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Case Report

Neoplastic leptomeningeal disease masquerading as central serous retinopathy. A case report

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Abstract

A 69-year-old man became aware of people's speech being out of synch with their lip movements alongside persistent headaches, both of which progressively worsened. A few weeks later, he developed progressive and painless visual loss in one eye. Initial neurological evaluation, inflammatory markers and head computed tomography scan were normal. Ophthalmological examination and OCT scan revealed right macular subretinal fluid with choroidal indentation, which prompted urgent further investigations including head MRI revealing extensive leptomeningeal disease. The patient continued to deteriorate and deceased shortly afterwards.

This is the first reported case of neoplastic leptomeningeal disease presenting with loss of vision due to choroidal metastasis with localised exudative retinal detachment. Diagnosing neoplastic leptomeningeal disease requires a high index of suspicion from the treating physician. Symptoms may be nonspecific and/or subtle. Combining cerebrospinal fluid cytology from lumbar puncture with contrast-enhanced magnetic resonance imaging of the brain is considered the optimal diagnostic approach.

Keywords: Neoplastic leptomeningeal disease, Migraine, Choroidal mass

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Introduction

Leptomeningeal metastases are a late complication cancer that result from dissemination of malignant cells into cerebrospinal fluid (CSF), arachnoid mater, pia mater and subarachnoid space. It is estimated to occur in 5% of patients with malignancies but is likely to become more frequent as survival from systemic disease increases and many novel cancer drugs fail to achieve therapeutic concentrations in the central nervous system.[1]

The clinical manifestations of neoplastic leptomeningeal disease (NLMD) can be highly variable and may affect both central and peripheral nervous systems, making diagnosis difficult. The diagnosis of CSF metastases is often delayed and not appreciated until fixed neurological deficits become evident. Ocular symptoms may represent the initial manifestation of meningeal carcinomatosis, even in the absence of other clinical symptoms.

In this paper we would like to share a rare case of NLMD that initially presented with a classical optical coherence tomography (OCT) picture of central serous chorioretinopathy and evolved within a matter of weeks into an obvious choroidal mass. We would like to raise awareness of this devastating and fatal condition.

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Abbreviations: BCVA, best corrected visual acuity, CSF, cerebrospinal fluid, CSR, central serous retinopathy, CT, computed tomography, MRI, magnetic resonance imaging, NLMD, neoplastic leptomeningeal disease, OCT, optical coherence tomography

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Case report

A 69-year-old male presented to his local optometrist complaining of headache and people's speech being out of synchronicity with the movement of their lips for the past 1 week. His medical history was unremarkable.

The optometrist recorded a Snellen best corrected visual acuities (BCVA) of 6/6 in each eye and an unremarkable fundal examination. A diagnosis of migraine was given with simple reassurance and the patient was discharged. Six days later he revisited the same optometrist with persisting symptoms and increasingly blurred vision in the right eye. BCVA had fallen to 6/15 in the right eye and repeat fundal examination revealed a dull foveal reflex. The patient was therefore referred to the local eye department.

Ophthalmological assessment showed no abnormalities in the anterior segments of either eye. Dilated fundus examination revealed right macular subretinal fluid confirmed by OCT scan. As there were no obvious retinal pigment epithelial changes, no intraretinal fluid and no pigment epithelial detachment, a diagnosis of presumed central serous retinopathy (CSR) was made and explained to the patient. Further clinical review in 2 weeks was planned.

One week later, the patient's headache continued to worsen prompting emergency admission at a different medical centre. Neurological evaluation did not reveal any obvious focal deficits, inflammatory markers were within normal limits and a computed tomography (CT) of the head was reported as normal with no evidence of space-occupying lesions (Fig. 2a). The neurological team made a tentative diagnosis of severe migraine and sought an ophthalmic review prior to discharge.

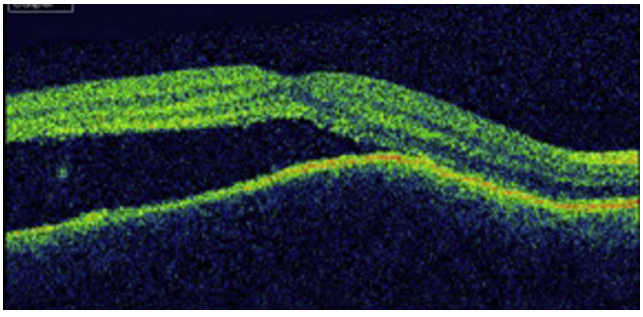

Figure 1. Right eye OCT shows subretinal fluid and choroidal indentation (day 14).

On presentation to the eye clinic, the patient's main ocular complaint was blurred vision in the right eye and he continued to complain of audible speech being out of synchronisation with lip movements. Examination showed BCVA of 6/36 (right eye) and 6/6 (left eye), anterior segment examination was unremarkable with no relative afferent pupillary defect and dilated fundus examination showed right localised macular serous detachment with no signs of optic neuropathy or papilloedema. OCT scan showed right macular subretinal fluid with choroidal indentation (Fig. 1).

The clinical findings together with the patient's symptoms were suggestive of choroidal malignancy. An urgent magnetic resonance imaging (MRI) of the brain with contrast was performed, providing radiological evidence of leptomeningeal disease (LMD) (Fig. 2b). Subsequently, a full-body staging CT scan showed a liver mass and pleural effusion (Fig. 3a and b). Histology of pleural paracentesis confirmed poorly differentiated malignant cells. Unfortunately, the patient continued to deteriorate and sadly deceased 36 h following presentation to our emergency department.

Discussion

Neoplastic leptomeningeal disease is a devastating condition due to dissemination of malignant cells into the CSF, leptomeninges and subarachnoid space. Although NLMD can present as a primary tumour, it is often secondary to a primary tumour elsewhere; most commonly breast carcinoma, lung carcinoma and melanoma.[2]

Neoplastic leptomeningeal disease has a wide variety of clinical presentations with symptoms potentially affecting both central and peripheral nervous systems. After headache, double vision is the second most frequent presenting symptom. Other common ocular complaints include painless loss of vision, which can either be acute or progressive over weeks, and papilloedema, which is often present (albeit not in this case).[3] NLMD carries a very poor prognosis with median survival rate of 3 weeks if left untreated, and this slightly improves to few months with systemic and intrathecal

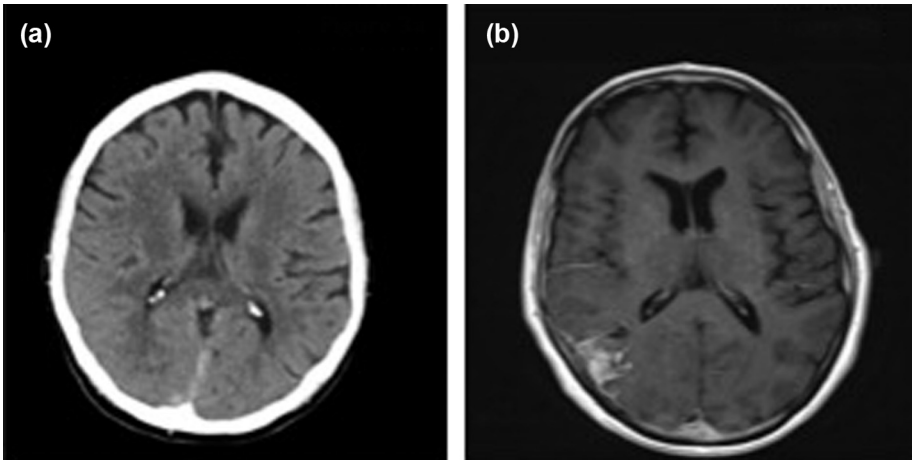

Figure 2. (a) Axial CT scan with no obvious pathology. (b) Axial gadolinium-enhanced MRI of the same area showing extensive meningeal enhancement typical of NLMD.

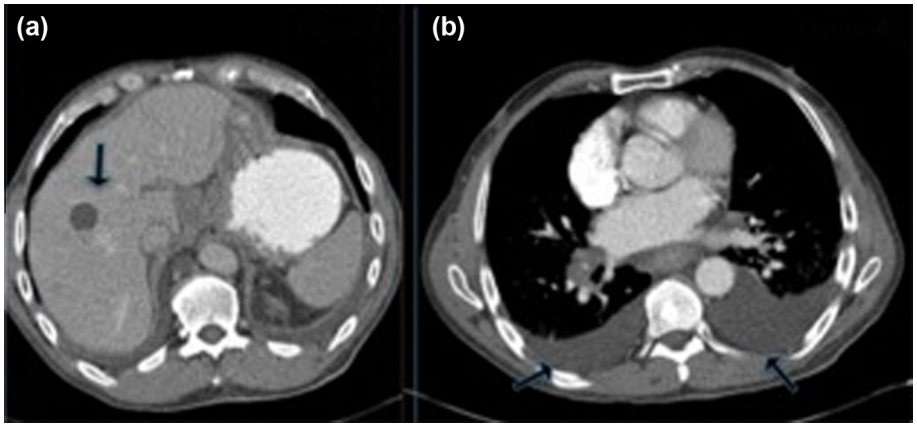

Figure 3. (a) CT scan with solitary liver lesion. (b) Bilateral pleural effusion.

chemotherapy (3–6 months survival rate).[4] It is therefore essential that ophthalmologists are aware of this fatal condition.

Diagnosing NLMD requires a high index of suspicion from the treating physician. It is often missed on CT and even with gadolinium-enhanced MRI, and specificity is only 77%.[5] The most specific diagnostic test for NLMD is CSF cytology by lumbar puncture[6]; initial one has a 50% sensitivity and increases with the number of lumbar punctures performed.[7] A combination of contrast MRI and CSF cytology is probably the optimal diagnostic approach to capture most NLMD patients.[4]

Our patient had painless progressive visual loss. However, unlike published cases where vision is reduced predominantly due to optic nerve involvement, in this case it was due to local choroidal metastasis with localised exudative retinal detachment. This initially resulted in an OCT picture similar to CSR. The patient's other main symptom of dissociation of images and sounds was most likely caused by NLMD affecting the visual cortex and occipito-temporal area.[8]

This is the first reported case of neoplastic leptomeningeal disease presenting with vision loss due to choroidal metastasis with localised exudative retinal detachment. As ophthalmologists we need to be able to differentiate between simple visual symptoms such as photopsia and floaters, where the source is ocular and requires detailed ophthalmic examination, and more complex visual symptoms such as palinopsia, cerebral polyopia and akinetopsia, for which neuroimaging in the form of MRI is required. Therefore, any history of severe, longstanding headaches, associated visual or auditory hallucinations should be treated with high index of suspicion and detailed ophthalmic and medical examination should be carried out. In this case the CT scan with negative inflammatory markers gave false reassurance of no sinister brain pathology, and the patient's headaches were attributed to migraine. Although migraine is a common neurological complaint, one should be suspicious of persistent migraine-like symptoms.[9]

Competing interests

All authors declare no conflict of interest.

Authors' contributions

IE wrote the introduction and performed the literature review, and HS wrote the conclusion both were responsible for drafting the manuscript; PC and HP reviewed and proofread the manuscript.

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