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SHORT COMMUNICATION

Paraneoplastic optic neuropathy in a patient with a non-small cell lung carcinoma: A case report

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PURPOSE. To report a case and describe the clinical approach to a patient with a non-small cell lung carcinoma and paraneoplastic optic neuropathy.
METHODS. Case report.
RESULTS. A 79-year-old woman with known non-small cell lung carcinoma was admitted with a swollen optic disc in the right eye. After detailed clinical, laboratory, and imaging studies, the authors diagnosed paraneoplastic optic neuropathy, excluding other possible diagnosis.
CONCLUSIONS. Paraneoplastic optic neuropathy is a clinical challenge and should be considered as a possible diagnosis in every cancer patient with optic disc edema. (Eur J Ophthalmol 2005; 15: 420-3)

KEY WORDS. Paraneoplastic optic neuropathy, Optic disc edema, Lung cancer

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INTRODUCTION

Paraneoplastic optic neuropathy is a true autoimmune syndrome that can occur in patients with underlying malignancy and should be considered in any patient who develops an optic neuropathy in the setting of systemic cancer, particularly when there is no evidence of central nervous system metastasis (1). Only a small number of cases have been reported in the literature so far. Most of them have been reported in patients with small-cell lung carcinoma, mainly as part of a paraneoplastic brainstem or cerebellar syndrome.

The poor visual acuity with optic disk edema was the main clinical ophthalmologic finding, usually bilaterally (1, 2). In this article we describe a patient with paraneoplastic optic neuropathy, highlighting the differential diagnosis and the step-by-step clinical approach.

Case report

A 79-year-old woman was admitted to our clinic with a 10-day history of poor visual acuity in the right eye. Eight months earlier she was investigated for consistent chest pain, fever, cough, and a lung mass on computed tomography. Fine needle aspiration and histopathologic examination revealed an undifferentiated non-small cell lung cancer with adenocarcinoma autoimmune phenotype. Six months after the cancer diagnosis, liver metastasis was also found on computed tomography examination.

The ophthalmologic examination on admission showed a marked visual acuity decrease in the right eye (1/10) and a fine optic disc edema (swollen optic disc) (Fig. 1). No hemorrhages or other abnormal findings were detected. The left eye was normal with 6/10 of visual acuity due to cataract formation. The kinetic perimetry showed normal

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visual field according to the present visual acuity in the left eye. A blind spot enlargement was detected in the right eye (Fig. 2). Fluorescein angiography showed a gradual late hyperfluorescence of the optic disc, mainly in the site of the ophthalmoscopically swollen disc (Fig. 3). Brain and orbit magnetic resonance imaging (MRI) with and without paramagnetic contrast agent (gadolinium) was also performed, with no abnormal findings. We particularly underline the normal appearance of the meninges, the subarachnoid space, and the orbit and the optic nerve. Our patient underwent a detailed neurologic clinical examination, which was normal. We underline the normal function of the rest cranial nerves and the absence of nystagmus, ataxia, and dysarthria, which excludes brainstem and cerebellar syndrome. The unremarkable neurologic examination of the patient, along with the normal MRI findings, excluded the need for lumbar puncture. Further investigation including cell blood count, serum biochemistry, C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR) were unremarkable according to the present patient's age and physical condition.

From the above clinical, ophthalmologic, and imaging data, we reached the diagnosis of paraneoplastic optic disc neuropathy. The treatment we chose was sub-Tenon corticosteroid injection of long-acting betamethasone.

The patient died 3 months later.

DISCUSSION

In a case of optic disc edema in an elderly patient with cancer and metastasis, the differential diagnosis should include anterior ischemic optic neuropathy, compressive optic neuropathy due to an intraorbital metastasis, carcinomatous meningitis, optic disc metastasis, and paraneoplastic optic disc neuropathy (2, 3).

Anterior ischemic optic neuropathy can be easily excluded because the visual field tests do not have the characteristic defects (sectorial or altitudinal scotoma). For the same reason and in the absence of any type of central or centrocecal scotoma, we were able to exclude all types of optic neuritis.

An intraorbital metastasis or other space-occupying lesion that could exert direct pressure on the optic nerve could be a possible pathogenetic factor in our case (2, 3). The detailed MRI of the orbit and the brain did not find any kind of metastasis, inflammatory, or space occupying lesion.

In patients with known cancer, anywhere in the body, carcinomatous meningitis can occur. This disorder may result from direct extension of a parenchymal metastasis of the leptomeninges at the base of the brain or from hematogenous dissemination of the tumor to the subarachnoid space. It can also develop when a cerebral metastasis impinges on the ventricular system (4). Although a low-grade meningitis, it is characterized by symptoms and signs of damage to the cranial nerves without the specific neurologic symptoms and signs suggestive of common meningitis. Additionally, MRI typically shows diffuse enhancement of the subarachnoid space, often with nodular thickening and enhancement of the dura (4). In our case the neurologic examination was normal and the MRI did not show any of the above findings. Carcinomatous meningitis is out of the question.

Another possible diagnosis is metastasis in the optic disc (3). This is a rare disorder accounting for 5% of all intraocular metastasis. In 27% of patients the lung is the site of the primary tumor. It is a condition that generally occurs unilaterally as enlargement of the optic disc due to tumor infiltration (3). However, it has a particular ophthalmoscopic appearance. In most cases there is a centrally located infiltration of the optic disc of an intensely white or yellow color. The margins are often sharply circumscribed with scallops or nodules. In some cases there is a distinct nodule in the optic disc. The majority of patients with metastatic cancer to the optic disc also have a characteristic contiguous choroidal metastasis (juxtapupillary choroidal component) (3). Such ophthalmoscopic findings were not detected in our patient. Fluorescein angiography also has an adjunctive role in the differential diagnosis. In metastatic neoplasms to the optic disc most tumors show a distinct gradual hyperfluorescence while hypofluorescence is detected in the early phases (3). In our case the angiography showed a gradual hyperfluorescence of the optic disc, compatible to a common swollen edematous optic disc. According to the ophthalmoscopic and angiography findings optic disc metastasis can be ruled out.

Our last consideration in this patient is the rare disorder of paraneoplastic optic neuropathy (1, 2). According to differential diagnosis described above and with the exclusion of the above mentioned possible diagnoses, the clinical and imaging data lead us to the diagnosis of paraneoplastic optic neuropathy. To our knowledge, this is a rather rare disorder with only a few cases reported, in patients mostly with small cell lung carcinoma or other types of carcinomas like lymphoma, neuroblastoma, pancreatic glucagonoma,

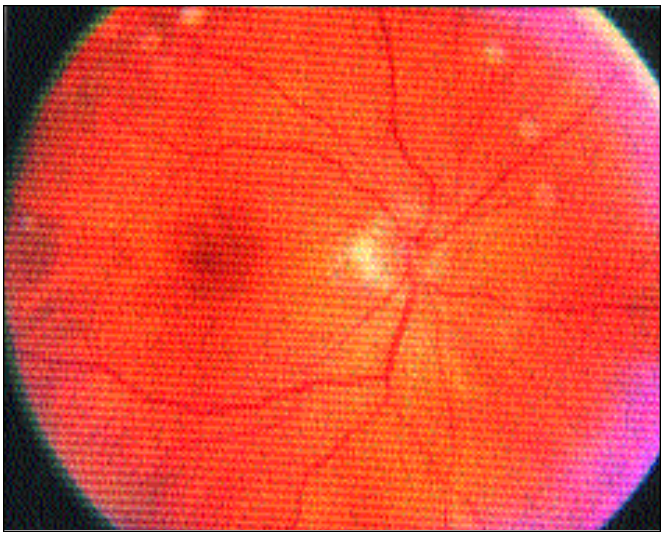


Fig. 1 - Swollen optic disc.

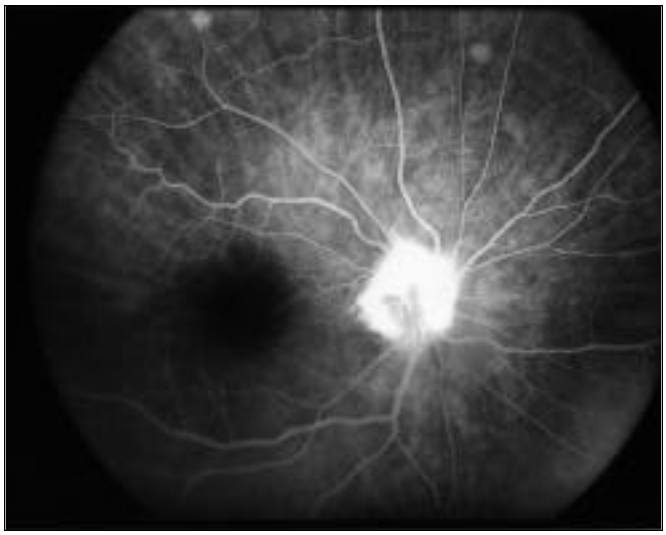
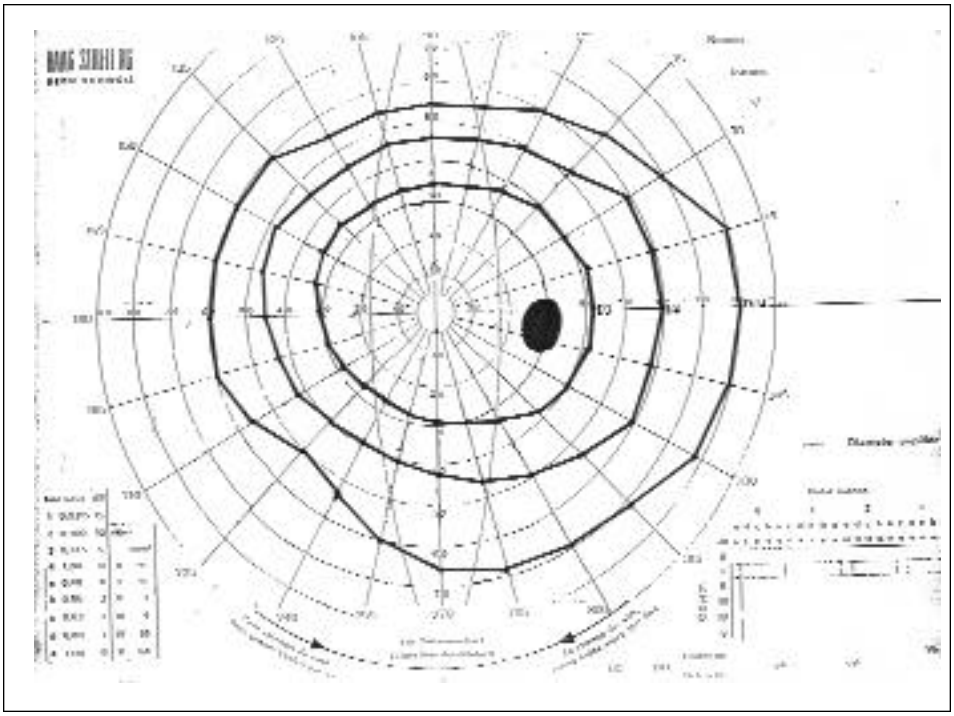


Fig. 3 - Late hyperfluorescence compatible to optic disc edema.

Fig. 2 - Blind spot enlargement of the right eye.



nasopharyngeal carcinoma, bronchial carcinoma, and thymoma. In our case the primary tumor was a non-small cell lung carcinoma, which is interesting as this syndrome is mostly related to small cell carcinoma (2, 5). It is of autoimmune pathogenesis and several neuropathologic findings have shown nonspecific perivascular inflammation, axonal loss, or demyelination of the optic nerve. Poor visual acuity and optic disc edema with or without visual field defects are commonly associated clinical findings (1, 2).

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REFERENCES

1. Jacobson DM. Paraneoplastic diseases of neuro-ophthalmologic interest. In: Walsh and Hoyt's Clinical Neuro-Ophthalmology, vol. 2, ch. 47, 5th ed. Baltimore: Williams & Wilkins; 1998: 2541-43.
2. Chan JW. Paraneoplastic retinopathies and optic neuropathies. Surv Ophthalmol 2003; 48: 12-38.
3. Shields JA, Shields CL, Singh AD. Metastatic neoplasms in the optic disc. The 1999 Bjerrum Lecture: part 2. Arch Ophthalmol 2000; 118: 217-24.
4. Garrity JA. Miscellaneous tumors of neuro-ophthalmologic interest. In: Walsh and Hoyt's Clinical Neuro-Ophthalmology, vol. 2, ch. 46, 5th ed. Baltimore: Williams & Wilkins; 1998, 118: 2485-8.
5. Blumenthal D, Schochet S Jr, Gutmann L, Ellis B, Jaynes M, Dalmau J. Small-cell carcinoma of the lung presenting with paraneoplastic peripheral nerve microvasculitis and optic neuropathy. Muscle Nerve 1998; 21: 1358-9.