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CASE REPORT

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Foster-Kennedy syndrome – a case report

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ABSTRACT
A 77-year-old female patient who had been treated for glaucoma for many years was referred to an outpatient clinic to extend the diagnostic work-up because of rapidly deteriorating vision despite anti-glaucoma treatment. During the visit, in addition to characteristic ophthalmic symptoms, behavioral and orientation disturbances were found, which raised a suspicion of Foster Kennedy syndrome and prompted the examining physician to refer the patient for an urgent brain scan. A computed tomography examination revealed the presence of focal lesions. Rapid diagnosis enabled immediate transfer of the patient to a neurosurgical center for further treatment.
KEY WORDS: optic disc edema, optic nerve atrophy, meningioma, behavioral disorders, Foster Kennedy syndrome, glaucoma, masquerading syndrome.

CASE REPORT

A 77-year-old female patient, under the care of an out-of-hospital ophthalmology clinic because of glaucoma of approximately 15 years' duration, presented to a hospital-based outpatient clinic to extend the diagnostic work-up following a significant progressive deterioration of vision in the right eye.

A week earlier, the patient was consulted by an ophthalmologist at an out-of-hospital ophthalmology clinic. Visual acuity examination revealed V_{OD} 0.3 and V_{OS} 0.63 (using the patient's own spectacle correction). Intraocular pressure measured with a Goldmann applanation tonometer was T_{OD} = 18 mmHg and T_{OS} = 16 mmHg. Ophthalmoscopically, the optical nerve discs were found to be pale pink in color, with clearly defined borders. Until that time, the patient had used dorzolamide with timolol twice a day in both eyes. Her ophthalmic history revealed cataract surgery in the right and left eyes, and trabeculectomy in the left eye. During the patient's ophthalmic appointment several months prior to the reported one, she was referred for additional examinations, including an ultrasound of the carotid arteries, because of a decline in visual acuity in the right eye (from 0.9 to 0.5). However, the patient did not have these assessments done, nor did she come for a follow-up visit, despite the fact that her vision deteriorated further.

It must be noted that the patient's previous treatment of glaucoma, spanning a dozen years or so, was uncomplicated, and the condition was well controlled for a long time despite the presence of stable defects in vision in subsequent visual field tests (Figures 1-4).

Considering the patient's ophthalmic history outlined above, and the decline in visual acuity, it was decided to refer her for further diagnosis and treatment at the hospital-based outpatient clinic, where further evaluations were scheduled, including a repeat visual field test. When the patient came for the additional assessments a few days later, a further decrease in visual acuity and edema of the optic nerve disc in the right eye were found. Consequently, she was urgently admitted to the hospital's ophthalmology department.

Upon hospital admission, the following parameters were recorded:

V_{OD}: counting fingers before the eyes (sc)
V_{OS}: 0.63 sc // 0.9 cc +1.5 Dsph –0.5 Dcyl ax 103
T_{OD} = 12 mmHg, T_{OS} = 12 mmHg

In view of the patient's very low visual acuity, a visual field test could not be performed.

No abnormalities were revealed in the anterior segment of the eye. A fundus examination showed congestion and optic nerve disc elevation, especially in the nasal region (not present 8 days earlier during the patient's assessment at the out-of-hospital ophthalmology clinic), and vascular tortuosity in the right eye. In the left eye, the examination revealed pallor of the disc with slight blurring of its inferior margin (Figures 5, 6).

During the medical appointment, in addition to a significant decrease in visual acuity, the physician's attention was drawn to the patient's behavior, which was perceived as in-

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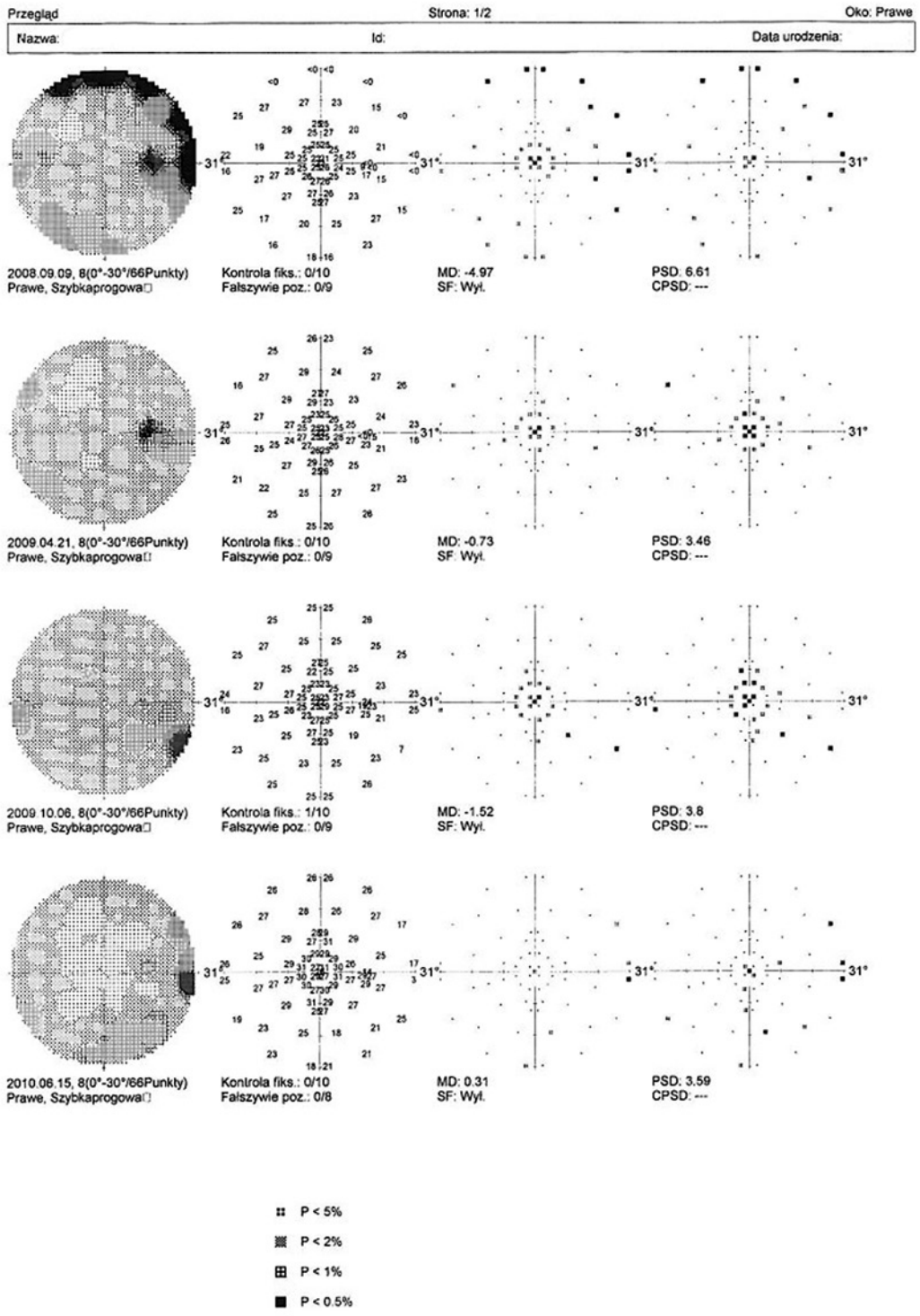

Figure 1. Results of the patient's right-eye visual field test (2008-2010)

adequate to the situation both during history taking and ophthalmic examination. The patient presented with a labile affect and a somewhat dysphoric mood, as well as minor memory impairments and a grandiose belief about her extraordinarily high status, which she had not displayed during previous visits. The patient herself reported that she had been slightly more irritable for approximately six months, which she attributed to the stress caused by the epidemiological situation (COVID-19 pandemic).

In view of the disorders described above, upon admission to the Department, the patient was urgently referred for a CT scan of the head with intravenous administration of a contrast agent. The scan revealed two focal lesions in the left frontal lobe, approximately 4 cm and 2 cm in diameter, with features most likely consistent with meningioma, causing deviation of the median structures to the right and deformation of the falx cerebri, narrowing the sulci over the cerebral convexities and compressing the anterior horn of the left ventricle. The in-

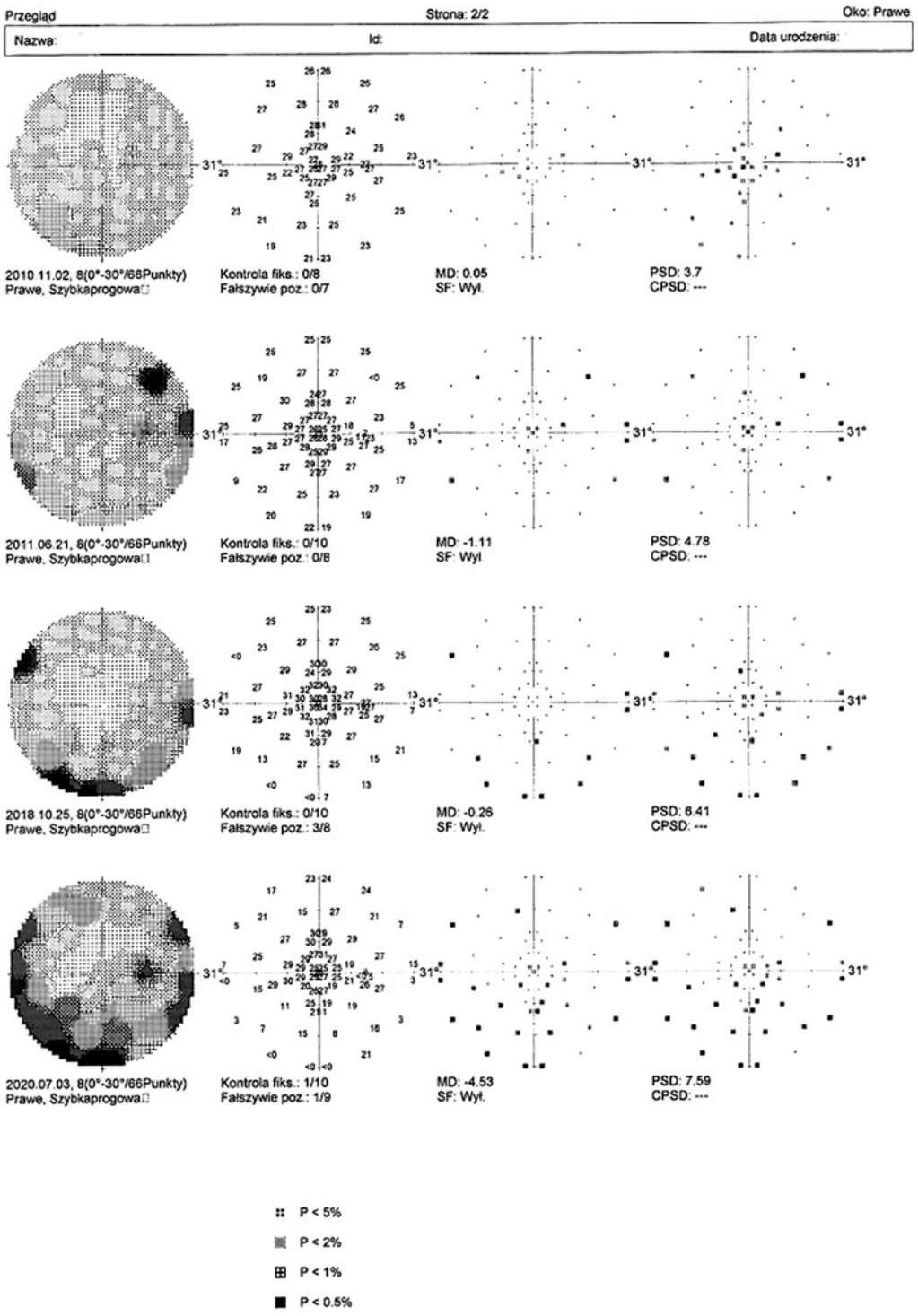

Figure 2. Results of the patient's right-eye visual field test (2010-2020)

ferior pole of the larger lesion was adjacent to the meningeal membrane at the base of the left frontal lobe. Following the intravenous administration of dexamethasone and 15% mannitol, the patient was transferred to the Neurology Department for further treatment (Figures 7, 8).

After performing an MRI examination of the head to confirm the preliminary diagnosis, the patient was referred to the Neurosurgery Department at another center. Left-sided meningioma of the olfactory groove, glabella and optic nerve junction area was diagnosed. The location of the focal lesion correlated well with the patient's fundus findings. Subtotal tumor removal was performed, leaving a part of the tumor mass penetrating bilaterally into the optic-nerve canals and into the right internal carotid artery canal. Histopathological analysis confirmed the diagnosis of meningioma.

Two months after hospitalization, the patient reported for the first postoperative ophthalmic follow-up examination.

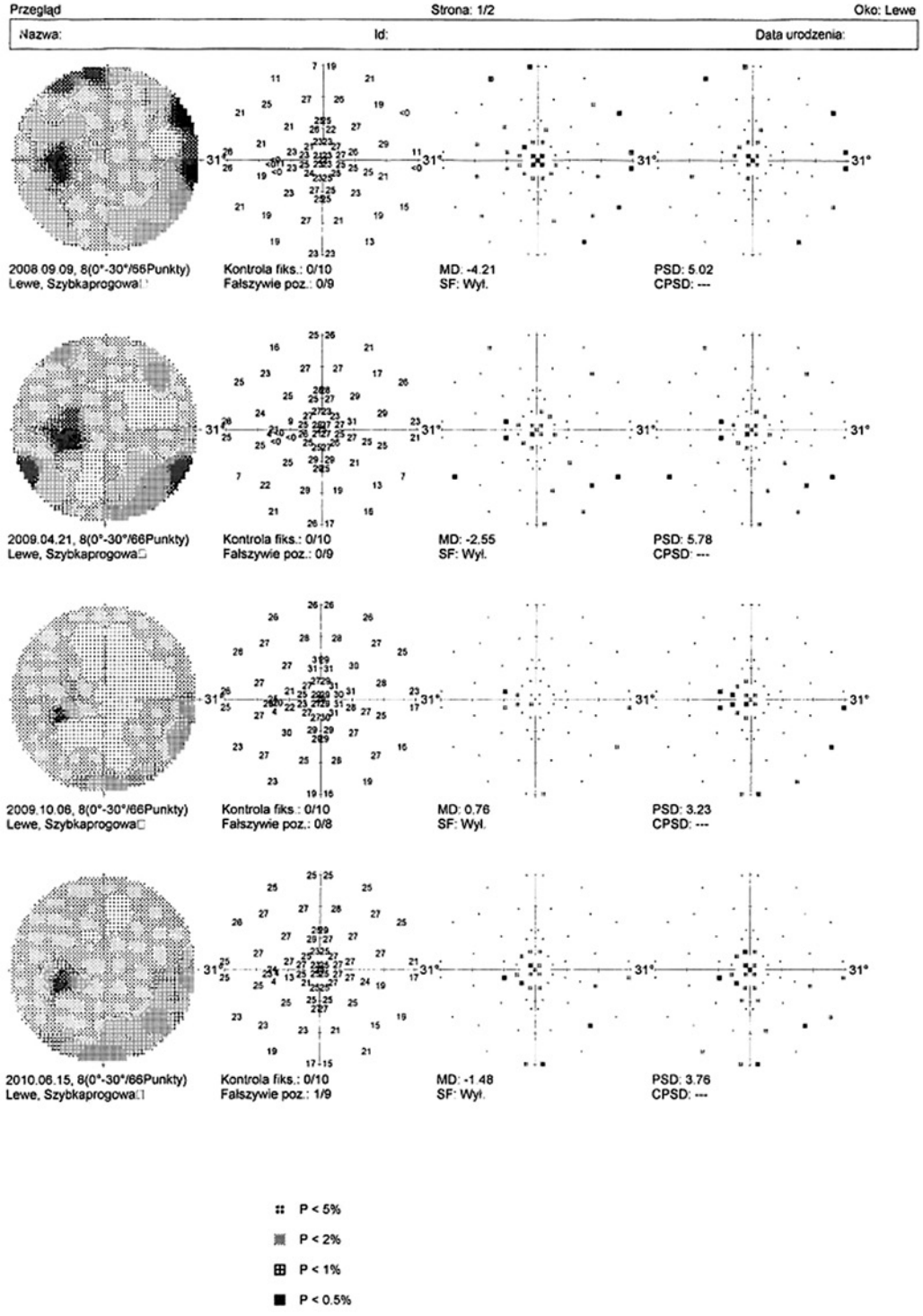

Figure 3. Results of the patient's left-eye visual field test (2008-2010)

Unfortunately, the visual acuity in the right eye declined to no perception of light. In the left eye, it remained at the level of incomplete 0.7. Optic disc edema in the right eye was found to have resolved, while optic disc atrophy in the left eye persisted. The patient did not present any neurological deficits. Behavioral and emotional disturbances were found to have disappeared. The next follow-up appointment took place two months later. The patient supplied a follow-up MRI scan of the head showing a recurrence at the tumor resection site and persistent thickenings in both optic nerve canals. The features of compression in the left lateral ventricle had resolved. As recommended by the attending neurosurgeon, the lesions were left for further observation. Ophthalmic examination revealed no changes in visual acuity in the right eye (absence of light perception) and a decrease in acuity in the left eye to 0.5. Both optic nerve discs showed features of atrophy, but no edema was found. At present, the patient is under the ongoing care of ophthalmology and neurosurgery outpatient clinics (Figures 9, 10).

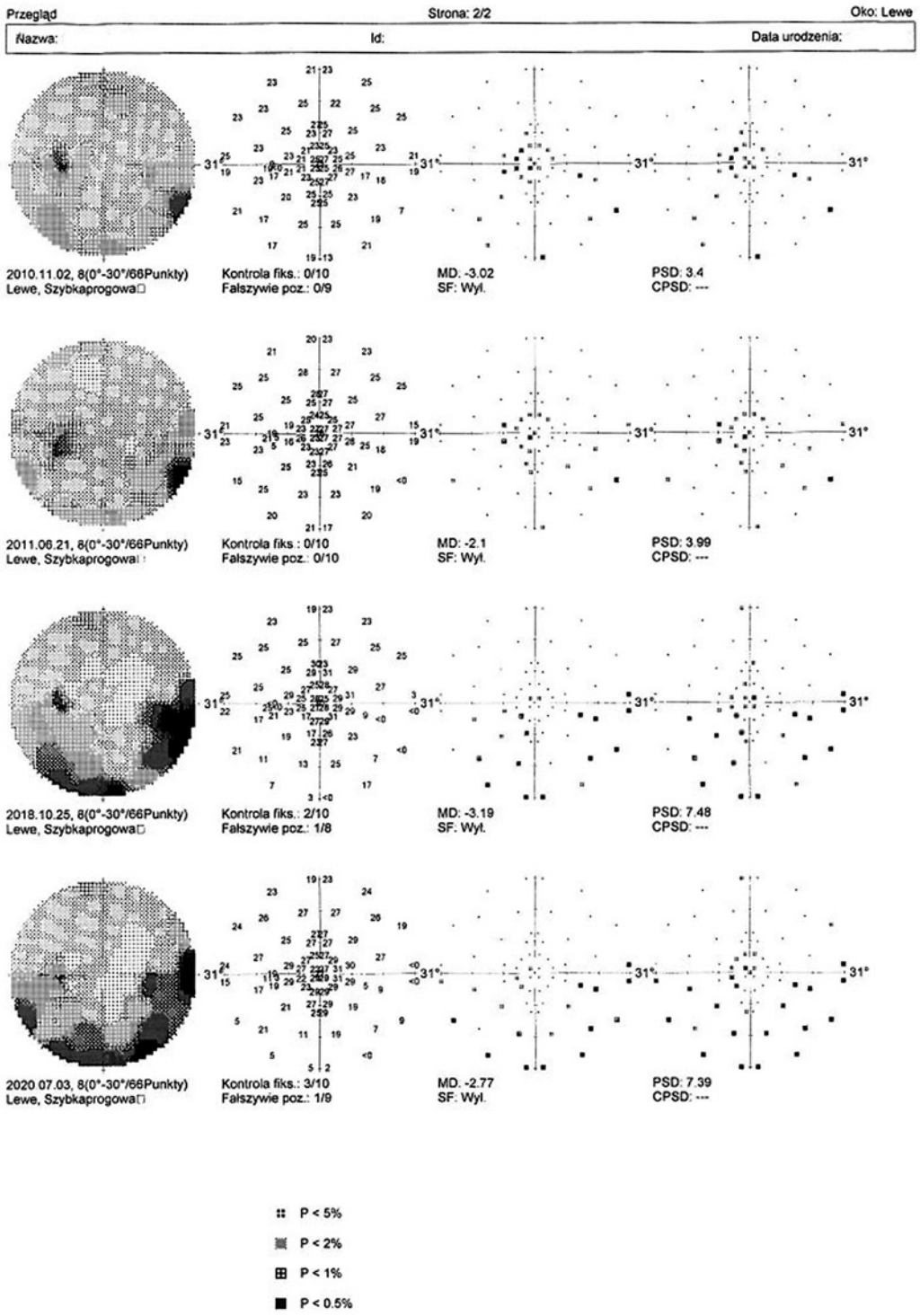

Figure 4. Results of the patient's left-eye visual field test (2010-2020)

DISCUSSION

Foster Kennedy syndrome was first described in 1911 by Robert Foster Kennedy [1, 2]. The diagnostic criteria for the syndrome are not clearly defined and vary to a certain extent depending on the cited source. From the point of view of clinical practice, the most significant manifestations of Foster Kennedy syndrome include unilateral optic atrophy and contralateral optic disc edema. Some authors also note the presence of a central visual field defect on the side of atrophy, as well as unilateral or bilateral anosmia, but from the clinical perspective these symptoms are frequently not amenable to objective assessment because of the patient's insufficient visual acuity or subjective impressions [3, 4]. Based on the available literature, Foster Kennedy syndrome can be classified into three types. Type 1 is the classic presentation with direct compression of the optic nerve and elevated intracranial pressure. Type 2 involves bilateral direct nerve compression without features of intracranial hypertension, while type 3 encompasses cases with-

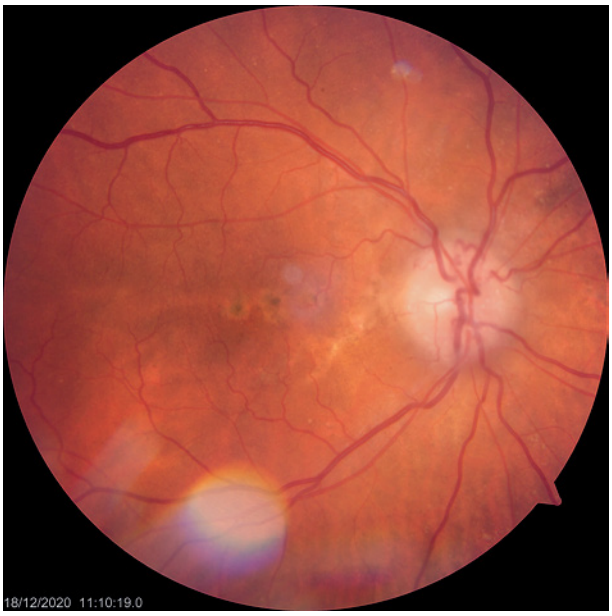

Figure 5. Congestion, elevation of the optic nerve disc in the nasal segment and vascular tortuosity in the right eye

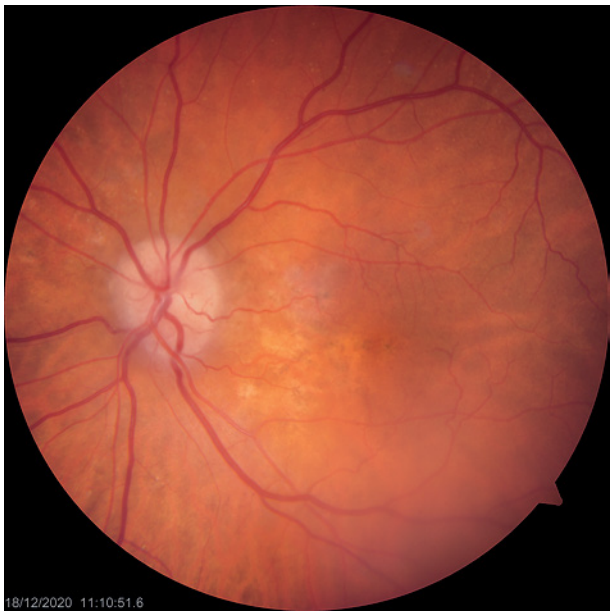

Figure 6. Optic disc pallor with blurring of the inferior margin in the left eye

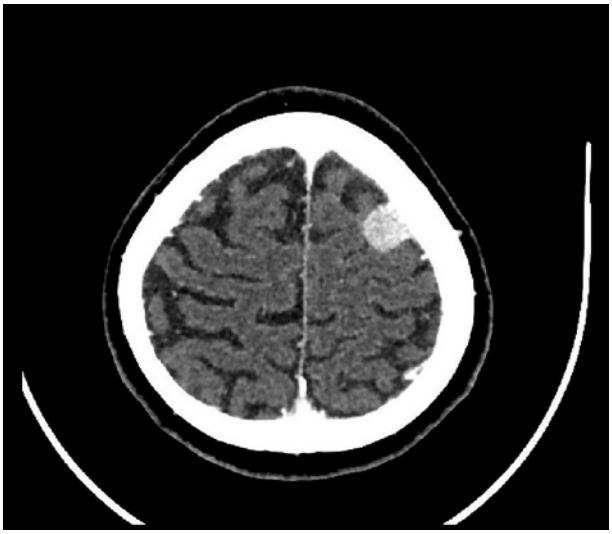

Figure 7. A 2-cm lesion consistent with meningioma in the left frontal lobe

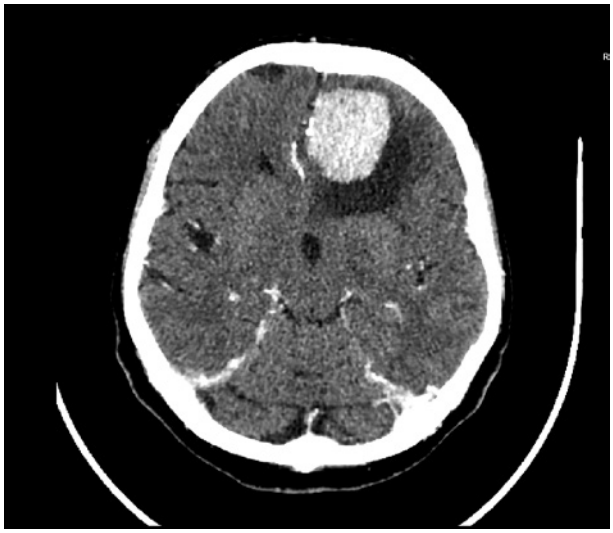

Figure 8. A 4-cm lesion consistent with meningioma in the left frontal lobe

out direct nerve involvement but with chronic intracranial hypertension [5].

The symptoms associated with Foster Kennedy syndrome are due to the presence of focal lesions in the anterior cranial fossa. Most typically, they are primary tumors (usually meningiomas), and less commonly metastases of primary tumors in distant body organs [4, 6]. There are several theories accounting for the development of optic nerve lesions. According to the most widespread and, it appears, the best substantiated theory neurons die as a result of compression of the optic nerve on the side of the tumor. Increasing volume of the lesion causes an elevation in intracranial pressure, which results in the development of edema of the optic nerve disc, but only on the contralateral side. Dead neurons on the tumor side do not enter the edema phase because of necrotic lesions [1-3, 5, 7, 8]. The available literature shows that in the eye affected by optic nerve atrophy, the decrease in visual acuity was very significant and in most cases led to the loss of light perception even after treatment. In the edematous eye, on the other hand, visual acuity ranged from hand movement to 1.0, and in the majority of cases surgical treatment resulted in improved vision [2, 3, 7]. The degree of improvement depends on several factors, the most important of which appears to be the duration of the edema phase [5].

Given the etiology of the syndrome, the clinical findings may also include symptoms resulting from the so-called tumor mass effect (e.g., nausea, vomiting, headaches) or damage to specific brain regions (e.g. emotional lability, impaired memory or orientation if the frontal region is involved) [3, 9]. However, the signs may be subtle. The literature also contains

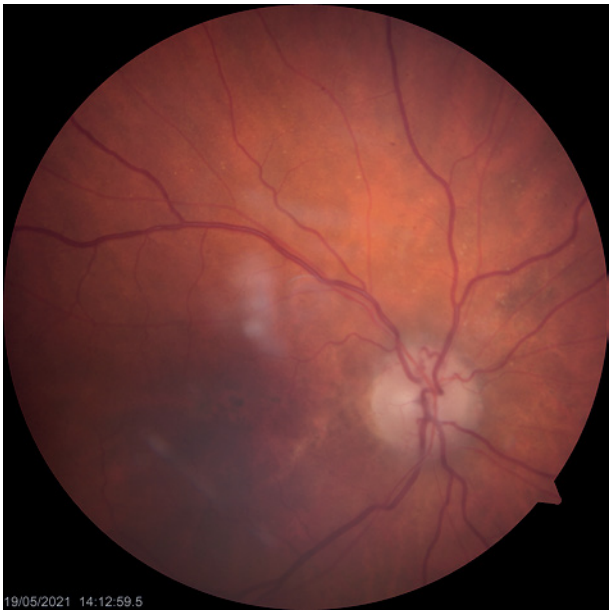

Figure 9. Resolution of optic nerve disc edema in the right eye

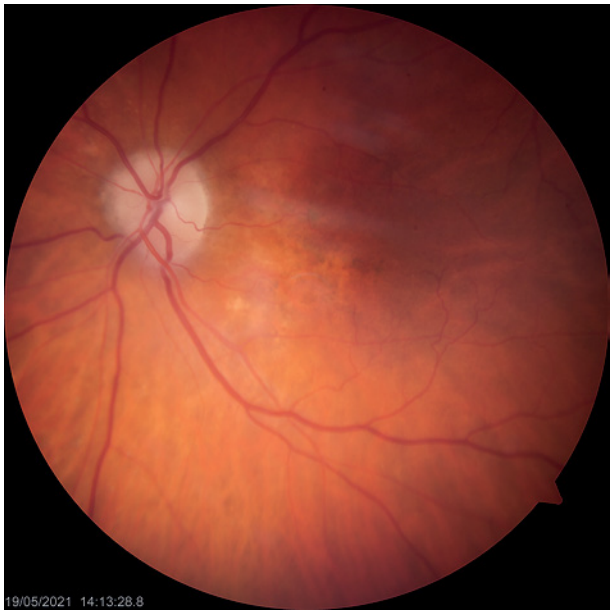

Figure 10. Atrophy of the optic nerve disc in the left eye

very rare reports of coexisting symptoms including diplopia, ptosis, disturbances of pupillary response or impaired function of the oculomotor muscles [2, 3, 6, 9].

The reported clinical findings are relatively rare, manifested by only 1-2.5% of tumors located in the anterior cranial fossa [7]. However, they are definitely very distinctive. Despite not being pathognomonic symptoms of focal lesions, the anomalies discussed above should prompt the physician to refer the patient for urgent diagnostic imaging of the brain and further neurological treatment. The presence of such disturbances always indicates severe damage to the visual pathways [4]. Although MRI of the head is considered the examination of choice, given its limited availability, it would be reasonable to perform a contrast-enhanced CT scan before [3]. Unfortunately, depending on the location, a growing tumor may remain clinically silent for a long time and be already large in size at the time of diagnosis [7].

The clinical manifestation of Foster Kennedy syndrome is not always typical. There are literature reports of focal lesions located beyond the anterior cranial fossa or replacement of contralateral optic disc edema by another acute condition, such as central retinal vein branch thrombosis [6, 10]. Further diagnostic challenges may be associated with pseudo-Foster Kennedy syndrome which, despite characteristic fundus abnormalities, involves no focal changes in the central nervous system, with the anomalies attributed to another pathology. These include idiopathic intracranial hypertension, asymmetrical atherosclerosis in the carotid arteries, carotid artery aneurysm, injury associated with meningitis, syphilis, hydrocephalus, Paget's disease, sarcoidosis, as well as vascular, endocrine or even allergic causes [1, 11].

The patient reported here presented with two main clinical signs (unilateral optic atrophy and contralateral optic disc edema). The patient's low visual acuity prevented a reliable assessment of the visual field. However, the course of the disease was masked by her long-standing diagnosis of glaucoma, which was deemed responsible for the optic disc findings and visual acuity. Some of the symptoms may have been early manifestations of atrophy secondary to meningioma, which is a slow-growing tumor, but the patient's medical records on the diagnosis of glaucoma are insufficient to verify this hypothesis. Nevertheless, the rapid decline in visual acuity correlated with a sudden change in the fundus image and behavioral disturbances that occurred within a week, helped to set the diagnosis on the right track. Still, there are grounds to presume that the neurosurgical diagnosis could have been made earlier, had it not been for the patient's earlier glaucoma diagnosis. Declining visual acuity and deteriorating results of the visual field examination are characteristic of the progression of glaucoma, which is a far more common condition than anterior cranial fossa tumors and was diagnosed in the patient years before. It is also important to highlight the misleading nature of the patient's fundus findings. Optic disc pallor is a symptom associated with a considerably earlier onset than the development of contralateral optic disc edema. In the reported case, it was attributed to glaucoma rather than other conditions requiring an extended diagnostic work-up. This explains why, despite regular follow-up examinations and diagnostic tests, the tumor was not detected until the patient developed symptoms caused by the mass effect.

Rather surprisingly, the patient's final visual acuity levels in both eyes show a better result in the eye in which optic nerve atrophy was originally observed. In most cases described in the literature, visual acuity in the eye with disc edema improved significantly after edema resolution, but in the presented patient it decreased until the loss of light perception. We speculate that various factors may have contributed to this effect, including the patient's long-standing glaucoma masking the early signs of meningioma as well as the tumor's location and the patient's history of

neurosurgical intervention. Importantly, the patient's tumor occupied both optic nerve canals, causing bilateral nerve damage. In typical cases, the tumor has a more superior location. Contralateral post-edematous atrophy of the nerve is due only to tumor mass, not direct infiltration. Also, compression is usually of a shorter duration. In our patient, however, there was bilateral canal involvement, preventing surgical excision of pathological tissues. Consequently, the final visual acuity may have changed in a far more unpredictable manner than in typical focal lesion locations.

Retrospectively, it is difficult to precisely determine the time point at which the process of atrophy associated with tumor compression began in the optic nerve. However, it seems to have persisted for several months, as evidenced by subtle, but nevertheless noticeable, changes in the patient's behavior, which she put down to a stressful life situation. A major role in the diagnostic work-up was played by the cautious attitude of the examining physician who did not ignore alarming signals associated with the patient's mental state.

CONCLUSIONS

The importance of talking to the patient must not be underestimated, and frequently collecting the patient's history provides extensive, and very valuable, diagnostic information. Atypical behavior of the patient, which seems inadequate to the situation, should always give rise to diagnostic caution, especially in the context of neurological conditions that may involve a range of coexisting somatic symptoms, very often of the ophthalmic type.

Unfortunately, the literature on this topic is very scarce, and the available evidence relies primarily on case reports. However, based on the current body of information, one may conclude that the presented collection of symptoms is of substantial clinical benefit in the diagnosis of brain tumors of various etiologies. We consider the case described above to be significant also for another reason, namely the unique course of the condition both due to the presence of glaucoma as a masquerading syndrome and the unusual location of the tumor resulting in unpredictable deterioration of visual acuity. Based on our current knowledge and understanding, this is the first published report of such a case. We hope that the case presentation will reaffirm the need to adopt a cautious clinical approach even in situations that may initially appear entirely typical.

DISCLOSURE

The authors declare no conflict of interest.

References

1. Massey EW, Schoenberg B. Foster Kennedy syndrome. Arch Neurol 1984; 41: 658-659.
2. Eidet JR, Biernat D, Dahlberg D, et al. Foster Kennedys syndrom [Foster Kennedy Syndrome]. Tidsskr Nor Laegeforen 2019; 139 (10). Doi: 10.4045/tidsskr.18.0719.
3. Ruben S, Elston J, Hayward R. Pituitary adenoma presenting as the Foster-Kennedy syndrome. Br J Ophthalmol 1992; 76: 117-119.
4. Von Wowern F. The Foster Kennedy syndrome an evaluation of its diagnostic value. Acta Neurologica Scandinavica 1967; 43: 205-214.
5. Liang F, Ozanne A, Offret H, et al. An atypical case of Foster Kennedy syndrome. Interv Neuroradiol 2010; 16: 429-432.
6. Yildizhan A. A case of Foster Kennedy syndrome without frontal lobe or anterior cranial fossa involvement. Neurosurg Rev 1992; 15: 139-142.
7. Pastora-Salvador N, Peralta-Calvo J. Foster Kennedy syndrome: papilledema in one eye with optic atrophy in the other eye. CMAJ 2011; 183: 2135.
8. Bellot L, Le Guyader F, Mouriaux F. Syndrome de Foster Kennedy révélant un volumineux méningiome [Foster Kennedy syndrome secondary to a large meningioma]. J Fr Ophtalmol 2019; 42: e305-e309.
9. Snyder H, Robinson K, Shah D, et al. Signs and symptoms of patients with brain tumors presenting to the emergency department. J Emerg Med 1993; 11: 253-258.
10. Parafita-Fernández A, Sampil M, Cores C, et al. Foster Kennedy Syndrome: An Atypical Presentation. Optom Vis Sci 2015; 92: e425-430.
11. Visa Reñé N, Paredes Carmona F. Pseudo-Foster Kennedy syndrome due to idiopathic intracranial hypertension. Arch Soc Esp Oftalmol (Engl Ed) 2019; 94: 598-601.