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Persistent Hyperplastic Primary Vitreous

— A report of a case associated with microphthalmia —

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Persistent hyperplastic primary vitreous (PHPV) is the term to designate "persistent posterior fetal fibrovascular sheath of the lens" (Reese, 1955). The posterior fibrovascular sheath usually shows complete regression at about the seventh fetal month, but remains of this structure are seen at birth, particularly in premature infants. Since the primary vitreous has two elements, the fibrovascular sheath and the hyaloid artery, one or both of these can be remained hyperplastic in the affected eyeball, which is called PHPV.

This is a case of persistent hyperplastic primary vitreous associated with intra-ocular adipose tissue and microphthalmia.

REPORT OF A CASE

This 10 months old boy was admitted to Seoul National University Hospital for the operation of a small eye in the right side on June 20, 1983. He was a product of normal full term spontaneous delivery to a 30 year old multigravid mother who took some unidentified drug for the emesis gravidarum during early phase of the pregnancy. The birth weight was 3.7 kg, and he did well pre- and post-natally except for the smallness of the rihgt eye.

Physical examination showed no remarkable abnormalities other than the right eyeball. The right orbit was small and the cornea measured 5 mm in diameter. The left eyeball was normal. Rest of the examinations including routine laboratory data and chest X-ray were unremarkable. Enucleation of the right eyeball was done for the sphere insertion.

Pathological examination (S83-7027):

Submitted eyeball in formalin measures 13 mm (AP diameter), 10 mm (height) and 10 mm (width) with a 4 mm stump of the optic nerve that has 4 mm in diameter. The cornea measures 6.0×5.0 mm in vertical and horizontal diameter. The sclera is white and thick. On sagittal sections there is no structure identified to be lens, and the vitreous cavity is filled with blackish soft tissue with central round grayish white area.

Microscopically, the cornea is small but no structural anomaly is present. The anterior chamber is abnormally shallow. The lens is shrunken to a small globule and is the site of multiple calcifications. The lens is degenerated and is subluxated forward and surrounded by fibrovascular tissue. The posterior lens capsule is not identifiable. The ciliary processes are elongated and are incorporated in the periphery of the fibrovascular tissue. The iris and ciliary process are matted together, and there is a heavy punctate melanin pigmentation along with fibrosis. A small focus of cholesterol clefts is noted in this area where

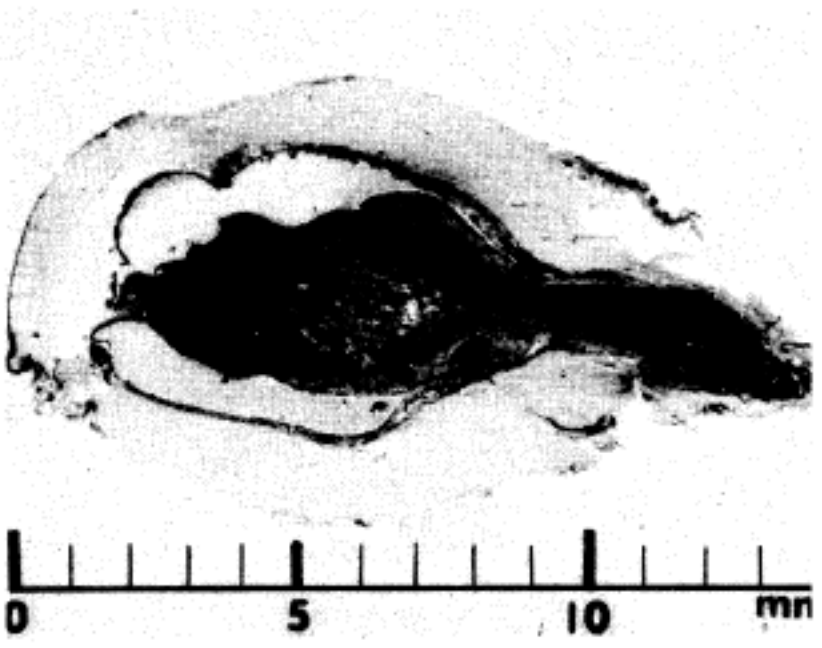

Fig. 1. Photomacrograph of the specimen showing microphthalmia and persistent hyperplastic primary vitreous.

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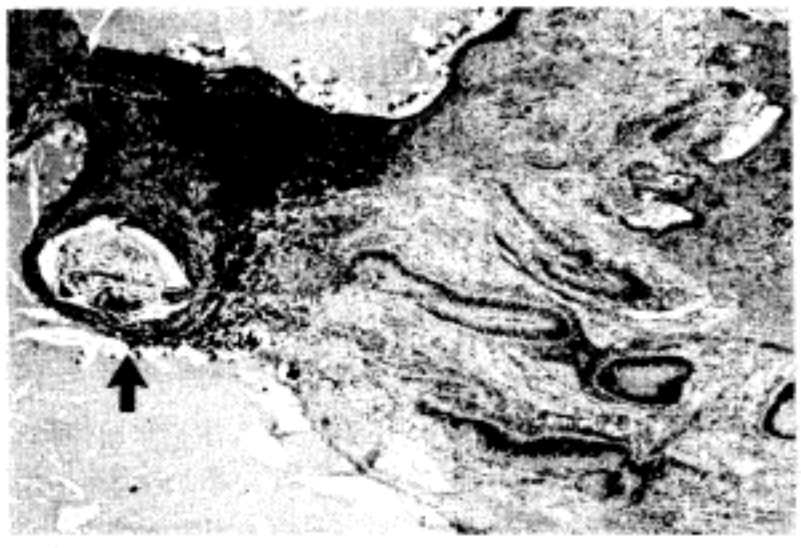

Fig. 2. Photomicrograph showing a focus of cholesterol clefts(arrow) and dysplastic retinal tissue around the fibrous core. (H&E, ×40)

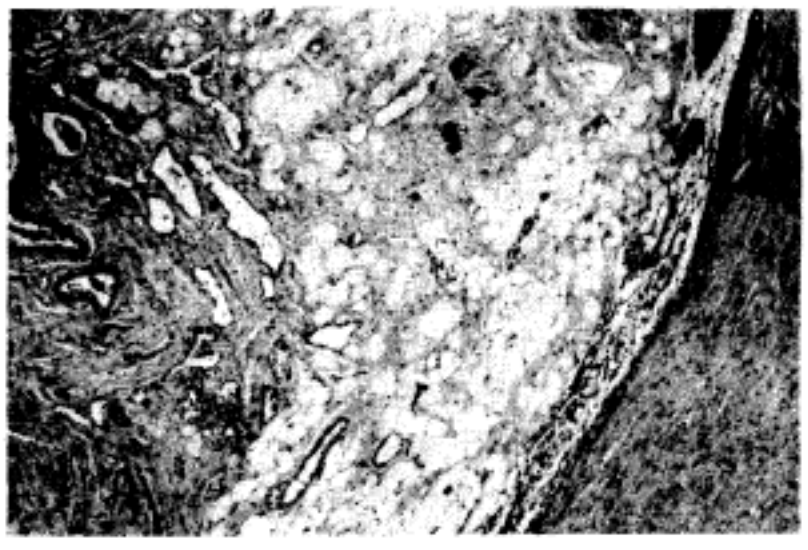

Fig. 3. Persistent hyaloid vessels and mature adipose tissue. (H&E, ×100)

dysplastic retinal tissue is adherent with it. Undifferentiated neuropithelial element is seen is this area. Radial and meridional smooth muscle fibers in the ciliary body are seen. But no Schlemm canal is seen. The vitreous body is reduced in volume and the surrounding sclera is fairly thick. There is a fibrovascular core in the vitreous cavity, where numerous thick-walled vessels and small amount of mature fat are included. This fibrovascular core starts from posterior margin of the lens and extends to the optic disc. It is surrounded by totally detached proliferative retinal tissue. The hyaloid artery is seen to be patent and to have smooth muscle wall. They extend into and along the fibrovascular sheath. Thee is eosinophilic amorphous exudate in the remaining vitreous cavity.

DISCUSSION

This case appears to be a typical example of persistent hyperplastic primary vitreous (PHPV). Unilateral involvement of the eye, association with microphthalmia, cataractous lens and persistent hyaloid vessels are in favor of PHPV and exclude the possibility of retrolental fibroplasia. The presence of long ciliary processes and shallow anterior chamber are not seen in congenital cataract which should be differentiated in this case.

Mature adipose tissue seen in this case is commonly observed in association with PHPV and microphthalmia. It can be seen in the vitreous body, the lens or retrolental masses. Font et al (1969) interprete the presence of intraocular far to represent a histologic variant of PHPV in which metaplastic changes have occurred either in intrauterine life or later after birth. Whether the fat was present congenitally or it developed after birth as a result of metaplasia from prexisting mesenchymal tissue is not known.

Although hyaloid vessels are always present in PHPV this case appears to represent an unusual accentuation of the hyaloid component. Accentuated hyaloid system is often associated with retinal detachment of varying degrees as in our case (Reese, 1955).

The presence of cholesterol clefts with foreign body reaction seen in this case represents old hemorrhage. It is often seen in PHPV (Reese, 1955). There are people who believe that hemorrhage plays an important role in producing PHPV (Lane, 1919).

SUMMARY

A case of persistent hyperplastic primary vitreous (PHPV) is reported in a 10 months old boy who had a congenitally small right eyeball. PHPV is associated in this case with cataractous lens, malformed anterior chamber, elongated ciliary processes, islands of mature adipose tissue, total retinal detachment and accentuated hyaloid vessels and microphthalmia.

As far as we could determine this case appears to be

isolated anomaly without associated malformation syndrome.

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=국문초록=

소안구증을 수반한 Persistent Hyperplastic Primary Vitreous 1예

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지제근 · 박인애

10개월 남아의 오른쪽 소안구를 검색한 결과 일차성 Vitreous의 잔종증이라고 생각되는 소견이 나타났는데 안구는 13×10×10mm였고, 각막은 6×5mm였다. 현미경적으로 렌즈의 석회화와 편위, 전방의 기형과 더불어 전반적 망막박리가 있었다. 한편 Vitreous 내에는 렌즈뒤로부터 시신경 출구에 이르는 hyaloid 혈관과 함께 이형성 망막조직의 증식이 있었고, 이와 더불어 성숙 지방 조직도 그속에 포함되어 있었다.