Rare disease

Interventricular low-grade oligodendroglioma with multiple parenchymal relapse

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Summary

Oligodendrogliomas can be found anywhere oligodendrocytes exist; however, they mostly occur in frontal lobes. Although intra- and extra central nervous system dissemination of anaplastic oligodendroglioma is a well-known property of this tumour, low-grade oligodendroglioma with intracranial relapse is a very uncommon finding. In this case report, a 37-year-old man with grade II oligodendroglioma relapsed after 18 months with multiple parenchymal masses is presented.

BACKGROUND

Oligodendroglioma constitutes 2–5% of primary brain tumours most commonly found in the frontal lobe.¹ It can invade the ventricular system; however, primary intraventricular cases are very rare.² Seeding through cerebrospinal fluid (CSF) usually is seen in primary parenchymal rather than intraventricular type.³ Although there are very few reports of intracranial dissemination of grade II type, they are mostly found in anaplastic oligodendroglioma (grade III). A case of interventricular grade II oligodendroglioma with intracranial dissemination is reported here.

CASE PRESENTATION

A 37-year-old man referred to our centre after stereotactic biopsy of ill-defined 4 × 3 cm mass in the frontal lobe, exactly between anterior horns of the lateral ventricles. Stereotactic biopsy was performed in Shohaday-e-Tajrish, one of the most famous neurosurgical departments in the Islamic Republic of Iran. Owing to the high risk of morbidity neurosurgeons did not propose surgery. When the patient was admitted in our centre he was in a good general condition and his Karnofsky score was 90%. The patient's history did not show any serious medical problem except for nausea in two previous months. Two weeks before undergoing biopsy, he had experienced an episode of tonic-clonic seizure. An MRI examination (figure 1) was carried out that revealed lateral displacement of lateral ventricles and moderate dilatation of lateral ventricles and non-communicating hydrocephaly due to Monro foramen obstruction. An ill-defined lobulated isosignal mass between the lateral ventricles and in genu portion of the corpus callosum extending to the suprasellar cisterna associated with marked surrounding oedema was seen. The lesion showed faint enhancement subsequent to Gadolinium injection. Biopsy consisted of multiple irregular pieces of tan brown colour between the soft tissues with a total size of 1.2 × 1 × 0.5 cm. Microscopic evaluation showed oligodendroglioma grade II (WHO classification) (figure 2).

DIFFERENTIAL DIAGNOSIS

The well-differentiated oligodendroglioma must be distinguished from the fibrillary astrocytoma and the ependymoma. However, oligodendroglioma lack the fibrillary background of the astrocytoma. Nuclear regularity associated with perinuclear halo and the general presence of calcification are additional features. The cellular ependymoma may also simulate an oligodendroglioma. Both tumours share monotonous sheets of glial cells and, on occasion, calcification. The focal presence of the epithelial surface clearly establishes an ependymal origin. In their absence, the distinction rests with perinuclear halos and the absence of pseudorosettes in the oligodendroglioma. An uncommon lesion that closely resembles the oligodendroglioma is the central neurocytoma. Its cells often

Figure 1 MRI shows an ill-defined lobulated isosignal mass between the lateral ventricles with significant oedema.

Figure 2 Microscopic feature of oligodendroglioma grade II.

align in column in a highly cellular background and in contrast to the oligodendroglioma the neurocytoma stains positively for neural proteins as synaptophysin.

TREATMENT

As no surgical plan was proposed for the patient, radiation therapy was done. The initial radiation field consisted of the whole ventricular system for treating any probable seeding till 4000 cGy and 3 cm margin to the tumour. Then the field shrank to the tumour and the margin decreased to 2 cm and finally the patient received a dose of 5400 cGy.

OUTCOME AND FOLLOW-UP

Repeating MRI 1 month after termination of treatment showed decreased tumour size and another MRI 1 year after treatment did not show any enhancing mass. The patient was well for 6 months when he complained of polyuria and amnesia. Laboratory examinations such as fasting blood sugar, sodium, potassium, renal function tests and urine analysis, especially specific gravity, were normal; however, brain MRI showed multiple enhancing brain lesions in both hemispheres (figure 3). Stereotactic biopsy was not possible because of the small size of the lesions. The most probable diagnosis was recurrence of primary tumour and treatment with PCV (procarbazine, CCNU and vincristine) was started. The patient received only one course but because his condition deteriorated the treatment was discontinued.

DISCUSSION

Oligodendrogliomas are the third most common glial tumours and account for 2–5% of primary brain tumours.¹ According to WHO classification, oligodendrogliomas are classified as low grade (grade II) and anaplastic (grade III).¹ Recently, recognition of molecular markers has changed the diagnosis and treatment of oligodendroglioma. Deletion of the short arm of chromosome 1 (1p), and especially when accompanied by deletion of long arm of chromosome 19 (19q), is a diagnostic marker for oligodendroglioma, an independent prognostic factor for chemosensitivity and better survival.¹⁴ Although Liwnicz and Rubinstein⁵ and most other authors believe that glioblastoma multiforme is the most common type of glial tumours that can be metastatic, some others state that

Figure 3 MRI shows multiple round enhancing lesions.

oligodendroglioma more likely metastasises to outside the central nervous system rather than to other gliomas.¹ Majority of oligodendroglioma dissemination cases occur in anaplastic type; however, there are few reports of dissemination in grade II disease.⁶ Primary brain tumours spread in four ways: (1) local invasion, (2) seeding through CSF, (3) through involvement of scalp lymph nodes and then spread via lymphatic system and (4) haematogenous spread through vascular channels.^{6 7} Spine is the most common site of metastasis.⁶ According to a review study by Zustovich *et al*,⁸ the most frequent extracranial metastatic sites are the bone and bone marrow followed by the lymph nodes, lung and pleura. Zustovich *et al*⁸ reviewed 33 cases of metastatic oligodendroglioma. They divided these patients into two different groups. The first group included patients who had synchronous or early metastatic disease, the latter cases not defined by the authors. The patients frequently had bone marrow involvement and a very aggressive course. The median overall survival was 17 months for patients with bone marrow involvement 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.⁸ According to previous studies, multiple craniotomies using ventriculoperitoneal shunt could be associated with tumour dissemination most probably due to dural damage.^{6 9 10} However, our patient did not have ventriculoperitoneal shunt and stereotactic biopsy was performed only once. In a study by Kanamori *et al*,¹⁰ unfavourable factors for dissemination of oligodendroglioma were grade III tumours, retention of chromosome 1p or 19q Ki-67 labelling index of C25%, diffuse expression of nestin, and p53 labelling index of $\geq 10\%$. Most of the patients with dissemination disease had received previous radiation therapy.⁹ The irradiation may be contributory towards

inhibition of the ability for immunological defence against the tumour. According to the study performed by Ng *et al*,⁶ only six cases of metastatic oligodendroglioma grade II were reported previously. All of them were older than our patient and their mean age was 49.5 years. Metastatic site included spinal vertebra in two patients; and leptomeningium, pleura and chest wall, parotid gland, vermis and occipital lobe and cellular region, medulla oblongata, and frontal lobe ventricles in the sixth patient. Time interval from initial diagnosis to dissemination was between 2 and 17 years. Histological transformation to a higher grade of tumour may occur late in the disease progression. Regardless of histological grade, metastatic oligodendroglioma has a poor prognosis.⁵ Sampling was not possible in our patient as a result of small deep lesions when the tumour recurred tissue. In the meantime, our patient experienced the shortest interval (20 months) between disease diagnosis and recurrence.

Learning points

- ▶ Although most cases of disseminated oligodendroglioma are found in grade III, it can also be seen in grade II.
- ▶ Dissemination of oligodendroglioma may be seen without previous ventriculoperitoneal shunting and multiple craniotomies.
- ▶ Prognosis of the disease is poor regardless of histological grade.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. Engelhard H, Stelea A, Mundt A. Oligodendroglioma and anaplastic oligodendroglioma: clinical features, treatment, and prognosis. *Surg Neurol* 2003;**60**:443–56.
2. Tekkok IH, Ayberk G, Saglam S, *et al*. Primary intraventricular oligodendroglioma. *Neurochirurgia* 1992;**35**:63–6.
3. Zada G, McNatt SA, Gonzalez-Gomez I, *et al*. Anaplastic intraventricular oligodendroglioma: case report and review of the literature. *Surg Neurol* 2009;**71**:693–700.
4. Blakeley J, Grossman S. Anaplastic Oligodendroglioma. *Curr Treat Options Neurol* 2008;**10**:295–307.
5. Liwnicz BH, Rubinstein LJ. The pathways of extraneural spread in metastasizing gliomas: a report of three cases and critical review of the literature. *Hum Pathol* 1979;**10**:453–67.
6. Ng WH, Lim TC, Tan KK. Disseminated spread of recurrent oligodendroglioma (WHO grade II). *J Clin Neurosci* 2006;**13**:602–7.
7. Kim JG, Park CO, Hyun DK, *et al*. Spinal epidural metastasis of cerebral oligodendroglioma. *Yonsei Med J* 2003;**44**:340–6.
8. Zstovitch F, Della Puppa A, Scienza R, *et al*. Metastatic oligodendrogliomas: a review of the literature and case report. *Acta Neurochir (Wien)* 2008;**150**:699–702.
9. Han SR, Yoon SW, Yee GT, *et al*. Extraneural metastases of anaplastic oligodendroglioma. *J Clin Neurosci* 2008;**15**:946–9.
10. Kanamori M, Kumabe T, Sonoda Y, *et al*. Predictive factors for overall and progression-free survival, and dissemination in oligodendroglial tumors. *J Neurooncol* 2009;**93**:219–28.

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