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CASE REPORT

Response of Intravitreal Bevacizumab in Choroidal Neovascularization Associated with Choroidal Osteoma

Muhammad Moniruzzaman[1] Ankur Gupta[1] P Tandava Krishnan[1]
Siddiqur Rahman[2] Ummay Kawser[3] Emranul Islam[4]

Abstract

Our aim of this case report to show the effect of intravitreal bevacizumab in the treatment of choroidal neovascularization secondary to choroidal osteoma. A lady of 21 yr old presented with best corrected Snellen's visual acuity (BCVA) 6/18 in right eye and 6/6 in left eye. On clinical examination, a choroidal mass was noted in the posterior pole and parapapillary area of right eye. Ultrasonography showed a high intensity echo spike from the inner surface of the tumor with posterior shadowing. CT scan revealed features of a radiopaque plaque of bone density at the level of the ocular coat suggestive of choroidal osteoma. FFA and OCT showed features suggestive of choroidal neovasularisation within the mass. Three injections of intravitreal Bevacizumab were given at monthly intervals, following which BCVA improved to 6/6 and membrane showed signs of resolution both clinically and on FFA & OCT. 2 months after the third injection, recurrence of CNVM was noted on OCT and a repeat intravitreal bevacizumab was given . One month after the fourth injection , patient had a BCVA of 6/6. So in our experience intravitreal Bevacizumab is useful in the management of CNVM secondary to choroidal osteoma .

Introduction

The term 'choroidal osteoma'was coined by Gass in 1978.[1] These tumors demonstrate evidence of bone formation in the choroid & are believed to be choristomatous in origin.[2] In a follow-up study of 36 patients, the probability of loss of visual acuity (20/200 or worse) was more than 50% over 10 years.[3] Choroidal neovascularization (CNVM) is the most frequent cause of visual loss (>50%) in choroidal osteoma.[4]

Patint and methods

We report a case of a 21 year old lady of choroidal osteoma with CNVM presenting with complaints of defective vision OD for past 6 months. Her past ocular, medical, and family histories were noncontributory. On examination, her best-corrected visual acuity was 6/18 N8 in OD and 6/6 N6 in OS. Her anterior segment evaluation was within normal limits in OU. Fundus examination OD showed an elevated, yellowish lesion on the posterior pole having a smooth surface with slight elevation with associated subretinal haemorrhage (Figure:1). B scan ultrasound showed high surface reflectivity with posterior shadowing suggestive of ossification (Figure:2). CT scan showed a radio opaque plaque of bone density at the level of posterior pole in OD (Figure:3). FFA showed a

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large area of early patchy hyperfluorescence with late staining in the area of lesion with an area of blocked fluorescence in area of sub retinal haemorrhage to it suggestive of a CNVM (Figure:4). Spectral domain OCT showed a high reflective sub retinal lesion with posterior shadowing with adjacent area of sub foveal sub retinal fluid (Figure:5) . A diagnosis of choroidal osteoma with secondary CNVM was made. A treatment option of anti VEGF (Vascular Endothelial Growth Factor) injection was offered to the patient after explaining its roles and limitations for which the patient consented.The patient was given 3 intravitreal bevacizumab injections (1.25 mg in 0.05 mL) at monthly intervals.

Reaults

Visual acuity improved to 6/6; N 6 & CNVM showed signs of regression, with resolution of sub retinal hemorrhage and fluid confirmed on OCT (Figure:6a) and FFA. 2 months after third injection, recurrence of CNVM was noticed on OCT (Figure:6b) and a repeat bevacizumab injection was given. Two months after the last injection (7 months), CNVM was inactive with a visual acuity of 6/6; N 6 which was also noted on OCT (Figure:6c) and FFA.

Discussion

LASER photocoagulation was initially used for Choroidal osteoma with extrafoveal secondary CNVM.[5] Aylward et al found the success rate in such cases to be only 25%.[2] Nearly half of Choroidal osteoma related CNVM were subfoveal, which could only be observed. Extrafoveal CNVM too was poorly responsive to laser photocoagulation, probably due to depigmentation of RPE and degenerated RPE/Bruch's membrane complex, resulting in the reduced laser uptake. Surgical removal of subfoveal CNV has been performed successfully; however, the visual result has been poor.[6] Reports of PDT for osteoma-related CNVM have been published. However, issues related to cost, risk of vision loss and need for multiple retreatments have been raised.[7] TTT has also been tried in this condition but there is lack of visual recovery.[8] Bevacizumab has been used successfully for the treatment of CNVM due to neovascular AMD.[9] We used intravitreal bevacizumab for the treatment of Choroidal osteoma related CNV and observed rapid regression of CNV with a marked improvement of visual acuity. Marked improvement of visual acuity and rapid regression of CNVM was observed in our patient following 3 intravitreal injections of bevacizumab. However, recurrence was seen two months later which was treated with one more injection after which CNVM got stabilised and patient is maintaining a visual acuity of 6/6. Narayanan R et al reported prompt visual recovery and rapid CNVM regression following bevacizumab injection for Choroidal osteoma related CNVM.[10] Similar finding were noticed by Ahmadieh H et al.[11] This treatment modality may be superior to previous methods in terms of anatomic and visual results. This dramatic effect may be attributable to the very thinned and degenerated RPE/Bruch's membrane complex, resulting in the better penetration of bevacizumab into the underlying subretinal space.

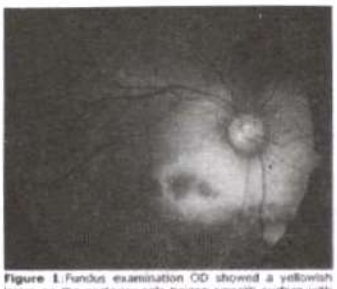

Figure 1: Fundus examination OD showed a yellowish lesion on the posterior pole having smooth surface with slight elevation associated with subretinal haemorrhage

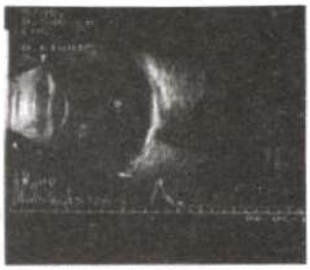

Figure 2: B scan ultrasound showed high surface Reflectivity with posterior shadowing suggestive of ossification

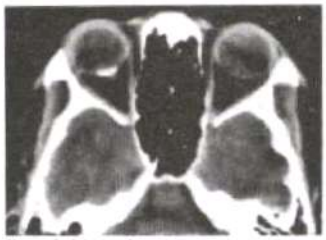

Figure 3: CT scan showed a radio opaque plaque of bone density at the level of posterior pole in OD

Figure 4: FFA showed a large area of early patchy hyperfluorescence (4a) with late staining in the area of lesion (4b) with an area of blocked fluorescence in area of sub retinal haemorrhage suggestive of CNVM

Conclusion

Intravitreal bevacizumab may be an effective treatment modality in management of CNVM secondary to choroidal osteoma

Larger series of such cases need to be studied to further validate our findings

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CASE REPORT

Clinical profile of Familial Optic Nerve Head drusen

Muhammad Moniruzzaman[1], Ankur Gupta[1], P Tandava Krishnan[1], Siddiqur Rahman[2], Ummay Kawser[3], Q M Iqbal Hussain[4]

Abstract

We describe here three siblings of a family who have been diagnosed to have optic nerve head (ONH) drusen. ONH drusen was bilateral in two siblings while the third had unilateral involvement. All the three children had best corrected Snellen's visual acuity of 6/6 in both eyes. Fundus examination revealed elevated disc with blurred margins in the affected eyes. B scan ultrasonography showed highly reflective echo at the level of ONH with acoustic shadowing in the medium gain scan with visual field examination showing variable field defects in the affected eyes of children.

***Conclusion** – ONH drusen should be ruled out in suspicious cases of optic disc edema and in confirmed cases family screening may be useful.*

Synopsis – We describe here three siblings of a family who have been diagnosed to have optic nerve head (ONH) drusen. ONH drusen was bilateral in two siblings while the third had unilateral involvement. All the three children had best corrected Snellen's visual acuity of 6/6 in both eyes. Fundus examination revealed elevated disc with blurred margins in the affected eyes. B scan ultrasonography showed highly reflective echo at the level of ONH with acoustic shadowing in the medium gain scan with visual field examination showing variable field defects in the affected eyes of children

Conclusion – ONH drusen should be ruled out in suspicious cases of optic disc edema and in confirmed cases family screening may be useful Optic nerve head (ONH) drusen are deposits of hyaline calcific material with an incidence of 0.3%.They are usually asymptomatic but occasionally can cause blurring or loss of vision.[1] They are frequently bilateral. They may be familial in nature with irregular dominant trait.[2] Superficial ONH drusen are easily diagnosed with fundoscopy having classic yellow-white glistening hyaline deposits. Buried ONH drusen may not be readily apparent on fundus examination.[3] Swollen appearance of the optic nerve in such cases may be mistaken for disc oedema. ONH drusen are autofluorescent in nature.They can be diagnosed by echography even when they are not evident on funduscopy.[4]

We describe the occurrence of ONH drusen in 3 siblings of a family.

Case 1: The first sibling had best corrected visual acuity (BCVA) of 6/6 in both eyes with normal anterior segment findings. Fundus examination elevated disc with blurred margins in both eyes (figures 1a and 1b). B scan evaluation showed high reflective echo in area of optic nerve head in

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both eyes (figures 1c and 1d). Visual field (30-2 full threshold) showed extensive visual field defects in both eyes (figures 1e and 1f).

Case 2: The second sibling had a BCVA of 6/6 in both eyes with normal anterior segment findings. Fundus examination was normal in right eye and showed disc elevation with blurred margins in the left eye (figures 2a and 2 b). B scan evaluation showed high reflective echo in area of optic nerve head in left eye (figures 2c and 2d).

Case 3: The third sibling had a BCVA of 6/6 in both eyes with normal anterior segment findings with bilateral disc margin blurring and disc elevation. The fourth sibling and the parents had normal findings.

Discussion: Incidence of ONH drusen in adults was described by Lorentzen as being 3.4 per 1,000.[2] The incidence in children is 4 per 1,000.[5] Histological studies of adult autopsy eyes have revealed a higher incidence of ONH drusen 0.5%.[6] Lorentzen found 3.1 % involvement of relatives of patients with ONH drusen i.e. 10 times the incidence in general population.[7] Primary pathology of ONH drusen - most likely an inherited dysplasia of the ONH & its blood supply, which would predispose.[8] ONH drusen are bilateral in 65-91%.[4,9] In our series we had 2 out of 3 siblings having a bilateral presentation. The impairment of visual acuity due to ONH drusen is rare. Mustonen found that in 307 drusen eyes, drusen was responsible for impaired VA in only 2 eyes.[10] All the three siblings in our series had a BCVA of 6/6. Erkkila found visual field defects in 10 of 89 eyes (11%) in children under 15 with buried drusen.11 Hoover et al reported a high frequency of visual field defects in 51% eyes, with 9 eyes showing an enlarged blind spot, 6 eyes an inferior arcuate, sector, or altitudinal defect, and 3 eyes presenting both kinds of visual field defects.[12] We found a visual field abnormality in one amongst the three siblings. B-scan echography has been shown to be the most reliable method in detecting ONH drusen.[13] It is noted as a highly reflective round lesion with acoustic shadowing in the medium gain scan and is characteristically detectable even in low-gain scans due to calcium content. ONH drusen can also be detected by CT scan but they may not be as sensitive as B scan as a slice thickness of 1.5 mm used at optic nerve head may miss smaller drusen.[14,15] CT alone is apt to miss a substantial number of ONH drusen cases. High cost and radiation exposure makes CT unsuitable as routine.

Conclusions

- ONH drusen should be ruled out in suspicious cases of optic disc edema
- Neuroimaging not mandatory as routine
- In confirmed cases family screening may be useful.

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Figure 1

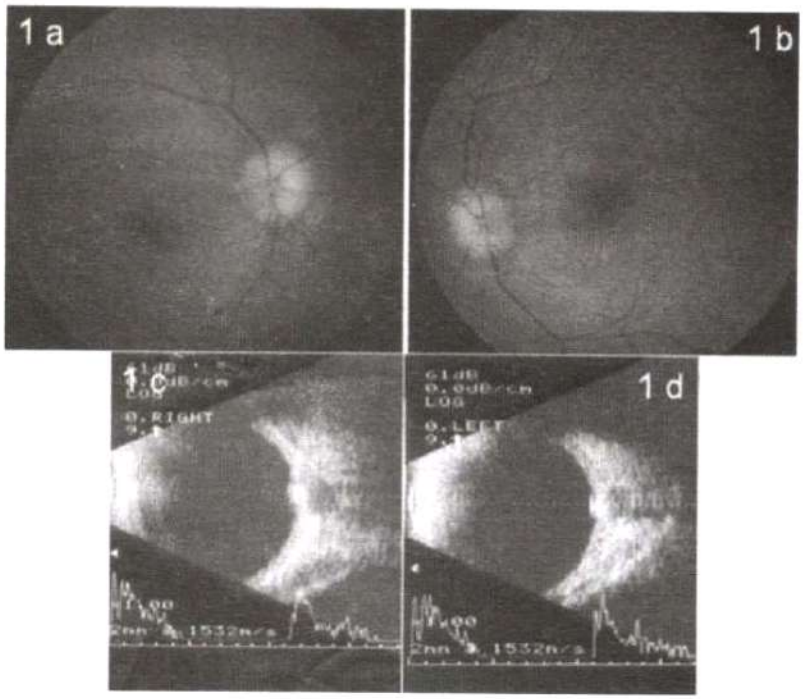

Figure 2

Legends:

Figure 1: Blurring of disc margins are seen in both eyes (figure 1a and 1b) which on echography is seen as a high reflective echo at the level of ONH shadow (figure 1c and 1 d).Extensive visual field defects are also seen in both eyes (figure 1 e and 1f).

Figure 2: Blurring of disc margin is seen in left eye (figure 2b) with a high reflective lesion seen at the level of optic nerve head (figure 2d). The corresponding images for right eye are normal (figure 2a and 2 c).

CASE REPORT

Clinical Profile of Familial Exudative Vitreoretinopathy

Muhammad Moniruzzaman[1], Ankur Gupta[1], Siddiqur Rahman[2], Ummay Kawsar[3] Q M Iqbal Hussain[4], Atiquzzaman[5]

Abstract

Objectives: *To report a case of familial exudative vitreoretinopathy (FEVR) with its clinical presentation, diagnostic procedures and its management.*

Patient and methods: *A 19 years girl who presented with sudden loss of vision in right eye for three months. The diagnosis was established by clinical examination, fluorescein angiography and family screening. Prophylactic photocoagulation and surgical treatment done according the stage of disease in the respective eyes.*

Results: *On clinical examination best corrected Snellen's visual acuity (BCVA) was finger count 2 feet in right eye and 6/12 in left eye. Indirect ophthalmoscopy with scleral indentation showed mild vitreous hemorrhage and tractional retinal detachment in right eye. Left eye showed temporal dragging of major blood vessels, peripheral neovascularization and characteristic peripheral avascular zone with abrupt cessation of the vasculature. Fluoresceine angiography findings were corresponding the clinical picture of the left eye. Prophylactic photocoagulation in left eye and surgery done in right eye. On family screening father had peripheral avascular zone in both eyes but the disease process was not progressing.*

Conclusion: *FEVR appears to be more common than reported. A high index of suspicion, careful examination, family screening and early prophylaxis is very important to prevent avoidable blindness from this underdiagnosed disease.*

Introduction

Familial exudative vitreoretinopathy (FEVR) is a hereditary retinal vascular disorder first described by Criswik and Schepens in 1969.[1] They reported six patients from two families with fundus changes remarkably similar to those seen in retinopathy of prematurity (ROP), but had no history of premature birth or perinatal complications. The major ocular menifestations included peripheral neovascularization, peripheral retinal traction with temporal dragging, falciform folds, retinal detachment and lipid exudation. As the disease is frequently asymptomatic and due to its variable presentation FEVR is frequently unrecognized or diagnosed incorrectly. It can resemble many ocular conditions, depending on the stage of the disease at presentation and the age of the patient. Much has been learned about the natural history, genetics and phenotypic expression of the disease. Despite many advances the pathogenesis and management of this condition remains uncertain.[2]

Patient and methods

A 19 years girl who presented with sudden loss of vision in right eye for three months. Patient had history of high myopia from childhood with no

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history of premature birth or perinatal oxygenation. The diagnosis was established by clinical examination, fluorescein angiography and family screening. Prophylactic photocoagulation and surgical treatment done according to the stage of disease in the respective eyes.

Results

On clinical examination best corrected Snellen's visual acuity (BCVA) was finger count 2 feet in right eye and 6/12 in left eye. Indirect ophthalmoscopy with scleral indentation showed mild vitreos hemorrhage and tractional retinal detachment in right eye (Figure:1). Left eye showed temporal dragging of major blood vessels (Figure:2), peripheral neovascularization and characteristic peripheral avascular zone with abrupt cessation of the vasculature (Figure:3). Fluorescein angiogram showed peripheral abrupt ischemic zone with leaking new vessels at the junction between perfused and non perfused retina in left eye (Figure:4). In right eye 360^0 band with pars plana vitrectomy, membrane peeling, endolaser and silicon oil injection done. Prophylactic peripheral retinal photocoagulation done in left eye. In between laser sittings vitreous hemorrhage occurred which resolved spontaneously. Silicon oil was removed from right eye one month after due to early emulsification. Upto six months follow up retina was attached in right eye and BCVA was 6/60 and 6/12 in left eye. Among her family members her father had peripheral retinal avascular zone but it was asymptomatic and not progressive.

Discussion

FEVR is most commonly an autosomal dominant disease with near 100% penetrance.[3,4,5,6] Autosamal recessive, X – linked recessive inheritance and sporadic cases have also been reported.[7,8,9] Two genes are known to be associated with autosomal dominant FEVR: *FZD4, LRP5*.[10,11] It is characterized by failure of peripheral retinal vascularization which by itself usually causes no clinical symptoms. The visual problems and variable presentation associated with autosomal dominant FEVR result from secondary complications caused by retinal ischemia. The retinal avascularity is probably present from birth and generates sequelae that stabilize in early adult life or progress in later life. This region of retinal avascularity is presumed to be present from birth and initially was thought to generate sequelae that stabilized in early adult life but now it is suggested that disease progression is possible into later life also.[5,2] Expressivity may be asymmetric and is highly variable, ranging from mild or asymptomatic to severe (e.g., registered as blind) within the same family.[12]

The diagnosis of FEVR is based on a family history compatible with autosomal dominant inheritance and bilateral peripheral retinal avascularity, seen temporally by indirect ophthalmoscope and scleral indentation, or by fundus fluorescein angiography. Other retinal abnormalities include retinal neovascularization, peripheral brush border anastomoses, falciform retinal folds, retinal traction associated with macular ectopia and/or straightening of vessels at the posterior pole, acute-angle vascular branching, retinal detachment, retinal tears or holes. Exudation may be associated with retinal detachment. Rare retinal features include retinoschisis and giant retinal tears.[12] In our case there was vitreous hemorrhage with tractional retinal detachment in one eye. Other eye showed characteristic peripheral avascular zone, peripheral neovascularization and temporal dragging of major blood vessels. Fluorescein angiogram also showed peripheral abrupt ischemic zone with leaking new vessels. On family screening we found her father having peripheral retinal avascular zone which is asymptotic and not progressive.

The important differential diagnoses are retinopathy of prematurity (ROP), high myopia,

Eales disease, Coat's disease, juvenile retinoschisis, persistant hyperplastic primary vitreous (PHPV) and toxocariasis. Patients with ROP will have a history of prematurity and perinatal complications and will develop vascularization of retinal periphery in the majority of eyes after involution of the diseases.[13] High myopia does not show the typical peripheral avascular zone in fluorescene angiogram. Eales disease show inflammatory vascular sheathing; exudation and macular ectopia are not seen.[14] Coat's disease is typically unilateral, has male predilection and shows no evidence of vitreoretinal traction.[15] Juvenile retoinoschisis is bilaterally symmetrical, affects only male children, has typical foveal schisis and peripheral new vessels with exudation are rare.[13,16] PHPV is also unilateral and usually associated with micropthalmia.[15] Toxocara granuloma in children is typically unilateral and there is associated vitritis and uveitis.

Avascular retina does not require treatment until it develop peripheral neovascularization which can be treated with prophylactic argon laser photocoagulation or cryopexy to induce regression of the new vessels. Tractional retinal retinal detachments with or without retinal breaks are repaired surgically. Exudative retinal detachments may be stabilized with cryotherapy, but the prognosis is poor.[12] We had done vitrectomy and silicon oil injection due to tractional retinal detachment in right eye but silicon oil was removed one month after due to early emulsification. In left eye prophylactic laser done and in between laser sittings vitreous hemorrhage occured which was resolved without any intervention. After six months follow up BCVA was stable as 6/60 in right eye and 6/12 in left eye.

Children should undergo regular fundus examination to evaluate disease progression. Asymptomatic individuals with a small region of peripheral avascularity may require annual review only. Those with active neovascularization or exudate may well require treatment and close observation thereafter.[12]

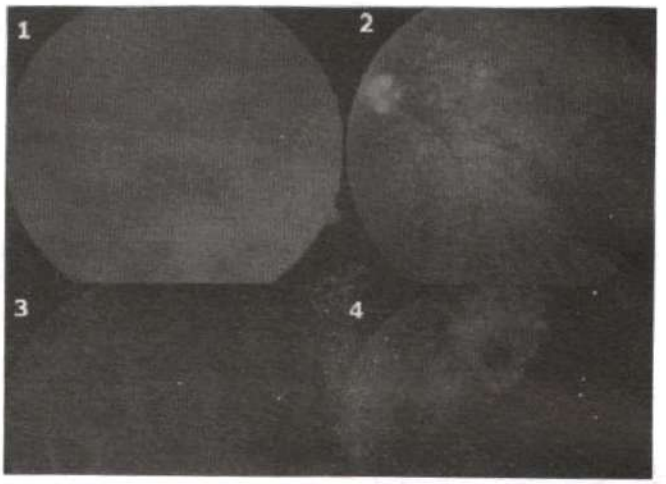

Figure 1: Fudus photograph showing mild vitreous hemorrhage with tractional retinal detachment in right eye, Figure 2: Fundus photograph of posterior pole of left eye shows temporal dragging of macula and major vessels, Figure 3: Fundus photograph of periphery showing the charachteristic peripheral avascular zone with abrupt cessation of the vasculature in left eye. Fioure 4 : Fluorescein angioaram.

Conclusion

FEVR is more common than it is reported. Careful family screening, high index of suspicion is essential for early detection of the disease in view of its progressive nature, unfavorable visual outcome of surgery and to prevent avoidable blindness.

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