

Central retinal vein occlusion as the initial presentation of isolated optic nerve sheath metastasis from breast cancer

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Abstract

Background: Isolated metastasis to the optic nerve (ON) and its sheath from breast cancer (BC) without involvement of other ocular structures is extremely rare. However, it is a pivotal diagnosis to rule out as it is a both sight- and life-threatening condition. We report a case of isolated ON sheath metastasis from BC presenting with central retinal vein occlusion (CRVO).

Case presentation: A 47-year-old woman with known metastatic BC presented with painless, progressive vision loss in the left eye. Visual acuity was hand movement with ipsilateral relative afferent pupillary defect. Fundal features suggested CRVO. Atypical rapid resolution of these features led to suspect ON metastasis. Magnetic resonance of the brain showed perineural enhancement of the optic nerves. Vision improved with radiotherapy.

Conclusion: Isolated ON sheath metastasis from BC is rare and may present with CRVO. High degree of suspicion is warranted in patients with metastatic disease and atypical findings.

Keywords: breast cancer, metastasis, neoplasms, optic disc, retinal vein occlusion

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Abstrak

Latar belakang: Kes metastatik kepada saraf optik dan sarungnya daripada kanser payudara tanpa penglibatan struktur okular yang lain jarang berlaku. Akan tetapi, pengecualian penyakit kanser metastatik adalah penting untuk mengelak daripada ancaman nyawa dan penglihatan. Kami melaporkan satu kes luarbiasa di mana kanser payudara metastatik hanya merebak kepada saraf optik beserta sarungnya dan menunjukkan ciri-ciri 'central retinal vein occlusion' (CRVO).

Kes: Seorang wanita yang berumur 47 tahun dengan kanser payudara metastatik mengadu bahawa penglihatan mata kirinya semakin merosot. Tahap penglihatannya cuma 'hand movement' dan terdapat 'relative afferent pupillary defect' pada mata tersebut. Pemeriksaan funduskopi menunjukkan ciri-ciri CRVO. Walaubagaimanapun, ciri-ciri ini semakin pudar dengan tempoh yang pendek tanpa apa-apa rawatan. Kadar resolusi yang pantas ini menjadikan metastatik saraf optik disyaki sebagai punca. Pengimejan otak menunjukkan tanda-tanda penglibatan saraf optik. Dengan rawatan radioterapi, tahanan penglihatannya berunsur baik.

Konklusi: Meskipun jarang, kanser payudara metastatik boleh merebak kepada saraf optik sahaja tanpa penglibatan struktur mata yang lain dan boleh menunjukkan ciri-ciri CRVO.

Kata kunci: kanser payudara, metastatik, ketumbuhan, saraf optik, retinal vein occlusion

Introduction

Metastatic disease to the eye is uncommon, with an incidence ranging from 0.07% to 12%.¹ The commonest primary tumour site was found to be breast cancer (BC).² Metastases from BC usually involve the uveal tract or orbital structures in these cases. However, isolated metastases to the optic nerve (ON) and ON sheath without involvement of other ocular or orbital structures are extremely rare. As the clinical signs associated with ON metastasis are not pathognomonic, the diagnosis may be easily overlooked. We report a case of isolated ON sheath metastasis from BC presenting with central retinal vein occlusion (CRVO).

Case presentation

A 47-year-old woman presented with painless and progressive visual loss in her left eye for 11 days. She denied having eye redness, eye swelling, diplopia, floaters, or light flashes. There was no history of headache, nausea, vomiting, numbness, or weakness of the body. She had underlying right BC, which was diagnosed in 2012, when breast conserving surgery with axillary clearance was performed, followed by

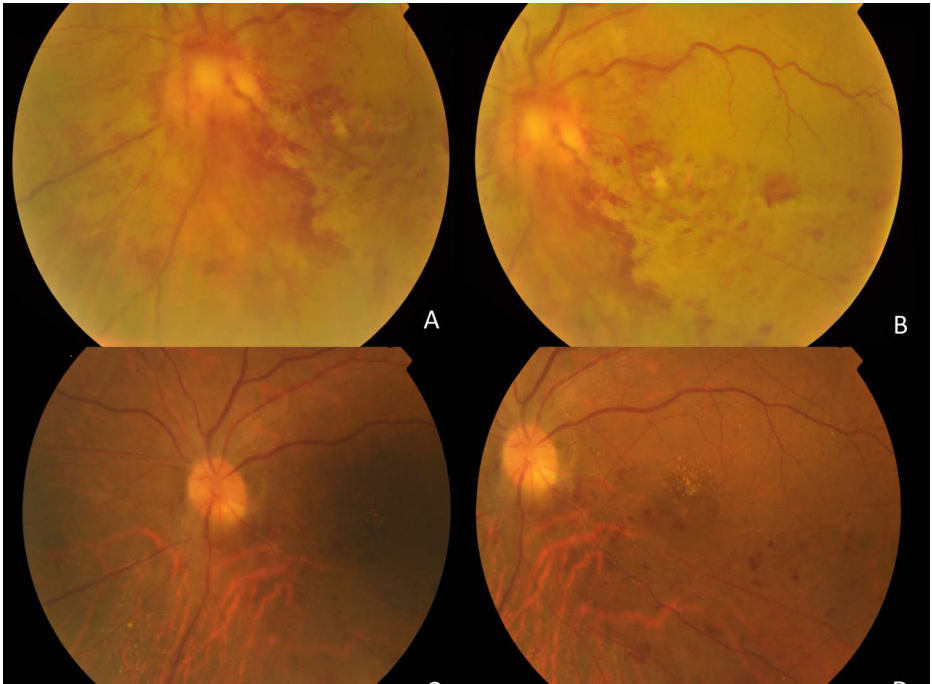


Fig. 1. (A-B) Fundus images of the left eye demonstrating marked disc swelling, flame haemorrhages, multiple intraretinal haemorrhages with tortuous retinal vessels. (C) Spontaneous partial resolution of retinal haemorrhages and macula oedema. (D) Four weeks later, with blurring of inferior optic disc margin remaining.

adjuvant chemotherapy and radiotherapy. Unfortunately, she defaulted treatment after 1 year of tamoxifen. In 2015, she presented with symptoms of local recurrence and lung metastasis; thus, another cycle of chemotherapy was given, and tamoxifen was recommenced. Her disease was stable until March 2018, when she developed supraclavicular lymph node enlargement. Core biopsy of the lymph node revealed metastatic BC. She refused further chemotherapy and was started on anastrozole, an anticancer hormone therapy. Four months later, she presented with the ocular symptoms. She denied having any other significant medical illnesses.

On examination, there was no obvious proptosis, lid swelling, or ophthalmoplegia. Her blood pressure was 136/86. There were no significant findings of the anterior segments of both eyes and the intraocular pressures were normal. Visual acuity was 6/6 in the right eye but reduced to hand movement in the left eye with ipsilateral relative afferent pupillary defect. Fundus examination of the left eye revealed a swollen optic disc with peripapillary haemorrhages. The retinal veins appeared dilated and tortuous. There were flame-shaped haemorrhages and intraretinal haemorrhages in all quadrants but denser in the inferotemporal region (Fig.1A-B).

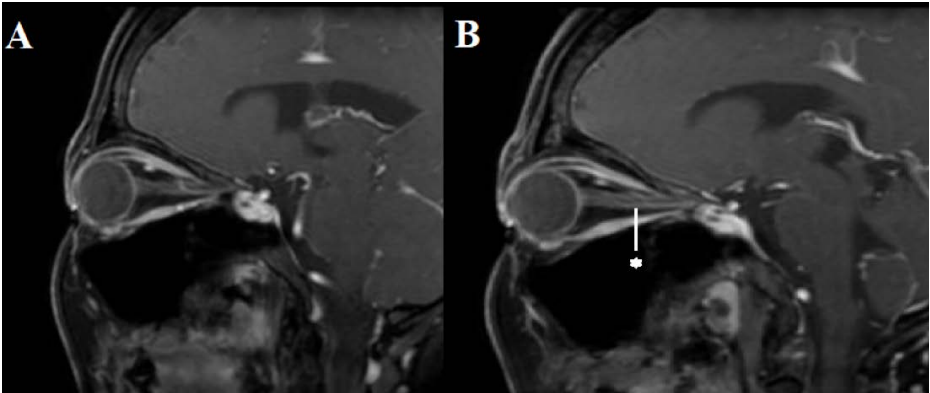


Fig. 2. Right eye (A) and left eye (B) sagittal views of orbit MRI with post-contrast fast spoiled gradient echo sequences. The left optic nerve (B) demonstrates increased perineural enhancement (asterisk) compared to the right optic nerve (A). No mass seen within both intraconal structures.

The macula appeared oedematous, but no hard exudates or cotton wool spots were visualised. The vitreous was clear and no neovascularization was seen at the disc or elsewhere. The fundus of the right eye was unremarkable.

A diagnosis of left CRVO with macular oedema was made. Preliminary blood investigations such as full blood count, coagulation profile, renal profile, and fasting blood glucose were normal. Fundus fluorescein angiography (FFA) was scheduled for a later date, to avoid the inaccuracy of capillary non-perfusion evaluation from the masking effect by the haemorrhages. Options for intravitreal anti-vascular endothelial growth factor injection were given but deferred due to financial constraints. However, upon review 4 weeks later, we observed marked spontaneous improvement of the disc swelling, retinal haemorrhages, and macular oedema with only blurring of the inferior optic disc margin remaining (Fig.1C-D). This unusual phenomenon triggered the suspicion of ON metastasis. Urgent contrasted magnetic resonance imaging (MRI) of the brain revealed perineural enhancement of both optic nerves, which was more prominent on the left, with no intraconal or extraconal mass (Fig. 2). The oncology team was alerted, and third-line hormonal therapy, exemestane was commenced. She was also given radiotherapy, 20 grays in 5 fractions with 6-MV photons to the orbit bilaterally. Eight months later, visual acuity in the left eye was limited to counting fingers due to a superior altitudinal defect, but she was able to achieve vision of 6/45 in a chin up position. Further resolution of the disc swelling and retinal haemorrhages was observed. The right eye retained visual acuity of 6/6 with a normal ocular examination.

Discussion

The most common site for intraocular metastasis is the uveal tract, likely attributed to its rich vascular supply. BC is the most frequent source of ocular metastases (43%), followed by lung cancer (27%).² Even so, ocular metastases is uncommon with bone, brain, liver, and lung being the usual sites of distant BC metastasis.³

Isolated ON and/or ON sheath metastasis from BC without the involvement of other ocular or orbital structures is very rare. Metastatic cancer to the optic nerve can occur as a direct hematogenous spread to the neural tissue or to the overlying meninges.² Optic nerve metastases from BC in general exhibit abnormalities of the optic disc such as pallor or oedema. However, our patient presented with CRVO.

Features of CRVO include dilation and tortuosity of all branches of the central retinal vein, retinal haemorrhages scattered throughout all four quadrants, and optic disc oedema, all of which were present in our patient. CRVO has only been reported in one case⁴ before, although the temporal relationship between the two was unclear, as CRVO was noted 7 months prior to the diagnosis of metastatic disease.

Although the exact pathogenesis remains elusive, occurrence of CRVO is thought to follow the principles of the Virchow's triad of thrombogenesis, including venous stasis, hypercoagulability, and vessel damage. In malignancies, CRVO is regarded as a paraneoplastic process where a complex interplay between tumour cells, leucocytes, platelets, coagulation system, and vascular endothelium contribute to the pathogenesis of CRVO.⁵ Certain therapeutic agents for the treatment of BC such as tamoxifen have also been associated with retinal vaso-occlusive disease, such as branch retinal vein occlusion.⁶ In our case, this could have been compounded by the impediment of venous outflow from the compression by the ON sheath metastases itself.

Surprisingly, marked spontaneous partial resolution of the retinal haemorrhages, optic disc swelling, and macular oedema was observed within 4 weeks without any change in systemic hormonal therapy or chemotherapy. This atypical rapid improvement was an unusual observation in CRVO considering the median time to resolution of retinal haemorrhages was reported to be 9.5 months in the posterior pole and 20.7 months in the peripheral retina for non-ischemic CRVO.⁷ This duration was even longer for ischemic CRVO. We postulate that perhaps the occlusion site of the central retinal vein was more posterior, where a greater number of tributaries anterior to the occlusion allowed re-establishment of collateral flow leading to a transient, yet significant disturbance of the retinal circulation.

The prognosis of patients with metastasis to the ocular structures is poor, with a mean survival of 9–13 months in those with metastasis to the retina and ON.^{2,4} Our patient remains a cancer survivor at the time of writing, 14 months since the diagnosis of CRVO was made.

Conclusion

Isolated ON sheath metastasis of BC is extremely rare and may present with CRVO. Although CRVO is not an unusual occurrence in the context of systemic malignancies, the presence of atypical features and/or optic neuropathy with a background history of metastatic BC should alert the clinician with the possibility of this grave diagnosis.

Declarations

Ethics approval and consent to participate

Not required.

Consent for publication

Informed consent was obtained from the patient for publishing this case report.

Competing interests

None.

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