Choroidal osteoma in a preterm infant

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Choroidal osteoma (CO) is a rare, benign, and usually unilateral intraocular tumor composed of mature bone affecting the choroid. It appears as slightly elevated, yellowish-white, or orange choroidal mass with well-defined borders. It is commonly encountered in young female adults. Here, we report findings of a 4-week-old premature baby with CO in her left eye which was detected during a routine examination for "retinopathy of prematurity." We believe that this case is the youngest patient reported with CO which showing that this pathology can be encountered even in a newborn.

Key words: Choroidal osteoma, cystoid macular edema, optical coherence tomography, prematurity

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Choroidal osteoma (CO) is a rare, benign, and usually unilateral intraocular tumor composed of mature bone affecting the choroid. It appears as slightly elevated, yellowish-white, or orange choroidal mass with well-defined borders. Although benign in nature, it usually grows very slowly over months to

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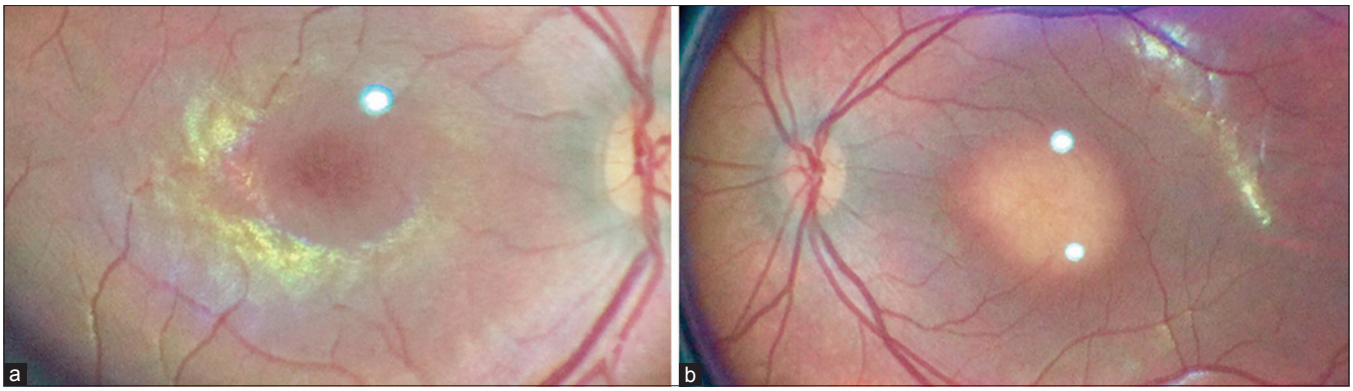

Figure 1: Fundus appearance of the right (a) and left (b) eyes of the case. Cystoid macular edema is seen in both eyes but more prominent in the right eye. In the left eye, a well-demarcated yellow-orange colored macular lesion in is seen

years. It is commonly encountered in young female adults.[1-4] Here, we report a 4-week-old premature baby with CO in her left eye which was detected during routine "retinopathy of prematurity" (ROP) examination.

Case Report

A 4-week-old girl baby born at 34th gestational week was examined for ROP. On examination, the baby was fixing to the light with her right eye and could not fix with her left eye. The external structures and anterior segment were normal in both eyes. Fundus examination showed irregular foveal reflex in both eyes. In the left eye, we also detected a well-demarcated yellow-orange mass in the center of the macula matching CO appearance. It was slightly elevating the fovea and was measuring about three-disc diameters. No ROP sign was detected in both eyes [Fig. 1]. Enhanced depth imaging optical coherence tomography (EDI-OCT) showed cystoid macular edema (CME) in eyes and a well-demarcated subretinal, hyporeflective, and homogeneous choroidal mass in the left eye [Fig. 2]. It was elevating the fovea about 250 μm from the retinal surface. The baby had no other systemic disease, and there was no history of trauma. We followed the baby without any treatment. On control examination at the 36th day, the fovea had a normal reflex in both eyes. OCT showed that CME had resolved spontaneously in both eyes [Fig. 3]. Besides this, osteoma had the same appearance on fundus examination and OCT.

Discussion

CO is an ossified tumor first described in 1978 by Gass and associates.[1] It is a rare, benign intraocular tumor affecting the choroid and unilateral in about 80% of the cases. The exact etiology of the CO is unknown.[1-4] Histopathology illustrates dense bony trabeculae with marrow spaces traversed by pathognomonic dilated thin-walled blood vessels.[2,5]

The diagnosis of the CO is mainly clinical. A yellow-orange well-demarcated lesion on fundus examination is typical.[1-4] The COs have high acoustic reflectivity due to the presence of calcified components in ultrasound. A characteristic shadowing or sound attenuation can be seen posterior to the lesion.[1-4] OCT, computed tomography, magnetic resonance imaging, and fluorescein angiography are other ancillary diagnostic tests for diagnoses.[1-4] The diagnosis of our case was based on the typical fundus appearance and OCT findings. We use a

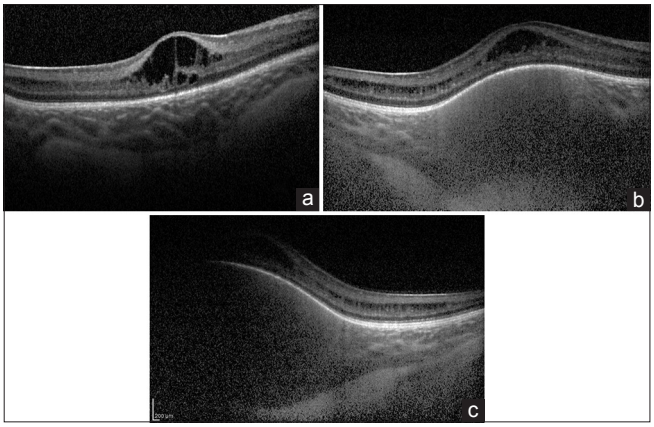

Figure 2: Optical coherence tomography images of the right (a) and left (b and c) eyes of the case at first examination. Optical coherence tomography shows cystoid macular edema in both eyes. In the left eye foveal elevation, separation at outer nuclear layer and choroidal hyporeflective mass is also seen

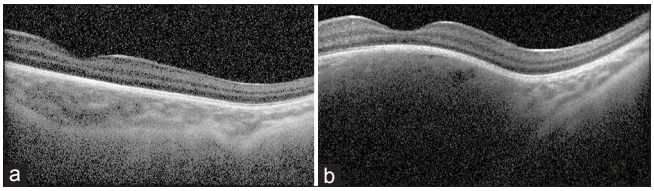

Figure 3: Optical coherence tomography images of the right (a) and left (b) eyes taken at 36 days after the first examination. Cystoid macular edema is resolved in both eyes. In left eye, outer nuclear layer separation is disappeared. Foveal elevation and subfoveal hyporeflective well-demarcated mass can be seen in the left eye

Heidelberg Engineering Spectralis HRA-OCT. Our biomedical unit made some mechanical modifications to lean forward the OCT device to take OCT images of a lying baby. During this performance, at least two doctors were needed. One of them controlled the position of the head of the baby and the other one controlled the OCT device and took the OCT images. We also added some information about this performance in the text (at the end of the first paragraph in discussion part).

EDI-OCT provides a more detailed view than spectral-domain OCT of deep anatomic structures such as choroid.[6] Shields *et al*.

reported that EDI-OCT of CO reveals unique features including subtle horizontal hyperreflective lamellar lines (bone lamella) with occasional denser lines, speckled-spongy, tissue, and horizontally or vertically oriented tubular channels representing Haversian or Volkman canals or cavernous vascular spaces which correlate with mature bone histology.[7] Besides this, in OCT of our case, we saw a well-demarcated relatively hyporeflective homogeneous choroidal mass which had slightly elevated the fovea, pushed back the sclera and surrounded by normal spongious choroidal tissue. The reason that we did not see the OCT findings reported by Shields *et al*. may be related with the very early age of our case. Possibly, similar findings would be able to see in further ages of the patient.

The CO is encountered predominantly in young adults in their early twenties.[1-4] Many CO cases have been reported in different ages ranging from few months old to sixties in literature.[3] Kida *et al*. reported an 8-month-old girl infant with CO in her both eyes in 1997 as the youngest case. They had followed up this case for 8 years, and during this period, the fundus had become yellow-white and irregularly scalloped. At the age of 8 years, her visual acuity was 20/15 in both eyes.[3] We think that our patient is the youngest case reported as CO until now.

It is reported that CO may keep growing in about 50% of the cases followed for 10 years.[8] The increase in mean basal diameter of around is about 0.37 mm/year. In this process, the visual acuity may decrease due to choroidal neovascularization (CNV), atrophy of the retinal pigment epithelium, and photoreceptor overlying a decalcified osteoma and serous retinal detachment over the osteoma in about 60% of the eyes.[8,9] CNV is seen in about 31%–47% of the CO cases.[8] Overlying hemorrhage and irregular surface are reported as risk factors for developing CNV. The visual prognosis is influenced by the tumor location, decalcification status, overlying RPE atrophy, presence of CNV, persistence of subretinal fluid, and occurrence of subretinal hemorrhages.[8]

CME in our case was due to the prematurity. It is reported that CME can be seen in premature babies with ROP or without ROP as in our case. Spontaneous regression of CME has been reported in many studies.[10] In our case, CME has resolved spontaneously at about 36 days after the first examination.

Conclusion

We believe that this case is the youngest patient reported with CO. Our case shows that although CO is seen muchly in patients at twenties, it can also be encountered in newborns too. Therefore, ophthalmologic examination of newborns is very important for early diagnosis and early treatment of complications, especially the CNV, retinal detachment, and amblyopia.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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