE 2: The vitreous cyst echoes as seen in a B-scan ultrasonography.

REFERENCES