Nevus Sebaceous of Jadassohn Associated With Macro Optic Discs and Conjunctival Choristoma

Following Jadassohn’s report in 1895, congenital ocular findings such as epibulbar choristomas (lipodermoids, aberrant lacrimal glands, cartilage), corneal pannus, colobomas (eyelid, iris, retina or choroid, optic nerve), and optic nerve hypoplasia but not macro optic discs have been described in combination with a facial nevus sebaceus.

Report of a Case. A 42-year-old man was first seen with a dark lesion on the left side of the forehead (4 × 4 cm, Figure 1, A). Best-corrected visual acuity was 0.7 OD and 0.4 OS with a refractive error of – 0.5 diopter OU. The right eye showed a superior corneal pannus and an elevated yellowish subconjunctival mass parallel to the superior limbus. The left eye showed an incomplete coloboma of the upper eyelid and an elevated mass adjacent to the nasal limbus. A scleral staphyloma was located at the 5-o’clock position and a corneal pannus in the inferotemporal quadrant (Figure 1, B). In addition, there was a large yellowish mass extending from the lacrimal gland (Figure 1, C). Both optic discs were very large, with the size of the right exceeding that of the left disc. The margin of the right disc was clearly defined except for the inferotemporal quadrant that showed a rather indistinct margin (Figure 2, A). The margin of the left disc was less clearly defined and showed an irregular indistinct contour in both the nasal and the temporal inferior quadrant (Figure 2, B). The branching pattern of the vessels of the right disc was nearly normal except for a slight nasal dislocation. However, the branching pattern of the left eye appeared to be abnormal (Figure 2, B). Both discs appeared relatively pale with a cup-disc ratio of about 0.9 and a vital, irregular neuroretinal rim (Figure 2, A and B). Measurement with laser scanning tomography showed areas of 6.4 and 12 mm² (right and left, respectively) while the cup volume was 3.5 and 10.8 mm³ (right and left, respectively) (normal, 2.17 ± 0.55 mm² and 0.37 ± 0.31 mm³, respectively) (Figure 2, C). Results of function tests...
(eg, visual fields, electroretinography) were normal. Histologic findings from the epibulbar mass of the left eye showed ectopic lacrimal gland, fat, and cartilage (Figure 1, D). Histologic examination of the irregular, dark skin lesion on the left side of the forehead showed a nevus of Jadassohn. Findings from all of the neurological and neuroradiological examinations (computed tomography, magnetic resonance imaging) were unremarkable.

**Comment.** Jadassohn originally used the term “organoid nevus” to describe the combination of sebaceous gland alterations with nevi. Later the term “nevus of Jadassohn” was coined for lesions of the facial skin with sebaceous gland enlargement. Although better known in dermatology, sebaceous nevus with ophthalmological abnormalities and often neurological abnormalities (not present in our patient) has rarely been mentioned in ophthalmological literature. Such an association is distinct from the linear nevus syndrome described by Feuerstein and Mims as the triads of linear facial nevi, seizures, and mental retardation.

The most striking finding in our patient was macro optic discs that rarely occur in the general population. Based on the Gaussian distribution curve of the optic disc size, a macrooptic disc can be defined as a disc with an area exceeding that of the mean + 2 SDs. According to our data on the normal German population, the minimal size for a macro optic disc would be 3.27 mm², which correlates with other data (4.4 mm² for fundus photography). Based on these data, less than 1 of 1000 emmetropic eyes would have the disc size observed in our patient. Interestingly, most of such eyes do not have the visual impairment as noted in our patient.

Although the pathogenesis is unknown, a developmental arrest in the seventh or eighth month of gestation has been suggested in the linear nevus syndrome. This hypothesis of a disturbance of cell migration and differentiation due to toxic or infectious agents is shared by others. The corneal pannus in our patient could therefore be explained by a disturbance of differentiation of corneal epithelial stem cells at the limbus. Also a defect of fusion and separation of tissue during the early development of the retina may have contributed.

**Figure 2.** A, Fundus photograph of the macro optic disc of the right eye. B, Fundus photograph of the macro optic disc of the left eye. C, Laser scanning tomographic measurement of the same disc reveals an area of 12 mm² (normal, 2.17 mm²).
weeks of pregnancy has been suspected in nevus of Jadassohn associated with ocular malformations. This theory could help in understanding the previously reported abnormalities of the optic disc such as colobomas as well as deep cupping and abnormal blood vessels as recently described by Shields et al and the macrooptic discs in our patient both of which might result from the same defect of fusion and separation.

We observed stable ocular findings over 2 decades. Since the nevus sebaceus may undergo benign or malignant transformation, patients with choristomas and/or colobomas should see a dermatologist for evaluation of their skin.

Friedrich E. Kruse, MD
Klaus Rohrschneider, MD
Reinhard O. W. Burk, MD
Hans E. Volcker, MD
Heidelberg, Germany

None of the authors has any proprietary interest in the methods or materials described in this report.

Corresponding author: Friedrich E. Kruse, MD, Department of Ophthalmology, University of Heidelberg, Im Neuenheimer Feld 400, D-69120 Heidelberg, Germany (e-mail: Friedrich.Kruse@ukl.uni-heidelberg.de).

Mooren Ulcer Following Epikeratoplasty for Keratoconus

Epikeratoplasty offers a reasonable therapeutic option for many young patients with keratoconus in Saudi Arabia who are contact lens intolerant because of unfavorable climatic conditions, often with concomitant vernal keratoconjunctivitis, which have a relatively clear visual axis, and for whom the higher risks of penetrating keratoplasty offset the benefits of a slightly better visual prognosis. We report the first case, to our knowledge, of Mooren ulcer occurring following epikeratoplasty performed for keratoconus.

Report of a Case. A 12-year-old Saudi girl was referred to the Anterior Segment Division of the King Khaled Eye Specialist Hospital in Riyadh for management of keratoconus. Her ocular history was remarkable for chronic vernal keratoconjunctivitis that was well controlled with topical mast cell inhibitors. Her best-corrected spectacle visual acuity was 20/160 OD with −6.00 −4.00 × 120 and 20/40 OS with −6.00 −4.00 × 25. Keratometry readings were 54.50/57.25 OD and 56.00/49.50 OS, with much more marked irregularity in the right eye. Findings from slitlamp examination showed apical corneal thinning and Vogt striae, more pronounced in the right than in the left eye. There was no visually noteworthy apical scarring in either eye. She was fitted with rigid gas permeable contact lenses and had a visual acuity of 20/25 OU, but proved to be contact lens intolerant.

She underwent epikeratoplasty on January 1, 1996, in the right eye with a 9-mm graft in an 8.5-mm bed with 16 interrupted sutures. Unfortunately, all sutures loosened prematurely, requiring complete suture removal between 2 and 4 months postoperatively. She returned on May 7, 1996, with a poorly adherent, edematous graft with peripheral melting and thinning of the graft with serrated edges. Following removal of the epikeratoplasty lenticule, the cornea reepithelialized, was clear and compact, and her uncorrected visual acuity was 20/80 OD.

On June 1, 1996, she returned with a complaint of moderately severe pain and poor vision in the left eye. On examination, the visual acuity was counting fingers at 6 ft OD and 20/25 OS. There were progressive peripheral thinning and vascularization involving the entire periphery of the right cornea, with marked centripetal extension in the superior cornea from the 10- to 2-o’clock position (Figure). The leading edge of centripetal extension was undermined with corneal stromal involvement 1 to 2 mm beyond the apparent clinical edge of involvement. There was an epithelial defect.
involving the entire cornea. Results from examination of the left eye were normal with the exception of the unchanged appearance of her pre-existing keratoconus. She was hospitalized and treated for 7 weeks with a bandage contact lens, topical corticosteroids, progesterational steroids, tetracycline ointment, lubricants, systemic prednisone, and doxycycline. Findings from her systemic evaluation (which included erythrocyte sedimentation rate, rheumatoid factor, antinuclear antibodies, immunoglobulin levels, and electrophoresis) were normal with the exception of a markedly elevated IgE level (687 U; normal <100 U [to convert to the SI units of milligram per liter, IgE first needs to be reported as milligram per deciliter; then multiply this value by 10 to get the SI value]). There was no clinical evidence of systemic helminth infection, although specific serum and gastrointestinal evaluation was not performed. Therapy resulted in reduction in inflammation, vascular regression, cessation of centripetal spread of the ulcer, and preservation of the clarity of the central visual axis. Conjunctival resection was not performed because of her favorable response to therapy with the medications prescribed. Systemic immunosuppression was not used because of her age. She was discharged with an uncorrected visual acuity of 20/80 OD. At no time during her hospitalization was there any inflammation or other evidence of any involvement of the left eye.

Over the next several months, there was progressive scarring, thinning, and vascularization of the central cornea, with a reduction in her visual acuity to counting fingers at 1 ft. With continuous therapy for her vernal keratoconjunctivitis and vascularization with topical cromolyn and medroxyprogesterone, there was gradual clearing of the visual axis over the next 12 months, although the entire cornea remained thin. At the time of her last examination, visual acuity had improved to 20/125 OD uncorrected, and to 20/100 OD with pinhole. At no time during the follow-up period was there any inflammation or other evidence of any involvement of the left eye.

Comment. Mooren ulcer is a painful, relentless, chronic ulcerative keratitis that begins peripherally and progresses circumferentially and centrally.1 It is by definition idiopathic, meaning that it occurs in the absence of any systemic disorder to which ulcerative keratitis may be attributed.2 There have, however, been a number of systemic diseases associated with Mooren ulcer, including helminthiasis, syphilis, tuberculosis, and chronic hepatitis C.2 In addition, there have been a number of reports of local corneal disorders associated with Mooren ulcer that include physical trauma, foreign bodies, chemical burns, and herpes simplex and herpes zoster infections,2 as well as surgical procedures such as cataract extraction and penetrating keratoplasty.2

With this case report, epikeratoplasty has been added to this list. While this patient is in the age range typical of the “malignant variety” of Mooren ulcer, her strictly unilateral course lends support to the observation that the epikeratoplasty was the initiating event. Although it is tempting to speculate that the concomitant presence of atopic keratoconjunctivitis, irritation from premature suture loosing, and/or antigenic reaction to the lenticule may have played a role in the pathogenesis of the Mooren ulcer, it must be emphasized that in more than 500 cases, epikeratoplasty has been performed for keratoconus on patients with similar ocular surface and allergic abnormalities without such a complication at the Anterior Segment Division at King Khaled Eye Specialist Hospital.

Klaus D. Teichmann, MD, FRCS, FRACO
Riyadh, Saudi Arabia
Michael D. Wagoner, MD
Boston, Mass
Riyadh

The authors do not have any proprietary interest in the epikeratoplasty lenticule or the manufacturer, nor do they have any financial interest or receive any payment as a consultant, reviewer, or evaluator for the manufacturer.

Reprints: Michael D. Wagoner, MD, Medical Library, King Khaled Eye Specialist Hospital, PO Box 7191, Riyadh 11462, Kingdom of Saudi Arabia.

Infectious Keratopathy Complicating Photorefractive Keratectomy

Excimer laser photorefractive keratectomy is becoming an increasingly popular method to treat ametropia. The main issue regarding the safety of this procedure has been the postoperative loss of best-corrected visual acuity from various origins, including irregular astigmatism, corneal scarring, and centered ablations.1 Infection following photorefractive keratectomy is rare; at least 7 patients developed infectious keratitis following excimer laser keratectomy, which in one case progressed to severe endophthalmitis.1,2 We report Staphylococcus aureus keratitis in the early postoperative period following photorefractive keratectomy with the development of severe degenerative keratopathy.

A 27-year-old man was seen for a corneal ulcer in the left eye that had developed 2 days following an uncomplicated photorefractive keratectomy to correct high myopia. A bandage contact lens was placed over the eye that also was treated with topical trimethoprim sulfate-polymyxin B sulfate, fluorometholone, and ketorolac tromethamine. One day postoperatively, the cornea was clear and there was a mild anterior chamber reaction. On the second postoperative day, there was a corneal infiltrate with hypopyon (Figure 1, left). Corneal cultures grew S aureus. The left cornea was treated with topical fortified ciprofloxacin and cefazolin eye drops every half hour and homatropine hydrobromide eye drops twice daily. The hypopyon resolved within 4 days. The ulcer slowly healed during the next 2 months with partial clearing of the central opacity thereafter. Fifteen months later, a chalazion developed in the right upper eyelid from which S aureus was cul-
tured, suggesting that the patient was a carrier of this microorganism. The pinhole visual acuity was 20/100 OS. A substantial ringlike central stromal scar associated with stromal thinning was present (Figure 1, right). Four months later, a penetrating keratoplasty was performed; the graft remained clear 12 months postoperatively.

Histopathologic examination of the corneal button revealed several...
large clusters of eosinophilic material containing occasional cholesterol clefts within the superficial half of the central stroma (Figure 2). These deposits exhibited congophilic with apple-green birefringence and stained positive for amyloid protein by immunohistochemistry and for glycosaminoglycans with the colloidal iron and alcan blue stains (Figure 2). Furthermore, electron microscopy showed clusters of nonbranching fibrils 10 nm in diameter consistent with amyloid. Otherwise, there was irregular thickening of the epithelium of the central cornea overlying an absent Bowman membrane, superficial stromal scarring, and mild endothelial attenuation.

The cause of postphotorefractive keratectomy corneal infiltrates may be infectious, toxic, hypersensitivity, or a combination of these processes. An infectious corneal ulcer is a rare but potentially devastating complication of photorefractive keratectomy that may lead to endophthalmitis and to substantial scarring, requiring penetrating keratoplasty, as in our case. Although a rare complication of photorefractive keratectomy, infectious keratitis is of concern, especially since bilateral photorefractive keratectomy is performed frequently in the same surgical intervention.

Our case is an example of secondary amyloid, lipid, and glycosaminoglycan degeneration of the cornea. Amyloid and lipid corneal degenerations are not uncommon sequelae of both infectious and traumatic keratopathies. Glycosaminoglycan deposition has been described as a component of subepithelial opacities following excimer laser photorefractive keratectomy in both human and rabbit corneas and may be responsible for the late haze following photorefractive keratectomy.

Vivian E. Hill, MD
Seymour Brownstein, MD
W. Bruce Jackson, MD
George Mintsiosids, MD
Ottawa, Ontario

We thank Merrill D. Benson, MD, professor and chairman, Department of Medical and Molecular Genetics, Indiana University School of Medicine, Indianapolis, for performing the immunohistochemical stains for amyloid protein.

Corresponding author: Seymour Brownstein, MD, University of Ottawa Eye Institute, 501 Smyth Rd, Room 3818, Ottawa, Ontario, Canada K1H 8L6.


Optic Neuropathy Resembling Normal-Pressure Glaucoma in a Teenager With Congenital Macrodiscs

Normal-pressure glaucoma is a special form of the chronic open-angle glaucomas characterized by loss of neuroretinal rim and retinal nerve fiber layer, perimetric defects, and normal intraocular pressure measurements. Age of the patients at the time of the diagnosis is usually in the range of 40 to 50 years. We report an unusual occurrence of an optic neuropathy resembling normal-pressure glaucoma in a 13-year-old patient with a follow-up time of 10 years.

Report of a Case. In 1987, at the age of 3 years, the patient was referred to us for exclusion of congenital or infantile glaucoma. During a routine ophthalmologic examination, the referring ophthalmologist had noticed an abnormally large optic cup. Visual acuity measured 12/20 OD, and repeated intraocular pressure measurements determined by applanation tonometry varied between 10 and 14 mm Hg. Horizontal and vertical corneal diameters ranged between 11.5 and 12.0 mm. Morphometric evaluation of color stereo optic disc photographs showed primary macrodiscs with pseudoglaucomatous but physiologic macrocupping. Visual fields were not examined owing to the age of the patient.

Ten years later, the boy complained of a shadow in his left eye. He attended high school with good to excellent performance in all subjects including sports. Repeated computerized and Goldmann perimetry revealed a deep Bjerrum scotoma in the inferior visual field of the left eye. Results of a perimetric examination of the right eye were unremarkable. Visual acuity measured 20/20 OU. Refractive error had changed from emmetropia to mild myopia (−2.5 diopters [D]). Axial length had increased from 21.3 to 25.4 mm. Relatively low keratometric readings of 40.5 D OD and 39.5 D OS with no marked astigmatism (<1.0 D) explained the relatively low myopic refractive error of −2.5 D despite the axial length of 25.4 mm. Intraocular pressure evaluated in day-and-night pressure profiles containing recordings at 5 PM, 9 PM, midnight, 7 AM, and noon ranged between 10 and 17 mm Hg with no significant difference (P > .50) between the right and left eye. Morphometric analysis of optic disc photographs showed that the optic disc shape had changed to a horizontally oval configuration.

In the right eye, rim shape was normal with the smallest rim part in the temporal disc sector (Figure 2, left). A disc hemorrhage was detected in the temporal superior disc sector. In another follow-up examination after an additional 4 months, the hemorrhage had recurred at the same location. In the left eye, neuroretinal rim notching was present in the superior disc region (Figure 2, right). In both eyes, parapapillary atrophy had developed in the temporal superior quadrant. Wide-angle red-free retinal nerve fiber layer photography showed a diffusely reduced visibility of the retinal nerve fiber layer in both eyes with an additional segmental loss in the temporal superior fundus region of the left eye. Results of repeated cranial computed tomography, magnetic resonance imaging, and neurologic and pedi-
atic examinations were unremarkable. Arterial blood pressure was normal for age. All examinations were repeated after 4 and 10 months and showed similar results, with no progression detectable in perimetry and morphometric optic disc analysis. All 4 other family members also had abnormally large optic discs.

Comment. With completely unremarkable neuroradiologic and pediatric examination results and normal recordings of intraocular pressure in day-and-night pressure profiles, our young patient with an inherited primary macrodisc may have an unusually early-onset optic neuropathy resembling normal-pressure glaucoma. Loss of neuroretinal rim and increase in the β zone of parapapillary atrophy, which is typical for glaucomatous optic neuropathy and which usually does not occur in nonglaucomatous optic neuropathies, point against a nonglaucomatous reason for optic nerve damage and visual field loss. The change in the optic disc shape, which was not paralleled by a marked increase in corneal astigmatism, deserves special attention, because after age 3 years a change in optic disc shape has so far been found only in eyes with progressive high myopia. One may discuss whether this change in optic disc shape, the abnormally large optic disc size, and the development of parapapillary atrophy may have resulted in mechanical stretching and tearing of the lamina cribrosa, including the microvasculature of the optic nerve head, or in other alterations, leading to focal loss of neuroretinal rim, flame-shaped optic disc hemorrhages, and deep Bjerrum scotoma in our patient with normal intraocular pressure.

Jost B. Jonas, MD
Claus Cursiefen, MD
Wido M. Budde, MD
Erlangen, Germany

This study was supported by Deutsche Forschungsgemeinschaft SFB 539.
Reprints: Jost B. Jonas, MD, Universitäts-Augenklinik, Schwabachanlage 6, 91054 Erlangen, Germany (e-mail: Jost.Jonas@augen.med.uni-erlangen.de).

2. Jonas JB, Gusek GC, Naumann GOH. Optic disc, cup and neuroretinal rim size, configuration, and

Figure 1. Optic disc photographs of the right eye (left) and left eye (right) at the age of 3 years. Disc area was 5.22 mm² OD and 4.99 mm² OS (means±SD optic disc area in 457 normal eyes: 2.69±0.70 mm²); optic cup area was 3.07 mm² OD and 2.97 mm² OS.

Figure 2. Optic disc photographs of the right eye (left) and left eye (right) at the age of 13 years. Note hemorrhage in the right eye, β zone of parapapillary atrophy (arrowheads), and rim notching in left eye (arrows).
Intraocular Tuberculosis Without Detectable Systemic Infection

Intraocular tuberculosis (TB) is rare; infection occurs via hematogenous spread, usually from the lung. Resurgence of tuberculosis in the developed world as a result of immigration, acquired immunodeficiency syndrome, and drug resistance has led to improved methods of diagnosis. Mycobacterium tuberculosis can be difficult to isolate in vitro. Amplification of DNA by polymerase chain reaction (PCR) has recently been used to identify M tuberculosis in ocular tissue samples of patients with systemic TB.1 We describe 3 patients with TB.

Case 1. A 53-year-old man of Indian ethnic origin was referred with a 6-day history of deteriorating vision in the left eye. There was no history of exposure to TB. Medical history was significant for ischemic heart disease. The visual acuity was hand movements OS and 20/30 OD. He had a left hypopyon, dense vitritis, and a choroidal lesion, 4 disc diameters across, superotemporal to the disc. Investigations revealed increased plasma viscosity (1.9 centipoise units), a negative autoantibody screen, and a normal serum angiotensin–converting enzyme level. A chest x-ray film was normal and Mantoux testing (10 TU/0.1 mL) was negative. The serum angiotensin–converting enzyme level was elevated at 62 U/L and a gallium scan was positive (Figure 2). A transvitreal biopsy specimen demonstrated noncaseating granuloma (Figure 3). No acid-fast bacilli were seen. The presumptive diagnosis of sarcoidosis was made and oral steroid therapy was begun. Following initial ophthalmic improvement, she developed ocular, spinal, and axillary abscesses, from which M tuberculosis was eventually isolated. She subsequently underwent enucleation of the affected eye with microbiological confirmation of M tuberculosis.

Case 2. A 27-year-old woman was seen with a 5-day history of floaters in the left eye. Visual acuity at the initial visit was 20/30 OU. Anterior segment examination was unremarkable. She had vitritis in the left eye, overlying an area of retinitis superotemporal to the disc with associated retinal neovascularization (Figure 4). She was generally well and initial investigations revealed only a mildly raised plasma viscosity and normal chest x-ray films. There was no history of recent exposure to TB. Her most recent visit to Pakistan was 5 years previously.

Mantoux testing (1 and 10 TU/0.1 mL) failed to provoke a reaction. Two chest x-ray films, blood cultures, and early-morning urine samples were negative for TB. Toxoplasma, herpes simplex, cytomegalovirus, and Toxocara titers were unremarkable. An anterior chamber tap was performed and samples sent for culture (tuberculosis, bacteriology, viral, and fungal), along with PCR testing for TB. Cultures were negative.

Case 3. A 32-year-old Pakistani woman was seen with a 5-day history of floaters in the left eye. Visual acuity at the initial visit was 20/30 OU. Mantoux screening revealed only a mild increase in plasma viscosity and normal chest x-ray films. There was no history of recent exposure to TB.

No alcohol or acid-fast bacilli were isolated from early-morning urine samples, blood cultures, gastric secretions, or pleural fluid. Mantoux testing (1 and 10 TU/0.1 mL) was negative.

An aqueous tap was obtained and samples sent for culture and PCR testing for TB, toxoplasma, herpes simplex, and Toxocara species. Microscopy and cultures failed to isolate any organism, but PCR testing identified M tuberculosis. Anti-TB therapy was begun with rapid resolution of the uveitis. The choroidal lesion involuted to a flat scar (Figure 4), with improvement in visual acuity to 20/40 OS.

References
negative but *M. tuberculosis* was identified by PCR testing. The patient is responding well to triple antituberculous chemotherapy and the retinitis is resolving (Figure 6).

**Comment.** Ocular manifestations of systemic TB include granulomatous uveitis, retinal periphlebitis, and choroidal tubercles. The most common manifestation of intraocular TB is choroiditis; tuberculous infection of the retina is very rare and is thought to occur via retinal blood vessels. In all cases described, ocular features were compatible with intraocular TB but conventional investigations for TB were unsupportive.

In all our patients, sputum, urine, and blood cultures failed to isolate the causative organism. None of our patients had chest x-ray films suggestive of TB. It is possible they did not have pulmonary TB. Furthermore, a normal chest radiograph does not exclude endobronchial infection. Early-morning sputum specimens have a higher yield and lower contamination rate compared with random specimens, but the sensitivity remains quite low. Positive urinary cultures in nongenitourinary TB may be as low as 1.2% and so cannot be relied on.

Recently evolved molecular biological techniques have provided rapid, sensitive, and specific methods of diagnosing *M. tuberculosis* infection. Polymerase chain reaction has been used to detect the presence of mycobacterial tuberculous DNA from sputum samples, gastric aspirates, and tissue samples. Only 0.01 mL of sample is required and is easily accessed via aqueous aspiration in a minimally invasive procedure with fewer associated complications than transvitreal biopsy. A definitive diagnosis of intraocular TB is obtained.

Tuberculosis may manifest itself in the eye without obvious involvement of other commonly affected organs or evidence of a
systemic illness. Our experience suggests that investigations conventionally used to detect TB are sometimes ineffective for diagnosing ocular infection. However, PCR testing on the aqueous humour samples made diagnosis of intraocular TB possible. We would advocate its early use in any instance where there is suspicion of ocular TB and conventional tests have been unhelpful. The consequences of a misdiagnosis or even a late diagnosis may be disastrous.

Nagini Sarvananthan, MBBS, MMEdSci
Martin Wiselka, FRCP
Kim Bibby, FRCS, FRCOphth
Leicester, England

Corresponding author: Kim Bibby, FRCS, FRCOphth, Leicester Royal Infirmary, Infirmary Road, Leicester LE1 5WW, England.


**Optical Coherence Tomography in Successful Surgery of Vitreomacular Traction Syndrome**

Vitreomacular traction syndrome (VTS) is a disorder caused by incomplete posterior vitreous detachment with persistent traction on the macula that produces in most eyes cystoid changes and decreased visual acuity.1 We report the use of optical coherence tomography (OCT) in a patient in whom clinical findings were characteristic of VTS, and the anatomical outcome of successful surgery.

**Report of a Case.** A 67-year-old woman was referred to our institution for assessment of visual loss. She complained of a 2-month history of blurred vision in the right eye. Visual acuity was 20/80 OD and 20/25 OS. Fundus examination by biomicroscopy revealed macular cysts with posterior hyaloid attachment at the posterior pole and no epiretinal membrane in the right eye. Fluorescein angiography confirmed macular cystoid edema. B-scan ultrasonography revealed vitreous adhesion to the optic nerve and macula with clear vitreoretinal separation peripheral to the vitreofoveal junction in the right eye. The left eye was normal.

Ocular coherence tomography (Humphrey Instruments, San Leandro, Calif) showed a thickened retina with localized nonreflective cystoid spaces in the outer retina of the right eye and a large central cyst that extended near the internal limiting membrane (Figure 1). Central macular thickness measured by OCT was 604 µm. The scan also showed the presence of a thin, moderately reflective membrane from the surface of the retina at the fovea, which followed the configuration of the posterior vitreous and was identified as the posterior hyaloid.

The diagnosis of VTS was made and a 3-port pars plana vitrectomy performed. After the excision of the posterior two thirds of the vitreous gel, the posterior vitreous surface was cut, thereby relieving anterior-posterior traction. The posterior hyaloid membrane was easily separated from the macula by gentle suction with the vitrectomy probe, and later it was clearly separated from the papilla. No gas or air tamponade was used.

Four weeks after surgery, visual acuity improved to 20/25 OD. Fundus examination revealed a red-dish, round (600 µm in diameter),
sharp circumscribed lesion, corresponding to the location of the preoperative central macular cyst, that had the clinical appearance of a lamellar macular hole. An OCT scan showed partial restoration of normal appearance with less retinal thickening, much less prominent cystic spaces, and no hyaloid adherence on the inner surface of retina, but a marked central depression similar to the appearance of a lamellar macular hole (Figure 2). Maximum retinal thickness at the margin of the hole after surgery was 422 µm at 2 weeks, 386 µm at 4 weeks, and 349 µm at 8 weeks. Retinal thickness at the base of the hole after surgery was 233 µm at 2 weeks, 225 µm at 4 weeks, and 204 µm at 8 weeks. Fundus examination results remained unchanged at all postoperative times.

Comment. High-resolution cross-sectional in vivo OCT imaging of the retina, with a longitudinal resolution of 10 µm, is a useful tool for identifying and monitoring macular diseases. In this case of type B VTS (as classified by Smiddy et al), OCT provided images of localized vitreous adhesion caused by VTS and also showed secondary cystoid changes in the macula. Features of VTS discovered with OCT have been previously reported, mainly associated with epiretinal membrane; however, in this case no epiretinal membrane could be detected either by ophthalmoscopy or OCT. Our findings are similar to those of stage 1 impending macular hole in that there is an absent foveal pit and minimal reflective spaces within the fovea without full-thickness loss of retinal tissue or vitreous separation. The main differences are the greatly increased macular thickness, the persistence of retinal tissue at the base of the fovea, and the hyaloid attachment limited to the fovea.

When patients have progressive visual impairment due to VTS, vitrectomy may provide functional and anatomical improvement. In our patient, postoperative OCT scans revealed that posterior hyaloid reflexes were absent and also that macular edema and cyst spaces were resolving, which was correlated with a substantial increase in visual acuity.

We conclude that macular examination with OCT allows confirmation of the diagnosis of VTS, helps to explain the pathogenesis of the disease, and can objectively assess the anatomical improvement of secondary macular changes.

B. 4 weeks after surgery, optical coherence tomography shows a faint decrease in retinal thickness and less intraretinal fluid at the base of the hole. C, 8 weeks after surgery retinal thickness is already normal. Intraretinal fluid, seen as low backscattering cavities, is markedly reduced.

Potential Role of Computerized Visual Field Testing for the Appraisal and Follow-up of Birdshot Chorioretinopathy

Birdshot chorioretinopathy (BC) is a chronic intraocular inflammatory disease of unknown etiology, characterized by multiple deep-retinal creamy lesions scattered throughout the fundus up to the equator. Other features include vitreitis, vasculitis, cystoid macular edema, and optic disc swelling. Evolution can be quite variable from case to case. Therapeutic intervention usually consists of the concurrent administration of 2 immunosuppressive drugs, in most cases cyclosporine and corticosteroids. Because of the severity and multiplicity of the side effects, the decision to treat depends on disease severity. The factors routinely used to evaluate disease severity are the clinical evaluation of inflammation, visual acuity, fluorescein angiography, and subjective visual disturbance. A fac-
tor that seems to have been neglected so far is visual field testing. The case reported herein indicates that computerized visual field testing seems to be an additional useful factor in the assessment of the severity of BC and in monitoring response to therapy.

Report of a Case. A 43-year-old white man was examined for the first time for flickering and floaters in his right eye. Best-corrected visual acuity was 20/20 OU for near and far. Anterior chamber inflammation in the right eye was discovered subclinically, amounting to 5.2 photons per millisecond measured by laser flare photometry. There was a slight vitreitis in both eyes, and on fundus examination scattered punched-out creamy lesions were seen in both eyes compatible with BC, a diagnosis confirmed by the presence of the HLA-A29 antigen. Fluorescein angiographic findings showed all the classic signs of BC, such as papillitis, diffuse vasculitis, massive fluorescein impregnation of the retina, and a pseudodelay in retinal arteriovenous circulation time. Results of indocyanine green angiography (ICG) was also typical for BC, showing scattered dark dots at the intermediate and late angiographic phases, fuzzy choroidal vessels at the intermediate angiographic phase, and late zonal hyperfluorescence (Figure 1, A and C).

The patient was not treated for 2½ years thereafter because his condition remained relatively stable. After 2 years, the patient complained of a decrease in visual performance despite having a visual acuity of 20/20 OU. Computerized visual fields (Octopus, program G1, Interzeag, Zurich, Switzerland) showed elevation of the mean defect (MD) (MD OD, 9.4 dB; MD OS, 5.1 dB) and of the loss variance (LV) (LV OD, 54 dB; LV OS, 23.5 dB) in both eyes, but more so in the right eye. Six months later, flickering in the right eye increased markedly, visual acuity decreased to 20/25 OD for far and 20/30 for near, and the visual field deteriorated further in both eyes (MD OD, 9.5 dB; MD OS, 6.1 dB; LV OD, 55 dB; and LV OS, 33.2 dB) (Figure 2, E). Laser flare photometry increased to 15.1 photons per millisecond OD and 5.1 photons per millisecond OS. Because of increased symptoms, decreased visual acuity, and marked disturbance of the visual fields, we initiated treatment consisting of oral prednisone, 1 mg/kg, and cyclosporine, 5 mg/kg. Five weeks after introduction of therapy, flickering in the right eye disappeared, visual acuity improved from 20/25 to 20/20 for far and from 20/30 to 20/25 for near, laser flare photometry diminished from 15.1 to 7.2 photons per millisecond, and indocyanine green angiographic signs improved (Figure 2, B and D) and remained stable thereafter. Visual fields improved bilaterally (MD OD, 9.5-3.5 dB; MD OS, 6.1-2.9 dB; LV OD, 55-16.5 dB; LV OS, 33.2-9.3 dB). Two and a half months later, the more severely affected right eye remained stable (visual acuity, 20/20; laser flare photometry, 7.2 photons per millisecond; visual field parameters: MD, 2.6 dB; and LV, 12.2 dB).

On the left side, however, far vision decreased to 20/50 due to central serous chorioretinopathy (CSC) that had a typical angiographic aspect on both fluorescein and ICG angiography. Accordingly, the visual field worsened slightly in the left eye because of the scotoma related to CSC with a stable MD (2.7 dB) and a slight increase in LV (10.5 dB) (Figure 2, E). As CSC was attributed to the corticosteroid therapy, the use of corticosteroids was rapidly decreased and acetazolamide, 500 mg twice daily, was introduced. Cyclosporine also had to be discontinued because of intolerable side effects (systemic hypertension and an increase of >30% in the serum creatinine level). After discontinuation of corticosteroid therapy, visual acuity again improved to 20/30 OS. Evidence of CSC by fluorescein angiography was no longer seen, but persistent staining with ICG remained. The fact that computerized visual fields contin-
ued to improve despite decreasing visual acuity in the left eye indicated that the visual acuity drop was due not to the primary disease process, but to secondary CSC. Six months after discontinuation of therapy, MD again increased from 2.6 to 3.9 dB (OD) and from 2.7 to 4.8 dB (OS) (not shown on graph).

**Comment.** Visual field testing has been neglected so far in the appraisal of BC. This case shows that visual field alterations can indeed be very extensive while central visual acuity is preserved. It also seems as if the visual field deteriorates before other factors and is more sensitive for follow-up. The striking feature here was not only the pronounced deterioration of visual fields, but also their marked improvement during treatment. The involved physiopathological mechanism has to be investigated. From the pattern of visual field alteration it is impossible to say whether the involvement is retinal rather than at the level of the optic disc. Diffuse and massive retinal impregnation by fluorescein and electrophysiologic findings point toward a retinal pathology. Systematic computerized visual field testing in BC is probably warranted and will enable us to gather more data to evaluate its value as a follow-up factor. Another striking feature in this case was the occurrence of CSC, which was most likely due to the corticosteroid therapy.

Christian de Courten, MD
Carl P. Herbst, MD, PhD
Lausanne, Switzerland

Corresponding author: Carl P. Herbst, MD, PhD, La Source Eye Center, 2 Avenue des Bergières, CH-1004 Lausanne, Switzerland.