Case Report
Persistence of Retinopathy of Prematurity in an Infant with Tetralogy of Fallot

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We report an infant with tetralogy of fallot (TOF) who was born at 35 weeks of gestation and of 1700 g birth weight and presented with persistent retinopathy of prematurity (ROP) at 6 months of age. Follow-up ophthalmic examinations were done at 2, 3, and 4 weeks of age. A demarcation line in Zone II was noticed on the first ocular examination done at 4 weeks of postnatal age. At 6 months of postnatal age, the infant still had an avascular peripheral retina with the demarcation line in Zone II. Even though this index subject did not have any typical risk factors for ROP, TOF seems to be the probable reason for developing as well as persistence of avascular retina.

1. Introduction
Tetralogy of fallot (TOF) is one of the most common cyanotic congenital heart diseases (CCHD). The prominent feature of the disease is cyanosis which depends on the disease severity [1].

It has been reported that presence of CCHD was related to the development of retinopathy of prematurity (ROP). It was indicated that cyanotic premature infants should undergo screening for ROP as other premature infants [2]. Paulus and Moshfeghi [3] demonstrated persistence of plus disease even after adequate laser therapy for ROP in a patient with TOF. Furthermore, studies have shown several retinal vascular abnormalities in association with TOF [4–6]. In the present report, we demonstrated a 6-month-old preterm infant with TOF who showed persistence of ROP with failed peripheral retinal vascularity.

2. Case Report
A female infant with gestational age (GA) of 35 weeks and birth weight (BW) of 1700 g was referred by a pediatrician on postnatal first month for routine examination for ROP to Ophthalmology Department of Zeynep Kamil Maternity and Children's Diseases Training and Research Hospital. The patient already was diagnosed to have TOF at Pediatric Cardiology Department at the same hospital. She also had a history of vaginal delivery after maternal induction of labor for oligohydramnios and intrauterine growth restriction (IUGR). No other significant risk factor was present before referral. At first visit, on binocular indirect ophthalmoscopic examination, a demarcation line in Zone II without plus disease was noted. ROP involved the 6 clock hours of the peripheral retina. No other abnormality was detected in anterior segment examination revealed by a hand-held slit lamp. Two weeks later, patient still demonstrated a demarcation line in Zone II. Consecutive examinations were performed with two- or three-week intervals up to a postmenstrual age (PMA) of 54 weeks in order to detect a possible progression of the retinal findings. The last examination at adjusted 6 months of age revealed persistence of the findings with demarcation line in Zone II and vascular dilatation and tortuosity (Figure 1). No additional abnormal finding was noted in both anterior and posterior segments of the eye. Fixation behavior was
central and steady and was maintained in both eyes. No
evidence of gaze palsy, strabismus, higher refractive error, and
 glaucoma was noted. Furthermore, clinical observation was
still considered for TOF by the Pediatric Cardiologist due to
the stable clinical course of the disease at the time of the last
ophthalmologic examination.

3. Discussion

Tetralogy of fallot is a common form of CCHD. Although
most of the patients have normal life with good function after
surgical intervention, regular follow-up is crucial following
surgery [7]. Several retinal abnormalities have been docu-
mented in patients with TOF. These include increased retinal
vascular tortuosity, [8] retinal arterial and vein occlusions,
[5, 9], and ischemic [6] and proliferative retinopathies [10].

The infant in our case had a GA of 35 weeks and BW of
1 700 g. Studies from several parts of the world have reported
that larger premature infants who received improper oxygen
therapy can develop ROP [11, 12]. Our case has already been
diagnosed with TOF at birth and required supplemental
oxygen administration due to systemic hypoxia. The devel-
optment of ROP might be related to this issue in our case.
Supportively, Johns et al. [2] have previously reported that
the presence of TOF has an association with the development
of ROP. Furthermore, it has been well known that peripheral
retinal avascularity is also not uncommon for ROP. But it has
been shown that this situation occurs commonly in smaller
premature infants with lower BW. It has been stated that
spontaneous involution of ROP follows a systematic pattern
wherein anteroposterior location of retinopathy changes
from Zone I to Zone II or Zone II to Zone III [13]. In our
case, we did not observe any sign of progression or regression
pattern. Demarcation line freeze at anterior Zone II location
until 6 months of age.

A reason for the development of retinal vascular abnor-
malities in TOF has been suggested to be ischemia. In
general, oxygen saturation of the arterial blood cannot be
maintained in infants with a CCHD; therefore an ischemic
environment of the body arises [11]. The role of hypoxia
has been demonstrated to be associated with nonperfused
peripheral retina in animal models [14]. Avascular retina
in our case was due to ROP. However, we can hypothesize
that the infant does not have very significant risk factors for
ROP and still has developed ROP. Hence, the deoxygenated
blood in TOF might be responsible for altered production
of factors responsible for normal retinal vascularisation and
hence might have contributed to causing ROP in this case.

In another study, Paulus and Moshfeghi [3] observed
persistence of plus disease in an infant with TOF after
appropriate laser treatment for ROP. They indicated that
vascular tortuosity following laser therapy in that case was
independent of ROP regression. We also observed minimal
vascular tortuosity in our case during the follow-up period.

In conclusion, we reported a larger premature infant
with persistence of ROP at 6 months of age which might be

Figure 1: Minimal vascular tortuosity is observed in both eyes (a and b). The arrows indicate the demarcation line in Zone II along the tem-
poral region in both eyes (c and d).
associated with TOF. However, further reports are needed to better clarify the underlying mechanism of the associated retinal findings in infants with TOF.

Competing Interests
The authors declare that they have no conflict of interests.

References


