Optic Neuritis with Various Manifestations of Visual Field

35

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Optic neuritis is a clinically common disease which has various clinical manifestations. There is no pattern to find in its visual field damage. Several types and outcomes of visual field defects in optic neuritis are reviewed through several cases and literature review in this section.

35.1 Case 1

35.1.1 Case Presentation

A 36-year-old male patient complained of sudden decrease in visual acuity in his right eye for more than 10 days with pain upon eye movement. But there was no red or sore eyes, metamorphopsia, etc. He had experienced a cold and fever 1 week before the onset of the vision change, which had resolved. Histories of trauma, other ocular diseases, systemic diseases, or familial diseases were denied.

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In the right eye, the uncorrected visual acuity (UCVA) was finger counting, and the best corrected visual acuity (BCVA) was 0.05 with refractive correction (−2.00DS − 0.75DC*180), the light projection was accurate, and red and green colors could be distinguished. In the left eye, the UCVA was 20/200, and the BCVA was 20/20 with refractive correction (−2.50DS−1.00DC*85). Intraocular pressure (IOP) was normal OU. Slit-lamp examination of the anterior segments was unremarkable except that the relative afferent pupillary defect (RAPD) was positive in the right eye. In the right eye, the optic disc exhibited an unclear margin, with congestion and edema. There was no abnormality in the left fundus (Fig. 35.1).

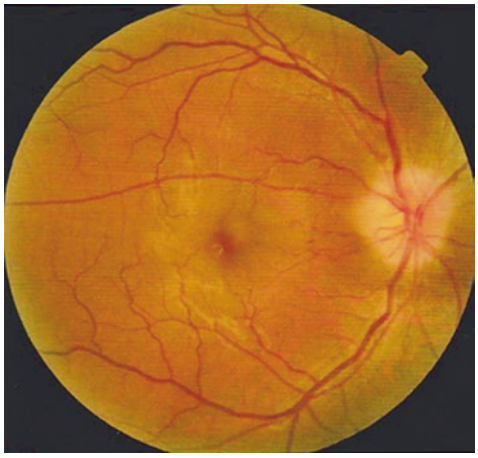

Fig. 35.1 Fundus photograph. The optic disc exhibited an unclear margin, with congestion and edema

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Standardized automated perimetry showed a severe damage to the central visual field in the right eye with residual nasal visual field only (Fig. 35.2).

The F-VEP test showed abnormal waveform and decreased amplitude of the P2 wave under the stimulation at 1.2 Hz and normal waveform, normal latency, and decreased amplitude of the P2 wave under the stimulation at 12 Hz in the right eye, while there was no abnormality in the left eye (Fig. 35.3).

F-VEP showed abnormal waveform and decreased amplitude of the P2 wave under the stimulation at 1.2 Hz and normal waveform, normal latency, and decreased amplitude of the P2 wave under the stimulation at 12 Hz in the right

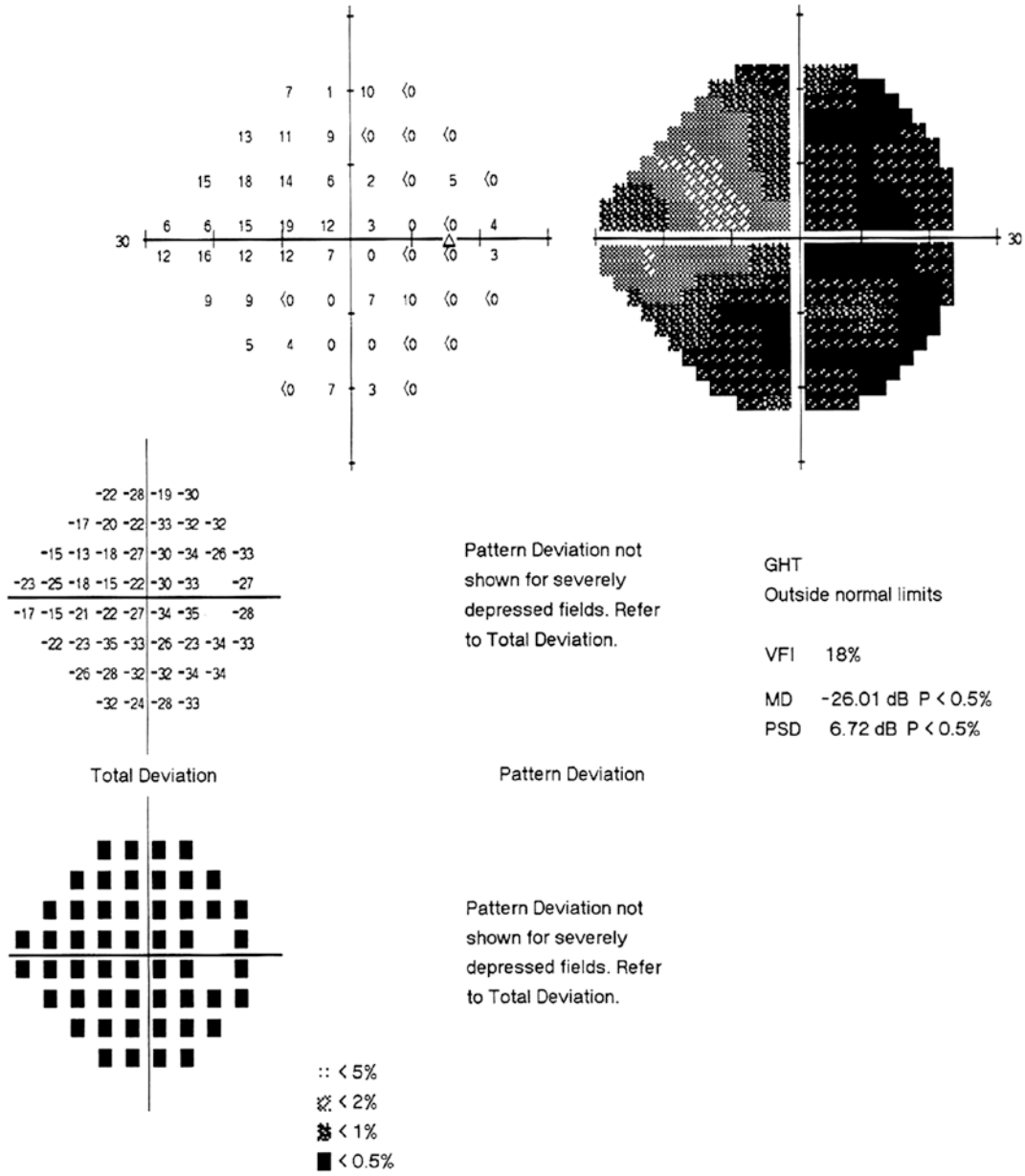
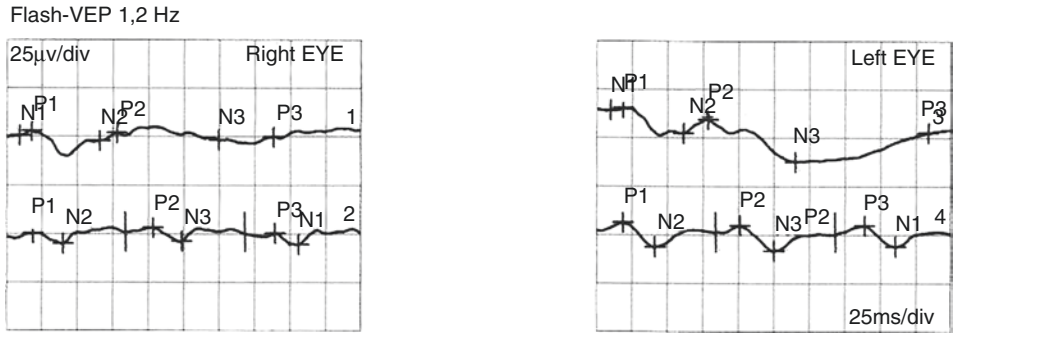


Fig. 35.2 Humphrey visual field analysis printout. The 24-2 test showed that there was only the nasal visual field remaining in the right eye, and the photosensitivity decreased diffusively and dramatically



Channel	N1 [ms]	P1 [ms]	N2 [ms]	P2 [ms]	N3 [ms]	P3 [ms]	N1-P1	N2-P2	N3-P3
1 R-1 1,2 Hz	9	18	66	78	150	189	2.24µV	4.13µv	4.47µv
3 L-1 1,2 Hz	10	19	62	79	140	234	1.17µv	6.97µv	4.7µv
2 R-1 12 Hz	206	19	40	104	124	190	5.94µv	8.39µv	1.93µv
4 L-1 12 Hz	211	19	41	101	125	189	12.4µv	11.1µv	1.2µv

Normals	-	-	-	-	-	-	-	-	-

Channel	Stimulus	Ampl., Range, Filter
1 R-1 1,2 Hz	GF LED Flash 0dB (2,00 cds/m? 1.199Hz, Avg:100	1, +/-100µv 0.5-50Hz
2 R-1 12 Hz	GF LED Flash 0dB (2,00 cds/m? 11.905Hz, Avg:100	1, +/-100µv 0.5-50Hz
3 L-1 1,2 Hz	GF LED Flash 0dB (2,00 cds/m? 1.199Hz, Avg:100	1, +/-100µv 0.5-50Hz
4 L-1 12 Hz	GF LED Flash 0dB (2,00 cds/m? 11.905Hz, Avg:100	1, +/-100µv 0.5-50Hz

Fig. 35.3 F-VEP examination printouts

eye, while in the left eye, the waveform, latency, and amplitude were normal under the stimulation at both 1.2 and 12 Hz (Fig. 35.3).

FFA showed telangiectasia in the optic disc and in the top layer around the optic disc at the early phase, fluorescein leakage at the venous phase, and strong fluorescence in the optic disc at the late phase in the right eye (Fig. 35.4).

35.1.2 Final Diagnosis

The final diagnosis was papillitis in the right eye.

The patient was given methylprednisolone pulse therapy and oral hormones at sequentially reduced doses, which was supplemented with supportive therapies such as vitamin B and blood circulation improvement for 10 days. At the follow-up examination, the best corrected visual acuity (BCVA) was 1.2. RAPD was negative. The optic disc edema was resolved, and the standardized automated perimetry results were unremarkable in the right eye (Fig. 35.5). The F-VEP results showed that the waveform, latency, and amplitude of the P2 wave were all normal in the right eye. The cranial MRI revealed no abnormalities.

35.1.3 Case Review

Papillitis is an inflammatory disease that occurs in the optic papilla and its surrounding areas. It is a primary demyelinating optic neuritis and is closely related to central nervous system demyelinating diseases, such as multiple sclerosis or neuromyelitis optica. In most of the typical cases, the patients are unilaterally affected young adults, and the most common manifestation is visual field damage at the central 20°. VEP examination will show that the conduction velocity of nerve potential slows down due to the demyelination of nerve fibers, and then the latency is prolonged. When the axons of the nerve fibers are damaged, the conduction velocity may be normal, while the potential intensity is weakened, resulting in a decrease in amplitude. FFA will reveal capillary dilatation on the optic disc surface and fluorescein leakage on the vascular wall. In the late phase of radiography, the strong fluorescence of the whole optic disc and its surrounding tissues can be seen. The patient's visual field test, VEP, and FFA results matched the typical manifestations of optic nerve papillitis.

Currently, adrenal glucocorticoid is the main treatment for acute demyelinating optic neuritis. The optic neuritis treatment trial (ONTT) con-

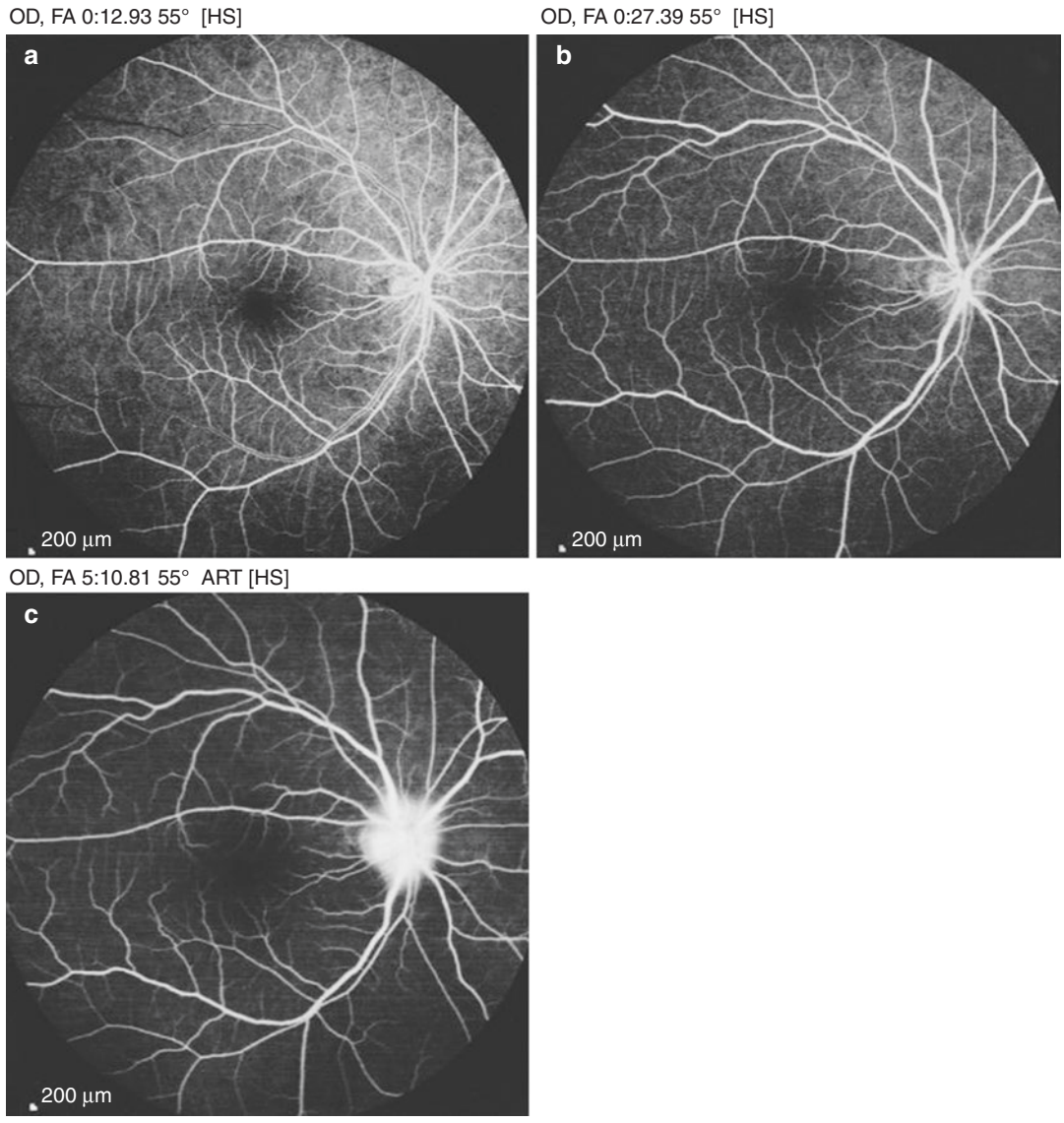


Fig. 35.4 FFA images. Panel a: telangiectasia in the optic disc and in the top layer around the optic disc at the early phase of angiography. Panel b: fluorescein leakage at the venous phase. Panel c: strong fluorescence of the optic disc at the late phase

firmed that pulse therapy with intravenous administration of high doses of methylprednisolone can obviously accelerate the recovery of visual function in these patients [1]. The patient in this case was treated according to the expert guidelines that were prepared by a neuro-ophthalmology group in China. After 10 days, the vision and visual field basically normalized without recurrence after reducing the dose of hormone sequentially.

For these patients, the cranial MRI examination is necessary, and clinical symptoms and necessary auxiliary examinations should also be considered to assist in determining whether there is any high-risk factor for progression into multiple sclerosis and neuromyelitis optica, especially in patients with recurrent papillitis and patients who are nonresponsive to hormone therapy.

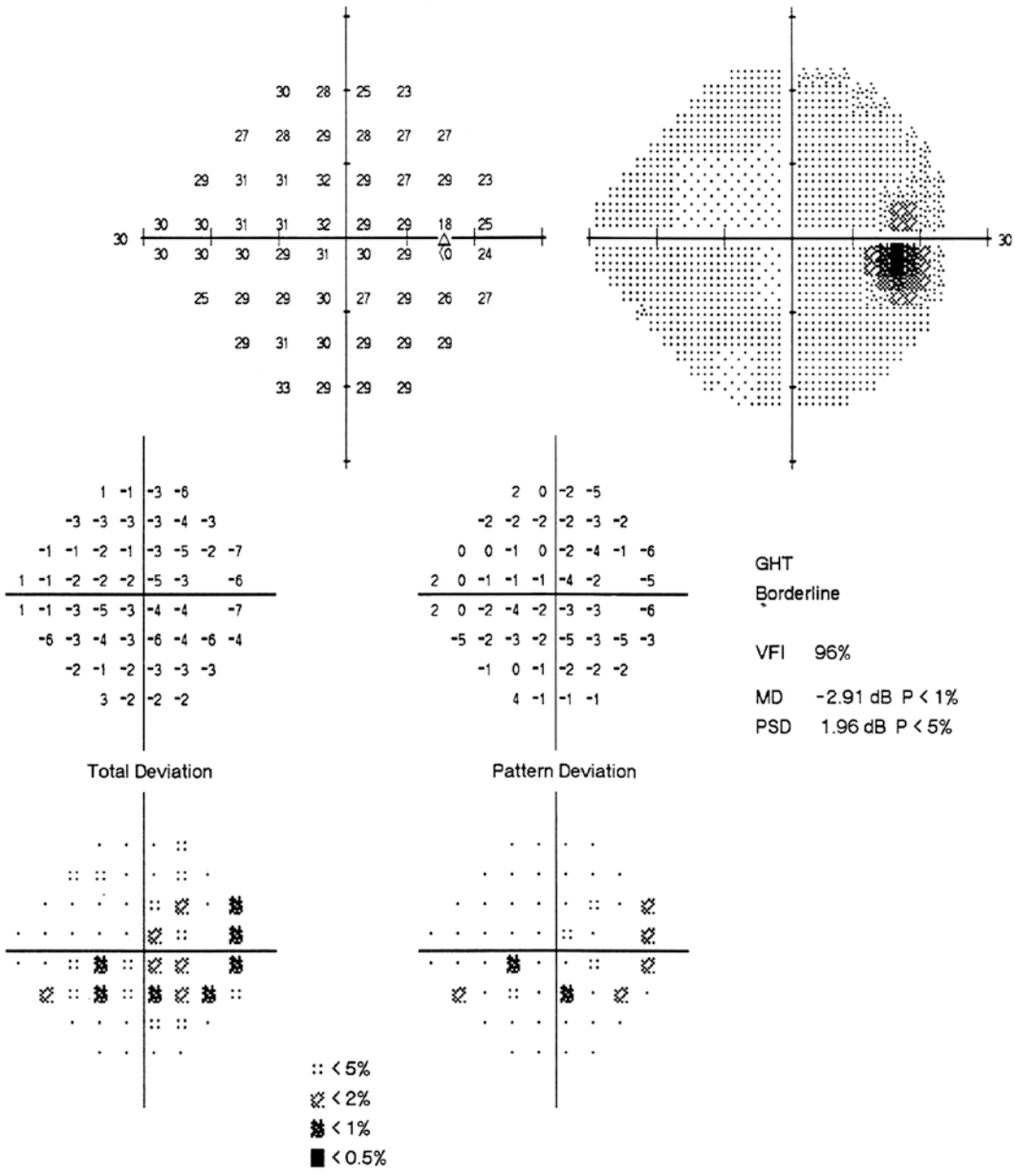


Fig. 35.5 Re-examination Humphrey visual field analysis printout obtained during the re-examination after treatment. The 24-2 test revealed a normal central field in the right eye

35.2 Case 2

35.2.1 Case Presentation

A 34-year-old male patient complained of feeling a shadow over the right eye accompanied by pain upon eye movement for 3 days. The patient did not feel any malaise. The patient did not have sore or red eyes, metamorphopsia, or other discomforts. There was no history of trauma, other ocular diseases, or systemic or familial diseases.

The UCVA was 1.2 OU. IOP was normal OU. Slit-lamp examination of his anterior segments was unremarkable. The RAPD was negative in the right eye. Fundus examination showed that the right optic disc was with an unclear boundary, congestion, and edema.

Standardized automated perimetry revealed nasal visual field defects in the right eye (Fig. 35.6).

The OCT results showed that the retinal nerve fiber layer (RNFL) thickness in the superior, inferior, and temporal quadrants increased significantly (Fig. 35.7).

FFA showed telangiectasia in the optic disc and the peripapillary surface layer at the early phase, fluorescein leakage at the venous phase, and obviously enhanced fluorescence of the optic

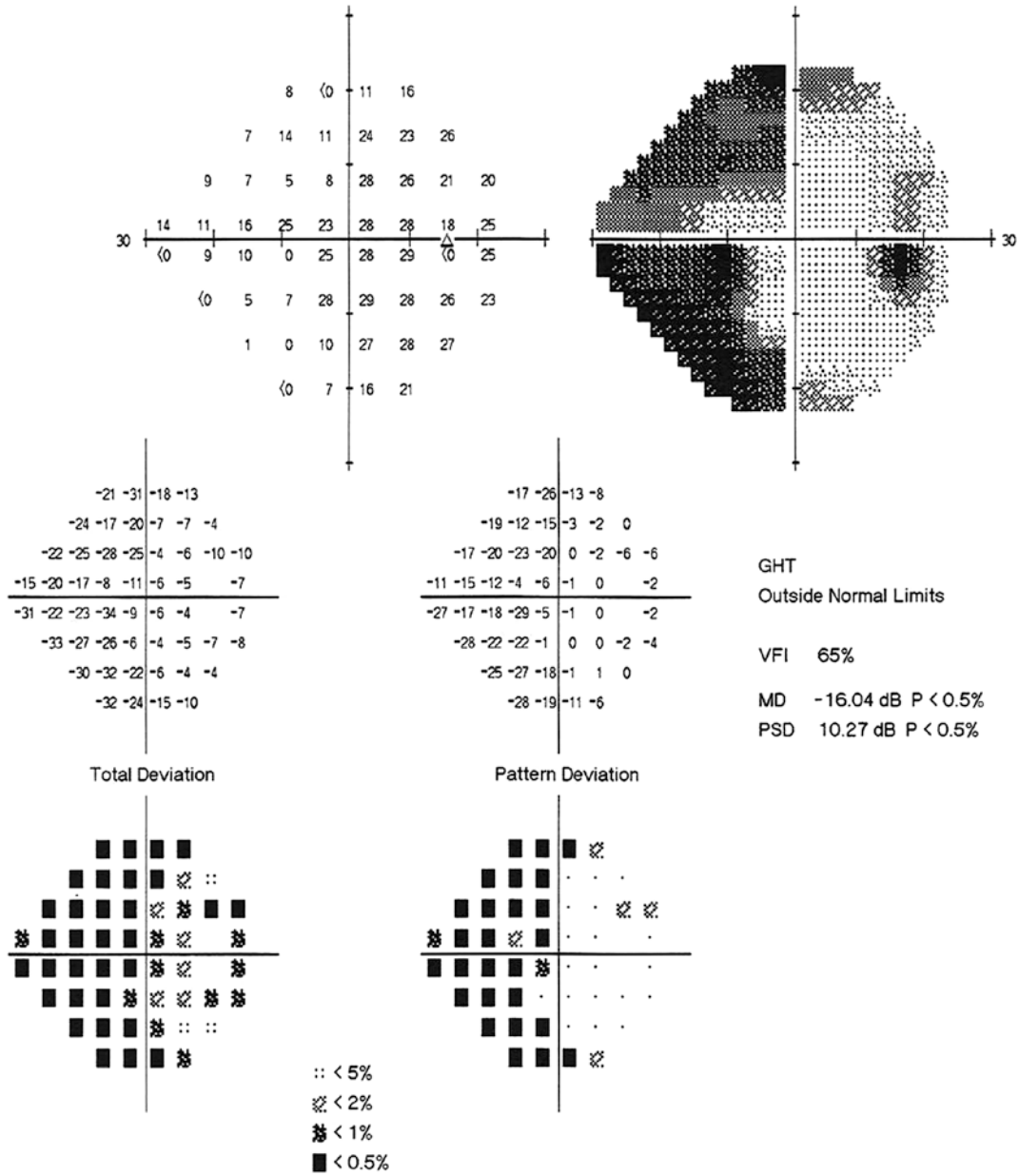


Fig. 35.6 Humphrey visual field analysis printout. The 24-2 test showed nasal visual field defects in the right eye

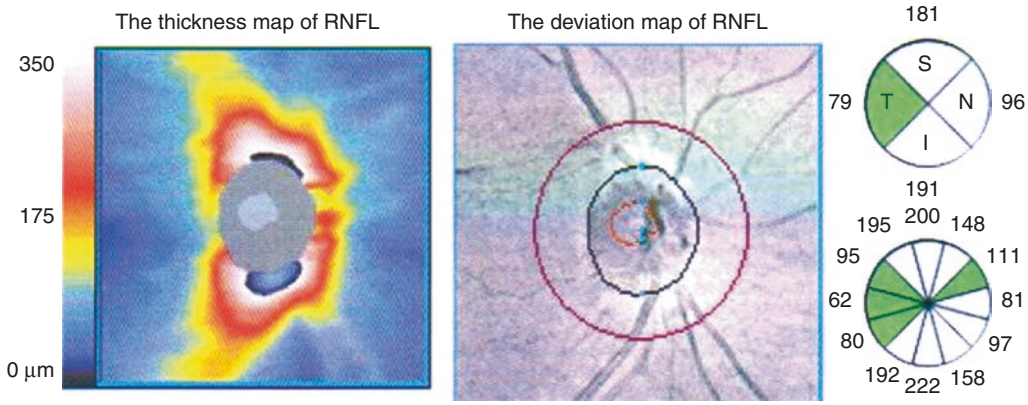


Fig. 35.7 OCT RNFL thickness analysis printout. The RNFL thickness in the superior, inferior, and temporal quadrants increased significantly

disc at the late phase in the right eye, but its disc boundary was still clear (Fig. 35.8).

The P-VEP results showed normal latency and a moderate decrease in the amplitude of the P100 wave in the right eye, and the P-VEP wave was normal in the left eye.

35.2.2 Final Diagnosis

The final diagnosis was papillitis in the right eye.

The patient was given methylprednisolone pulse therapy, oral hormones at sequentially reduced doses, and other supportive treatments including vitamin B and improvement of blood circulation. After 10 days, the follow-up examination showed a visual acuity of 20/20 OD, and the standardized automated perimetry revealed normal results (Fig. 35.9). No abnormalities were found during the head MRI examination.

35.2.3 Case Review

The patient was diagnosed with papillitis in the right eye. The visual field showed a nasal defect, which was consistent with the edematous RNFL areas (inferior, superior, and temporal quadrants) demonstrated by OCT. Considering the optic neuritis was localized around the optic disc, the local visual field damage was consistent, functionally and structurally, with the parapapillary RNFL thickness measured by OCT.

The amplitude of P-VEP in the right eye decreased significantly, and the latency was normal, suggesting that the lesion was mainly axonal injury of nerve fibers resulting in the reduction of the number of excitable axons. The normal latency indicated that the conduction function of myelin was normal, which suggested that demyelination was relatively mild.

35.3 Case 3

35.3.1 Case Presentation

A 25-year-old female patient complained recurrent vision loss in her left eye for more than 1 month. It had occurred without any obvious trigger. The patient had been treated with methylprednisolone, dexamethasone, prednisone, and other hormones in other hospitals. In the left eye, the visual acuity had been light perception at its worst, which was improved to 20/20 with refraction after treatment. However, the visual acuity in his left eye had decreased again 1 week before. He had no history of trauma, other ocular diseases, or systemic or familial diseases. His right eye was normal.

On examination, the UCVA was 20/20 OD and 20/28 OS, and there was no improvement with refraction. The IOP was normal. In both eyes, slit-lamp examination of anterior seg-

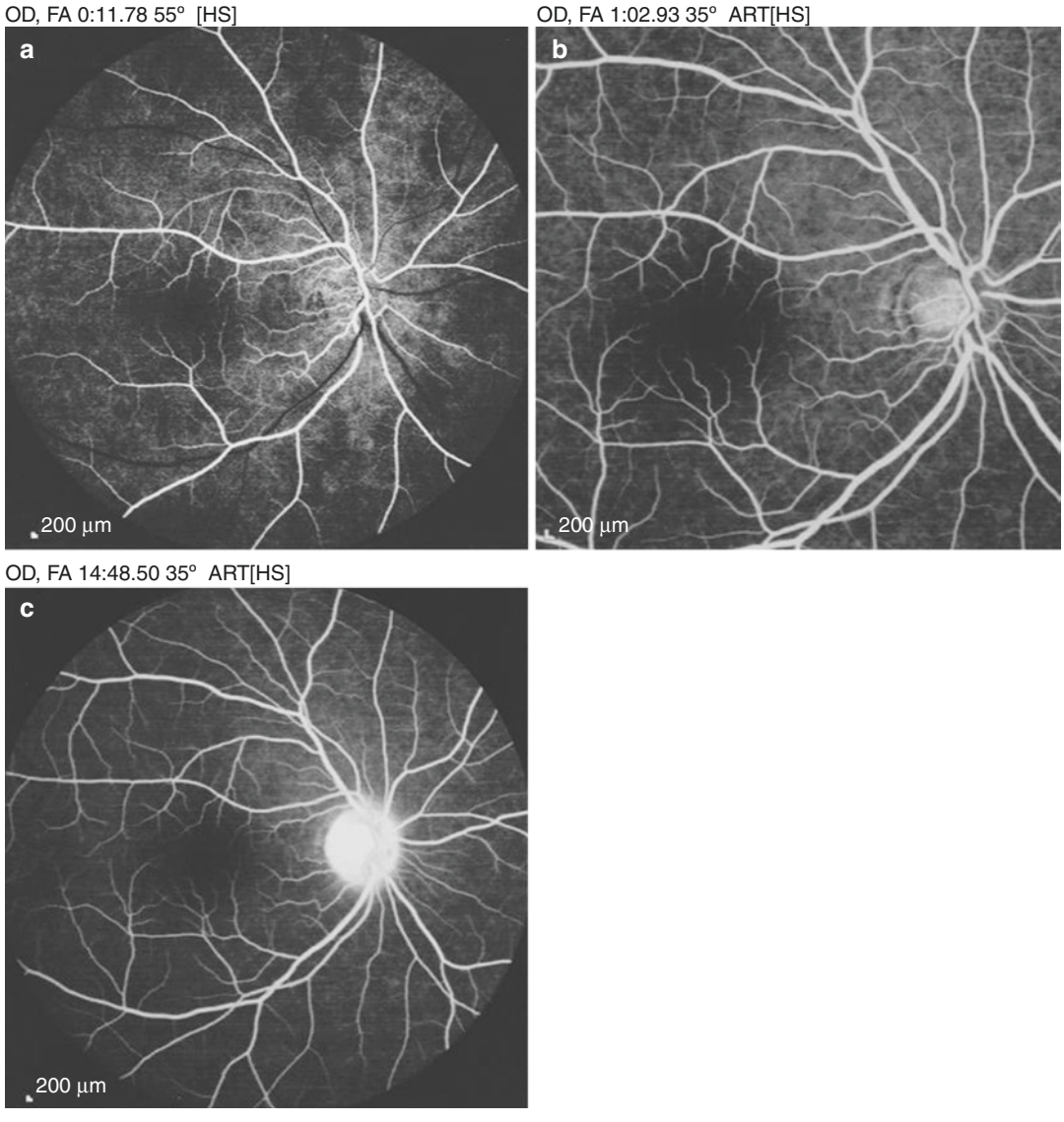


Fig. 35.8 FFA images. FFA showed telangiectasia in the optic disc and the peripapillary surface layer at the early phase, fluorescein leakage at the venous phase, and obviously enhanced fluorescence of the optic disc at the late phase in right eye, but its disc boundary was still clear. Panel a: the early-phase angiogram. Panel b: the venous-phase angiogram. Panel c: the late-phase angiogram

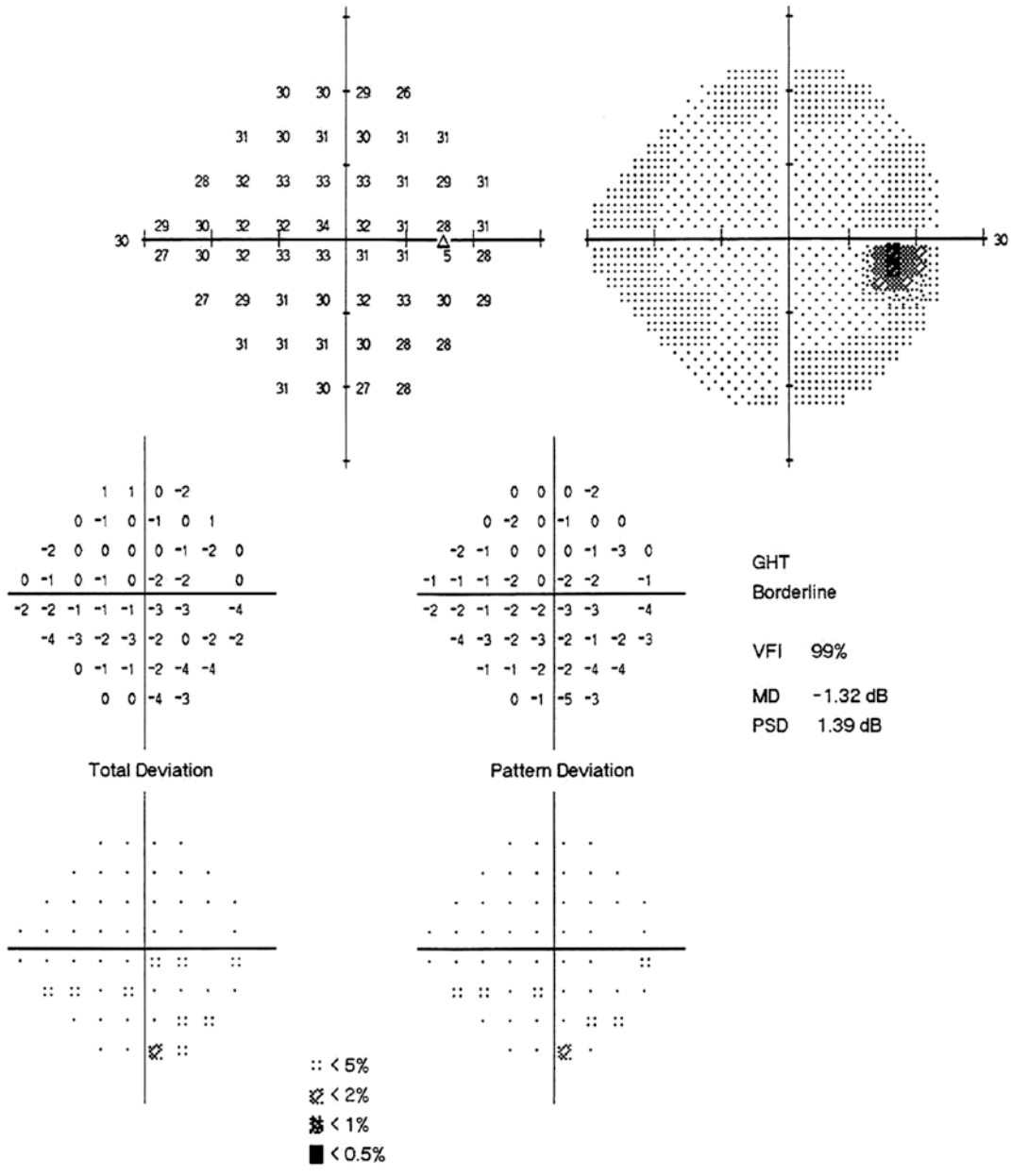


Fig. 35.9 Humphrey visual field analysis printout obtained during the re-examination after treatment. The 24-2 test showed normal results

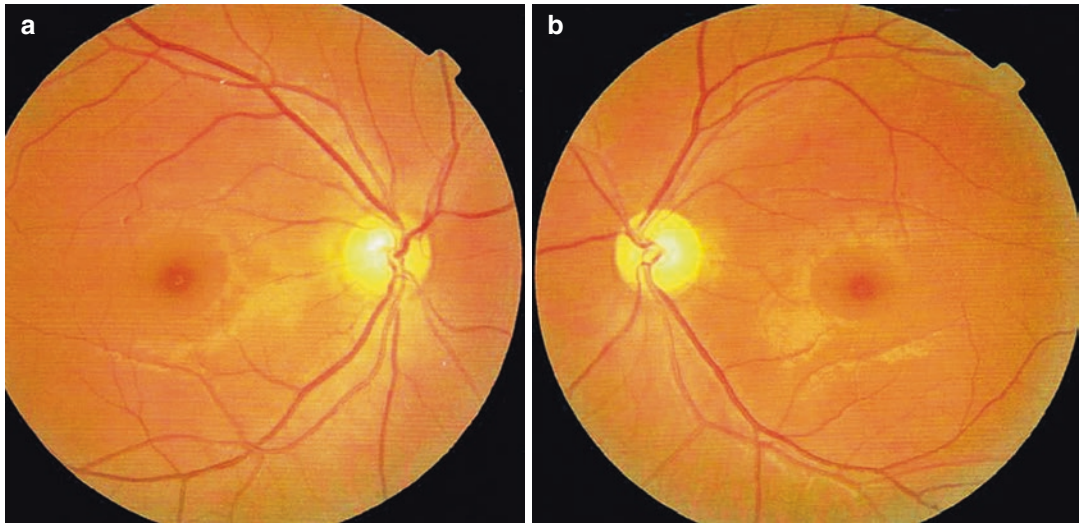


Fig. 35.10 Fundus photographs. Panel a: the right optic disc presented a clear boundary and a pink color, and the reflection of the RNFL was normal. Panel b: the left optic disc presented a clear boundary and a pale color, and the reflection of the RNFL was weakened

ments was unremarkable except that the RAPD was positive in the left eye. Fundus examination showed the left optic disc was pale in color with a clear boundary and a cup-to-disc (C/D) ratio of 0.5, and the reflection of RNFL was weakened. The right optic disc was pink in color with a clear boundary and a C/D ratio of 0.5, and the reflection of RNFL was normal (Fig. 35.10).

Standardized automated perimetry showed an irregular central defect in the left eye (Fig. 35.11).

The P-VEP results showed the waveform, latency, and amplitude of the P100 wave were severely abnormal in the left eye, while the P100 wave was normal in the right eye (Fig. 35.12).

FFA revealed normal findings in the left eye (Fig. 35.13).

35.3.2 Final Diagnosis

The final diagnosis was recurrent retrobulbar neuritis in the left eye.

35.3.3 Case Review

A central visual field defect is typical in optic neuritis, and it can appear in any phases of the course, such as the acute phase and recovery phase. At about 15 mm behind the eyeball or beyond, the core of the optic nerve is mainly macular fibers. Therefore, in case of retrobulbar neuritis, significant damage to macular fibers occurs first. As a result, a central defect appears in the visual field, similar to the visual field changes in patients with neuritis.

In this case, due to suboptimal hormone therapy after the onset, the condition recurred and deteriorated in a short period with thinning of retinal nerve fibers, suggesting optic atrophy. According to the natural course of idiopathic demyelinating optic neuritis, the visual function begins to recover 3–5 weeks after onset. When the visual acuity and visual symptoms improve at 4–6 weeks, the damaged nerve fibers begin to become thinner and the color of the optic disc paler.

35.4 Case 4

35.4.1 Case Presentation

A 34-year-old male patient complained of bulging pain for 1 week and decreased vision for 3 days in his right eye. The onset of disease had no

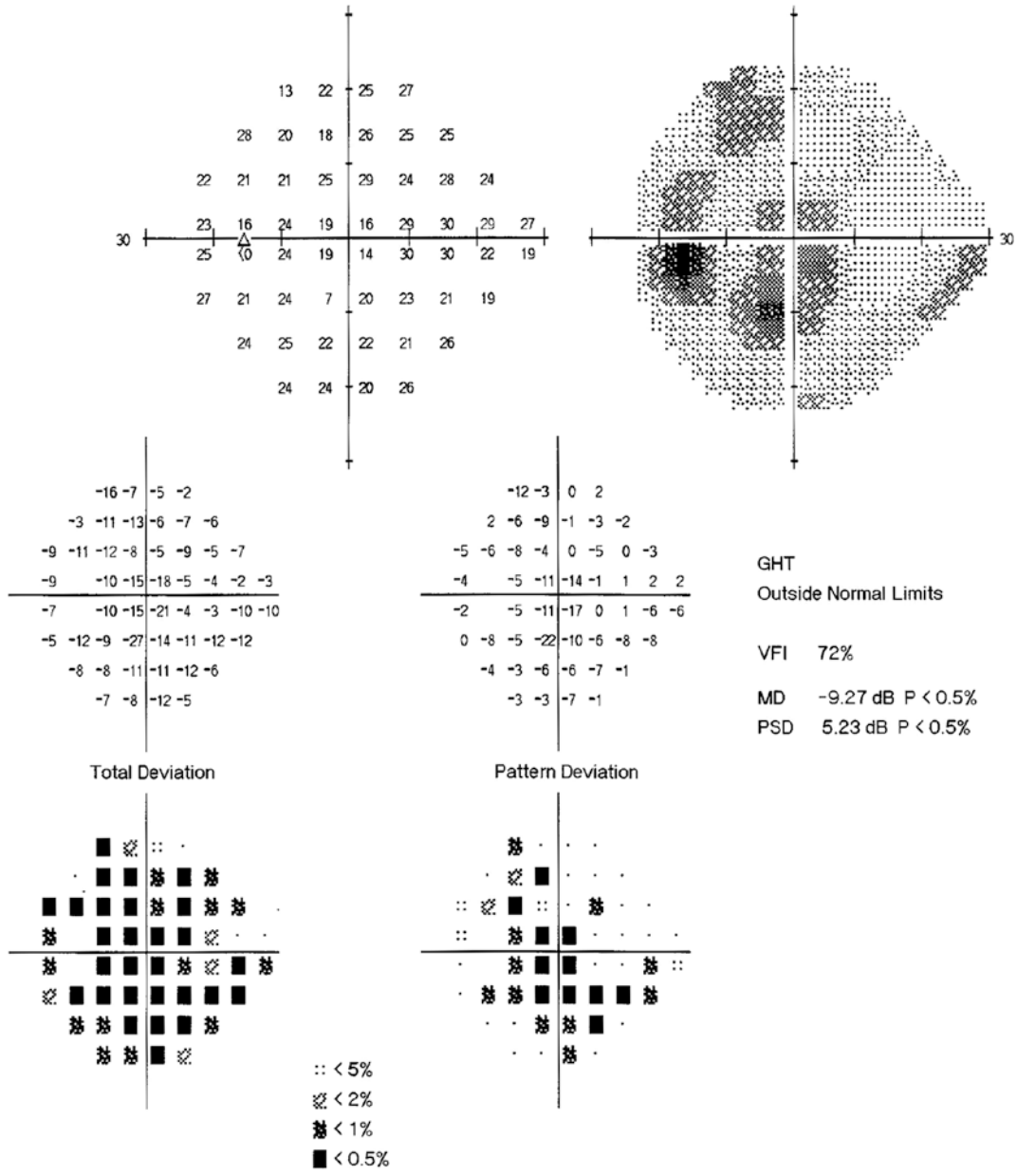
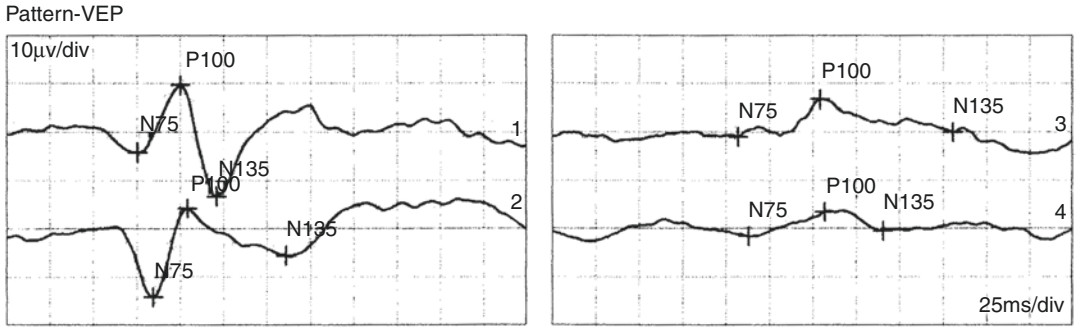


Fig. 35.11 Humphrey visual field analysis printout. The 24-2 test showed an irregular central defect in the left eye

obvious trigger. The patient denied any ocular discomfort. Histories of trauma, other ocular diseases, or systemic or familial diseases were denied.

The UCVA was 20/22 in the right eye, and it was not improved with manifest refraction. IOP was normal. Slit-lamp examination of his anterior segments was unremarkable except that the RAPD was positive in the right eye. Fundus examination revealed that the right optic disc had a clear boundary and was pale in color, with a C/D ratio of 0.6 (Fig. 35.14). No abnormality was found in the left eye.

Standardized automated perimetry showed quadrantanopia in the superior area of the right eye (Fig. 35.15).



Channel	N75 [ms]	P100 [ms]	N135 [ms]	N75-P100	P100-N135
1 R-1 1,0	76	100	122	13.8μv	22.9μv
3 L-1 1,0	107	154 (!)	231	7.65μv	6.7μv
2 R-1 0,15	85	105	161	18.3μv	9.73μv
4 L-1 0,15	113	157 (!)	191	5.11μv	3.99μv
Normals	-	90-116-90-116	-	-	-

Channel	Stimulus	Ampl., Range, Filter
1 R-1 1,0	MON Patt. Rev. CB, 1?Full Field, Ctr:97% 1.5Hz, Avg:96	1, +/-100μv 1-50Hz
2 R-1 0,15	MON Patt. Rev. CB, 0?5' Full Field, Ctr:97% 1.5Hz, Avg:98	1, +/-100μv 1-50Hz
3 L-1 1,0	MON Patt. Rev. CB, 1?Full Field, Ctr:97% 1.5Hz, Avg:85	1, +/-100μv 1-50Hz
4 L-1 0,15	MON Patt. Rev. CB, 0?5' Full Field, Ctr:97% 1.5Hz, Avg:87	1, +/-100μv 1-50Hz

Fig. 35.12 P-VEP examination printouts. The waveform, latency, and amplitude of the P100 wave in the left eye were severely abnormal, and these are normal in the right eye

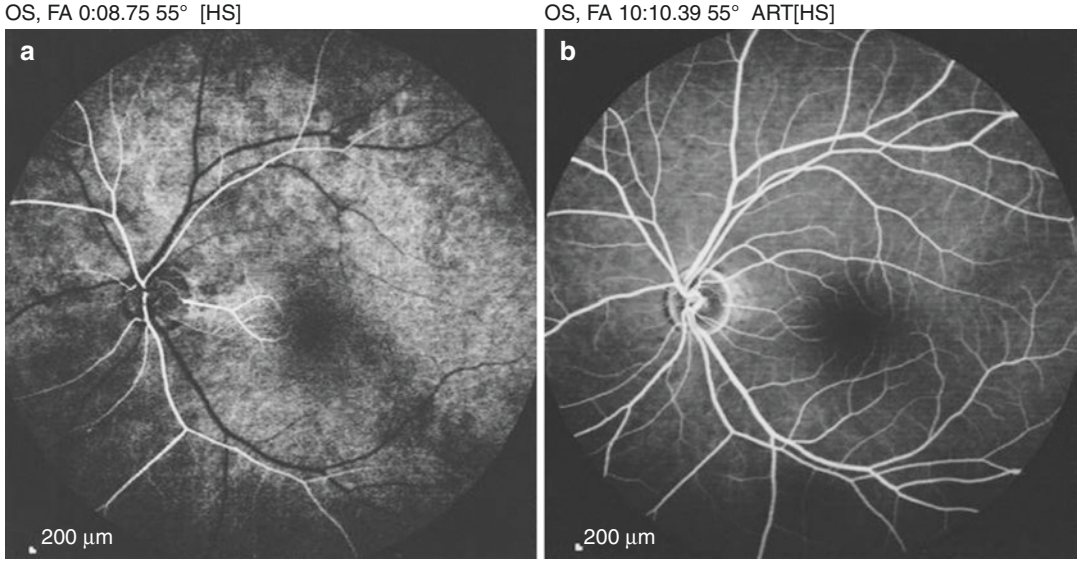


Fig. 35.13 FFA images. Panel a: FFA showed that the cilioretinal artery was filled with fluorescence at the early phase of angiography. Panel b: the FFA findings at the late phase of angiography were normal

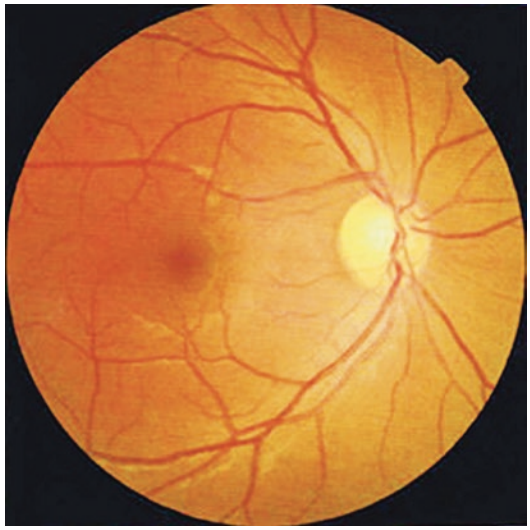


Fig. 35.14 Fundus photograph. The right fundus examination showed that the optic disc had a clear boundary and was pale in color with a C/D of 0.6

P-VEP showed that both the latency and amplitude of the P100 wave in the right eye were severely abnormal, and the P100 wave was normal in the left eye.

The FFA findings were normal in both eyes.

No abnormalities were found in the cranial MRI examination.

35.4.2 Final Diagnosis

The final diagnosis was retrobulbar neuritis in the right eye.

After 10 days of medical treatment, the follow-up examination showed that the visual acuity of the right eye was 20/20, and the visual field was normal (Fig. 35.16).

After 1 month of drug therapy, the patient was lost to follow-up and did not take medications as prescribed. The bulging pain and pain on eye movement in the right eye recurred after more than 2 months. The patient came back to the clinic 5 days later.

Eye examination showed the UCVA was 20/20 with no improvement with refraction in the right eye. IOP was normal. Slit-lamp examination of his anterior segments was unremarkable except that the RAPD was positive in the right eye. Fundus examination showed that the right optic disc had a clear boundary and was pale in color with a C/D ratio of 0.6. There was no abnormality in the macula. The findings in the left eye were normal.

Standardized automated perimetry showed an inferior arcuate scotoma in the right eye (Fig. 35.17).

P-VEP showed that both the latency and amplitude of the P100 wave were severely abnormal in the right eye, and the P100 wave was normal in the left eye.

FFA showed that no abnormalities were found in either eye.

35.4.3 Final Diagnosis

The final diagnosis was recurrent retrobulbar neuritis in the right eye.

The patient received medical treatment again. At the 10-day follow-up examination, a visual acuity of 20/20 and a recovering normal visual field were shown in the right eye (Fig. 35.18). The patient was discharged with medications and was asked to undergo regular examinations and take medicine as prescribed. During the 3-year follow-up, there was no recurrence of optic nerve disease.

35.4.4 Case Review

In this case, retrobulbar neuritis occurred two times within several months, which was mani-

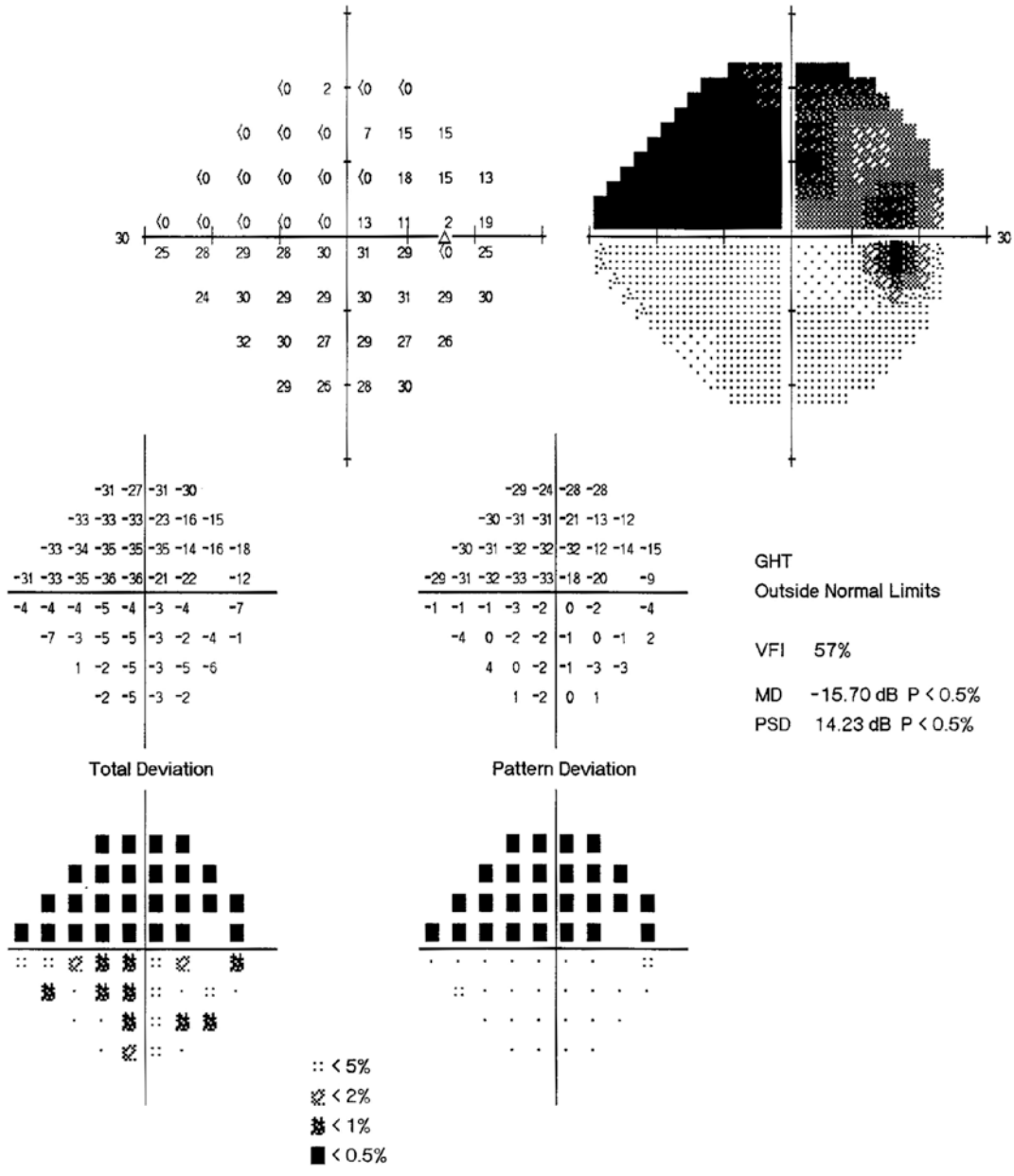


Fig. 35.15 Humphrey visual field analysis printout. The 24-2 test revealed quadrantanopia in the superior area

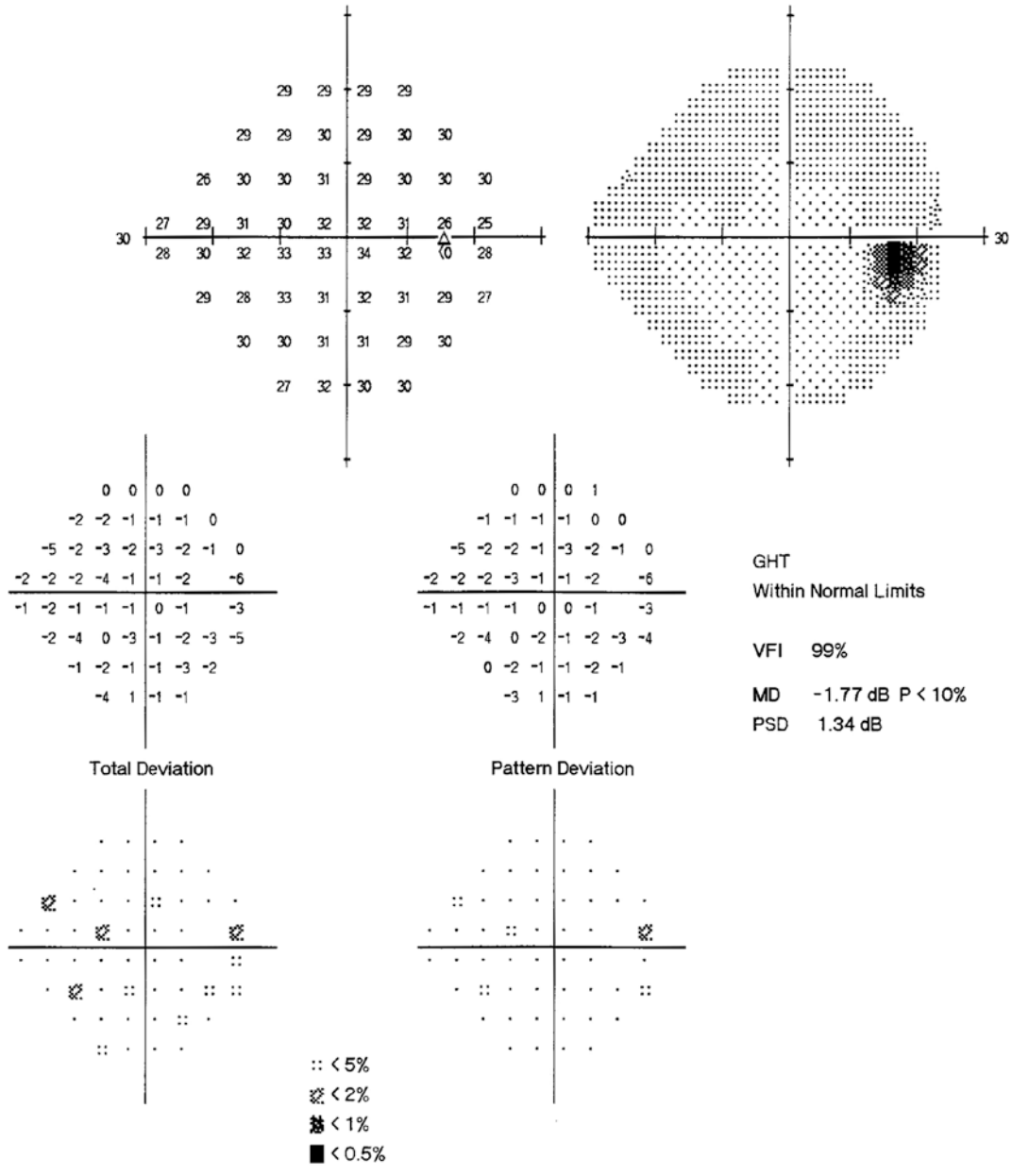


Fig. 35.16 Humphrey visual field analysis printout obtained during the re-examination after treatment. The 24-2 test showed normal results

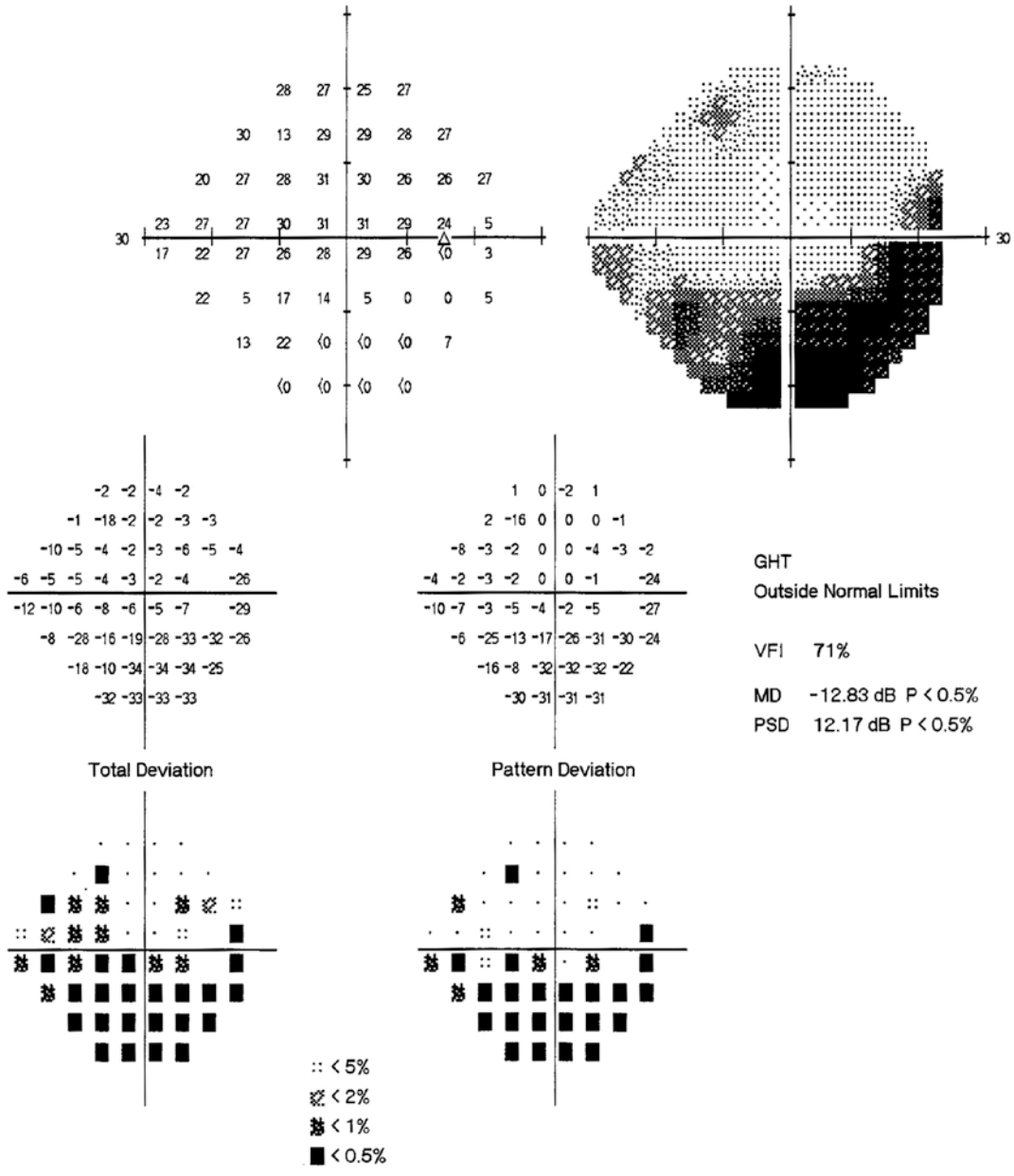


Fig. 35.17 Humphrey visual field analysis printout. The 24-2 test showed an inferior arcuate scotoma in the right eye

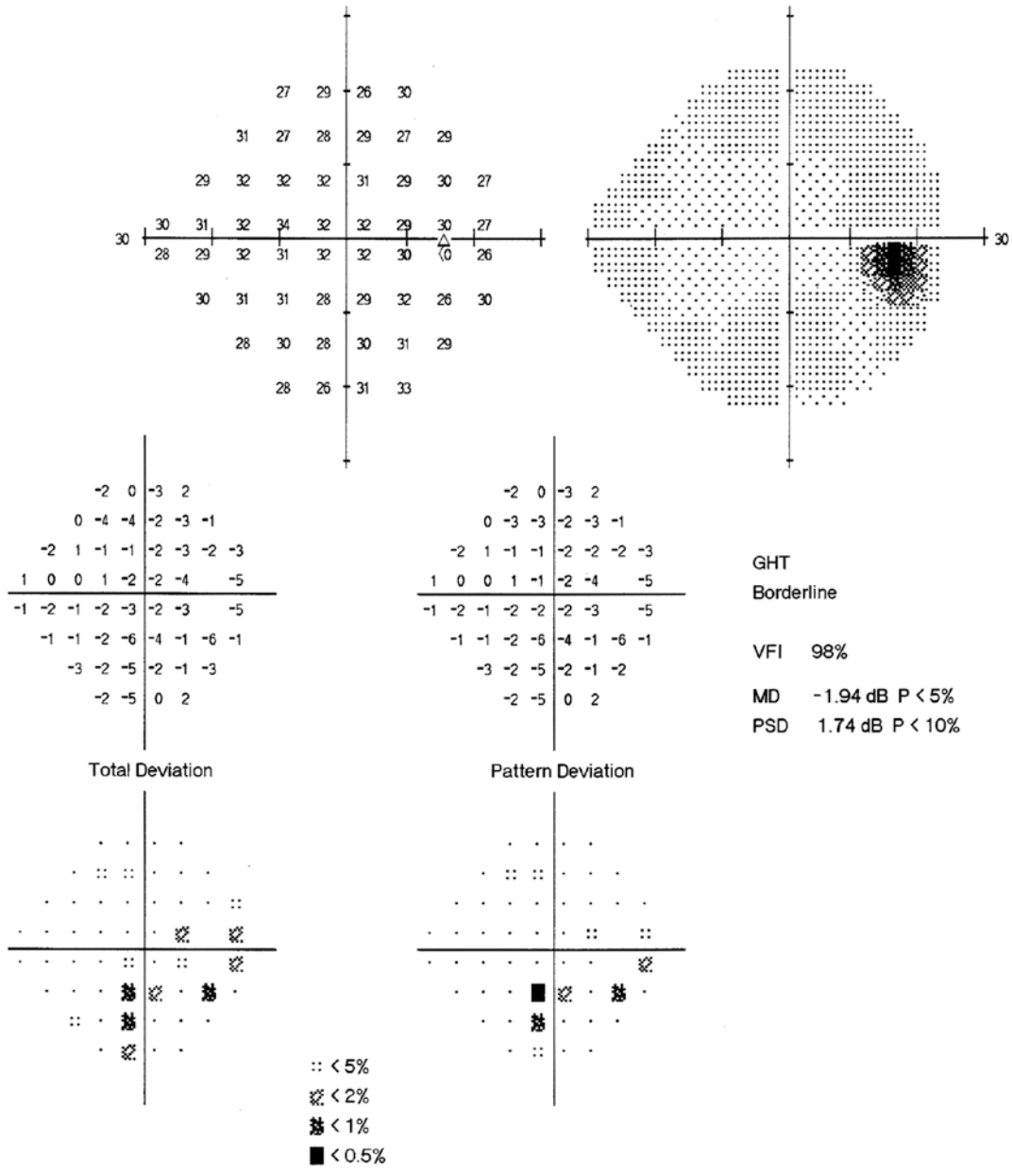


Fig. 35.18 Humphrey visual field analysis printout obtained during the re-examination after treatment. The 24-2 test showed a recovering normal result in the right eye

fested as a superior visual field defect and an inferior arcuate scotoma, respectively, further reflecting the variety of visual field changes in optic neuritis. In addition, it was recognized that standardized automated perimetry is not specific for diagnosing optic neuritis. In terms of morphology, the changes in the visual field in optic neuritis depend on the areas involved in inflammation and the composition of nerve fibers in these areas.

35.5 Discussion

The papillitis and retrobulbar neuritis discussed in this section are idiopathic demyelinating optic neuritis (IDON) that is the most common type of optic neuritis. This name is based on the previous knowledge that the disease is closely related to demyelinating diseases of the central nervous system, such as multiple sclerosis (MS) and neuromyelitis optica (NMO, also known as Devic disease). The clinical diagnosis is divided into papillitis, retrobulbar neuritis, optic perineuritis, and neuroretinitis according to the pathogenic site. Other types of optic nerve inflammation, including infectious optic neuritis, autoimmune optic neuropathy, and other optic neuropathy, like ischemic, genetic, compressing, nutritional and metabolic optic neuropathy, should be ruled out. According to the latest consensus on the diagnosis and treatment of optic neuritis, IDON may be MS-related optic neuritis. The latter is characterized by onset at multiple times and locations. The demyelination in NMO and its related optic neuritis is not exactly the same as that in MS, and the prognosis of NMO is also poorer [1, 2].

Of the 415 patients with defects at the central 30° of the visual field in the ONTT research, 48% were with diffusive defects, while 52% were with focal defects. In these patients with focal defects, 20% had nerve fiber bundle-induced focal visual field defects (including vertical, arcuate, and nasal step defects), only 8% had a central scotoma, and 5% had hemianopia [2].

The pattern of visual field damage in optic neuritis is rooted in the part of the retina corresponding to the nerve fibers involved. In the anterior optic nerve, because the central retinal arteries and veins occupy the center of the optic nerve, the fibers from the macula are squeezed in the lateral upper and lower position of the optic nerves, the retinal fibers from the superior and inferior nasal retina are located in the medial superior and inferior positions of the optic nerves, and the superior and inferior temporal fibers are located in the lateral superior and inferior positions of the optic nerves. In the optic nerve 15 mm behind the eyeballs or beyond, due to no passage of retinal vessels any more, macular fibers gradually shift to the axis part of the optic nerve, fibers from the temporal retina shift to the temporal part of the optic nerve, fibers from the nasal retina shift to the nasal part of the optic nerve, and a similar fashion applies to the fibers from the superior and inferior retina. Therefore, the location of the optic neuritis and the involved retinal nerve fibers determine the corresponding visual field changes. In the optic nerve 15 mm behind the eyeballs or beyond, macular fibers occupy about 1/4 of the core space, so the visual field damage with a central defect is relatively common, but other types of defect may also be found [3, 4].

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