

Leptomeningeal carcinomatosis: easy to miss

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Abstract

Leptomeningeal carcinomatosis is a devastating condition with very poor prognosis. With advances in cancer treatment leading to improved survival in solid tumours, the incidence of ante-mortem diagnosis of leptomeningeal carcinomatosis is likely to rise. Without a high index of suspicion and appropriate imaging the diagnosis can be missed. We report two cases of leptomeningeal carcinomatosis secondary to metastatic breast disease with varied clinical presentation where the diagnosis was initially missed.

Keywords: carcinomatosis, leptomeninges, MRI

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Case history

Case 1

A 67-year-old female presented with a 7-week history of increasing confusion, visual hallucinations and reduced mobility. MRI brain performed in a district hospital was incomplete as the patient was unable to remain still due to her mental status. Only the T2-axial weighted images could be acquired, which showed moderate ventriculomegaly. A provisional diagnosis of normal pressure hydrocephalus was made. She was eventually admitted to our hospital, the regional neuroscience centre, for therapeutic lumbar drainage. However, her level of consciousness deteriorated progressively.

On reviewing the history it became apparent that she had been treated for breast cancer 2 years previously with surgery, chemo- and radiotherapy. She had been discharged from follow up by her oncologist. Repeat MRI of the brain with contrast demonstrated diffuse leptomeningeal enhancement along the cerebral and cerebellar convexities as well as along the margins of the basal cisterns (Figure 1). A diagnosis of leptomeningeal carcinomatosis was made; it was confirmed by cerebrospinal fluid cytology. The patient received palliative treatment and died within a few days.

Case 2

A 58-year-old female presented to the local emergency department with acute onset, lower motor neurone-type facial weakness, impaired hearing and auricular headache. She was diagnosed with Bell's palsy and given a course of steroids and aciclovir. She re-presented to the emergency

department with ataxia. The initial non-contrast MRI showed only mild small vessel ischaemic changes. A new diagnosis of posterior circulation stroke was made and she was referred to the vascular neurology clinic. Physical examination revealed significant truncal and right-sided appendicular ataxia with impaired vibration and position sense. She also had ipsilateral hearing loss, prominent lower motor neurone-type facial weakness and impairment of taste sensation.

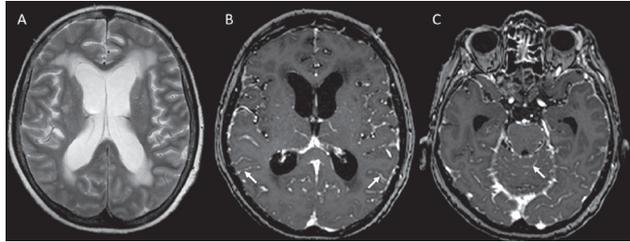
Further history revealed she had developed a new breast lump which was under investigation (she was due to have biopsy of the lump). This raised the suspicion of perineural metastases along the cranial nerves. MRI of the brain with contrast demonstrated intense enhancement of the cerebellar leptomeninges (Figure 2). It also showed enhancement of the seventh and eighth nerves bilaterally, in the internal auditory meati and in the intra-temporal course of the facial nerves. A diagnosis of leptomeningeal carcinomatosis with perineural spread along the seventh and eighth cranial nerves was made. A subsequent staging CT showed metastasis in the right lung. Cerebrospinal fluid analysis was unremarkable. The patient died before histological or cytological sampling could be obtained.

Discussion

Leptomeningeal carcinomatosis is defined as diffuse seeding of the leptomeninges by cancer cells. The condition was first reported in 1870; however, the term was not used until the early 20th century. Up to 30% of cancer patients can be expected to develop this. Breast cancer is one of the most common primary cancers that can have leptomeningeal spread. Breast cancer may spread to the pia and arachnoid

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Figure 1 A. T2W axial image showing moderate dilatation of the lateral ventricles. There is periventricular T2 hyperintensity which is likely to be a combination of small vessel ischaemic changes and transependymal cerebrospinal fluid seepage from hydrocephalus. B. Post contrast scan demonstrates enhancement of the leptomeninges along the cerebral convexity sulci (arrows). C. Post contrast T1W image showing leptomeningeal enhancement along cerebellar sulci (arrow).



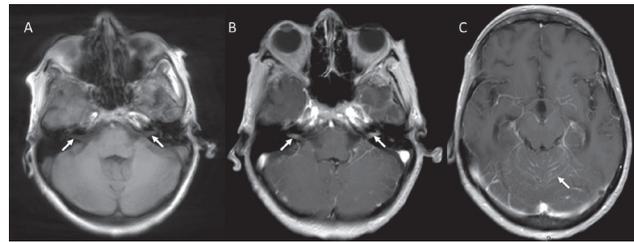
mater via an aggressive pathological process of dissemination known as carcinomatous meningitis.¹ Other malignancies such as lung cancer, melanoma and lymphoma can also show leptomeningeal spread.²

Patients typically present with symptoms caused by the effects of the direct tumour cell invasion into the subarachnoid nerve roots, invasion into the brain or spinal cord, or cerebrospinal fluid obstruction. The clinical picture can be varied and the diagnosis can be missed in the absence of a clear cancer history (as in case 1) and a thorough systemic examination (as in case 2).

Our second case also illustrates the importance of a thorough neurological examination and evaluation of the signs in establishing the correct diagnosis. Bell's palsy, a common cause of lower motor neurone-type facial nerve palsy, may be associated with hyperacusis rather than deafness (which suggests involvement of the eighth cranial nerve and is incompatible with the diagnosis of a simple Bell's palsy). The presence of ataxia when the patient re-presented was further evidence that this was not a simple Bell's palsy.

Early diagnosis is important as this may allow the oncologist to begin therapy prior to neurologic deterioration. The preferred initial investigation is a gadolinium-enhanced MRI. Contrast-enhanced CT is less sensitive and may miss the diagnosis in a third of cases. On CT, leptomeningeal spread may be interpreted as intraparenchymal in another

Figure 2 A. Normal appearances of the nerves (arrows) in the auditory meati on T1W non-contrast scan. B. Post-contrast T1W image shows enhancement of the nerves (arrows) in the internal auditory meati. C. Post contrast image shows intense leptomeningeal enhancement (arrow) along the cerebellar sulci.



third. Malignant cells, infiltrating into the subarachnoid space, disrupt the blood–brain barrier, thus allowing the contrast agent to permeate and outline the leptomeninges. The resulting enhancement can vary from thin and linear to thick and nodular.³ A post-contrast sequence with gadolinium is needed to demonstrate such enhancement. It is therefore important for the clinician requesting the MRI scan to provide the history of any previous cancer – treated, stable or progressive – so the radiologist is alerted to add contrast-enhanced sequences to the imaging protocol.

Definitive diagnosis requires cerebrospinal fluid cytology or leptomeningeal biopsy. The diagnostic accuracy of a single lumbar puncture is only 50–60%; multiple lumbar punctures may be necessary when clinical suspicion is high. **1**

References

- 1 Jayson GC, Howell A, Harris M et al. Carcinomatous meningitis in patients with breast cancer. An aggressive disease variant. *Cancer* 1994; 74: 3135–41.
- 2 Rhun EL, Taillibert S, Chamberlain MC. Carcinomatous meningitis: Leptomeningeal metastases in solid tumors. *Surg Neurol Int* 2013; 4(Suppl): S265–88.
- 3 Smirniotopoulos JG, Murphy FM, Rushing EJ et al. Patterns of contrast enhancement in the brain and meninges. *Radiographics* 2007; 27: 525–51.