Rare disease

Unusual intracranial spread of anaplastic oligodendroglioma

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Summary

Oligodendrogliomas are the third most common type of glioma. Oligodendrogliomas are typically indolent tumours, yet the majority of them are incurable and most patients will ultimately die. Anaplastic oligodendrogliomas are important to recognise from other primary brain tumours, since they have unique molecular, histological and clinical features. Oligodendroglioma, especially the anaplastic type, has a tendency to intracranial and extracranial metastasis. In this paper, we present an unusual manifestation of anaplastic oligodendroglioma.

BACKGROUND

Oligodendroglioma accounts for about 2–5% of primary brain tumours.^{1–3} It is mostly identified in adults, its peak of incidence occurring in the fifth and sixth decades of life, although not common in childhood and adolescence.^{1–4} Oligodendrogliomas have a tendency to invade the leptomeninges;¹ however, metastasis is more common from the ventricular ependyma than through the subarachnoid space.⁵ In this article we present an unusual case, with a history of anaplastic oligodendroglioma and bilateral optic nerves and lateral ventricle involvement.

CASE PRESENTATION

A 53-year-old woman referred to our centre subsequent to having undergone surgical operation for anaplastic oligodendroglioma (figure 1) of the right frontal lobe. Four years before surgical operation, she had a single episode of tonic-clonic seizure which had never repeated. No imaging studies had been done. One month prior to undergoing surgical operation she had been complaining of headache, nausea and vomiting. Brain CT scanning (figure 2) before starting radiation therapy revealed a large tumoral mass with intensive contrast enhancement and significant peripheral oedema.

DIFFERENTIAL DIAGNOSIS

Anaplastic oligodendroglioma can mimic other anaplastic gliomas and metastatic neoplasm.

To differentiate it from metastatic tumours the characteristic features of a well-differentiated oligodendroglioma should be sought in remote areas of the specimen. These features in combination with infiltrative properties, calcification, chicken-wire vasculature, vascular proliferation and frequent glial fibrillary acidic protein positivity serve to identify the neoplasm. In contrast to other anaplastic gliomas especially glioblastoma multiformis (GBM) the vascular proliferation in oligodendroglioma rarely attains glomerular formation and necrosis is generally devoid of pseudopalisading.

TREATMENT

Radiation therapy with two parallel opposed lateral fields was done with a total dose of 6000 cGy

OUTCOME AND FOLLOW-UP

One month after termination of radiation therapy, new CT scanning showed partial response. Then she received chemotherapy with PCV regimen (procarbazine, CCNU, vincristine) every 8 weeks. Her condition was improving; however, 6 months postradiation she complained of decreased visual acuity. Ophthalmological examination revealed optic nerve atrophy but repeating CT scanning showed a decreased size of her previous lesion, without any significant findings and unfortunately she lost her vision. Repeated ophthalmological examination once again confirmed bilateral optic atrophy. MRI performed

Figure 1 Section shows proliferation of oligodendroglial cells with hypercellularity and nuclear pleomorphism. High mitotic figures, vascular proliferation and necrosis are seen.

Figure 2 Brain CT scanning shows a large mass in the right frontal lobe with significant oedema, mass effect and small calcified area in its anteromedial aspect.

with gadolinium showed, in addition to the primary tumour in the frontal lobe, multiple enhanced small subependymal lesions in the left and right lateral ventricles (figure 3) and the right side of the pons (figure 4). Thickening and enhancement of optic chiasma and

Figure 3 Subependymal lesions evident in lateral ventricles, a lesion in the anterior part of right lateral ventricle and a smaller one in lateral of left lateral ventricle.

Figure 4 MRI shows an enhanced lesion in the right side of pons.

intracranial of both optic nerves due to metastatic lesions were seen (figure 5). Subsequently, we prescribed temozolomide for the patient but the proposal was refused due to the high price of the drug. She died 1 year postoperation.

DISCUSSION

Oligodendrogliomas are less prevalent than astrocytomas; however, they seem to be more common than previously thought.⁶ The multilevel grading system for oligodendrogliomas has been proposed. In the meantime a two-level classification of differentiated (grade 2) or anaplastic (grade 3) appears to be as useful.¹ Of these, roughly 70% are low-grade oligodendroglioma and 30% are anaplastic oligodendroglioma.⁶ Oligodendrogliomas have unique pathological and clinical characteristics. The recognition that codeletion of the short arm of chromosome 1 (1p) and the long arm of chromosome 19 (19q) is a diagnostic marker for oligodendroglial tumours was associated with better response to chemotherapy and improved clinical outcome.^{1 4 6}

Although oligodendroglioma can be found anywhere in the brain, its most common site is in the frontal lobes. Some believe that oligodendrogliomas arising from the frontal lobes are more aggressive and more probably metastatic to cerebrospinal fluid (CSF) and show widespread growth.¹ Cerebellopontine angle leptomeninges, retina, intraventricular system (including fourth ventricle), brainstem, septum pellucidum and optic nerve are among the unusual sites of primary oligodendroglioma.¹

Although Liwnicz and Rubinstein⁷ and most other authors believe that glioblastoma multiforme is the most common type of glial tumours that can spread through CSF and cause extracranial metastases, some others state that oligodendroglioma more likely metastasise outside the central nervous system than other gliomas.¹ Metastatic involvement of the CSF pathways in oligodendrogliomas probably occurs in 1–2% of patients.⁸ Even disseminated

Figure 5 MRI shows thickening and enhancement optical chiasma and intracranial of both optic nerves in axial(A) and coronal(B) views.

CSF spread of oligodendroglioma (grade 2) has been seen.⁹ Spine is the most common site of metastases.⁹ Extracranial metastases are more unusual but have been described in the bones, lymph nodes bone marrow, liver and other sites.^{8–10} Zustovich *et al* reviewed 33 cases of metastatic

oligodendroglioma. They divided these patients into two different groups. The first included the patients with synchronous or “early” metastatic disease, and the “early” not defined by authors. The studied patients frequently had bone marrow involvement and a very aggressive course. The median overall survival for patients with bone marrow involvement was 17 months and 38 months for the others. The second group had late metastatic spread. The patients in this group had mostly lymph node involvement and better median overall survival.¹¹ According to previous studies multiple craniotomies and using ventriculoperitoneal (VP) shunt could be associated with tumour dissemination, most probably due to dural damage.^{5–8} However, our patient did not have VP shunt. As we know it is the first case of bilateral optic nerve metastases besides lateral ventricle metastases. There are three ways for the spread of oligodendroglioma. The first pattern is following lymph nodes of scalp to distant areas. Another is through CSF and the third is through haematogenous metastases.⁹ Therefore, in patients with oligodendroglioma, especially anaplastic type, the physician must be warned about extracranial and intracranial extension of disease, and if this occurs prognosis would be poor despite chemotherapy and radiation therapy.

Learning points

- ▶ Oligodendrogliomas can spread intracranially and extracranially and when it occurs the prognosis will be poor.
- ▶ Intracranial dissemination can be seen without previous ventriculoperitoneal shunting.
- ▶ Disseminated oligodendroglioma may involve optic nerves and optic chiasma.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Akhavan A, Binesh F, Navabii H. Unusual intracranial spread of anaplastic oligodendroglioma. *BMJ Case Reports* 2012;10.1136/bcr-2012-006354, Published XXX

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