

Ziya Akar
Necmettin Tanriover
Ali M. Kafadar
Nurperi Gazioglu
Büge Oz
Cengiz Kuday

Chiasmatic low-grade glioma presenting with sacral intradural spinal metastasis

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Z. Akar
Department of Neurosurgery,
Cerrahpaşa Medical Faculty,
Istanbul University,
Istanbul, Turkey

N. Tanriover
Department of Neurosurgery,
Cerrahpaşa Medical Faculty,
Istanbul University,
Istanbul, Turkey

A.M. Kafadar
Department of Neurosurgery,
Cerrahpaşa Medical Faculty,
Istanbul University,
Istanbul, Turkey

N. Gazioglu
Department of Neurosurgery,
Cerrahpaşa Medical Faculty,
Istanbul University,
Istanbul, Turkey

B. Oz
Department of Pathology,
Cerrahpaşa Medical Faculty,
Istanbul University,
Istanbul, Turkey

C. Kuday
Department of Neurosurgery,
Cerrahpaşa Medical Faculty,
Istanbul University,
Istanbul, Turkey

N. Gazioglu (✉)
5. Gazeteciler Sitesi A 19/4,
Levent, 80620 Istanbul, Turkey
e-mail: nurperig@hotmail.com

Abstract Leptomeningeal metastasis of low-grade gliomas in children has been documented in several series, both at the time of diagnosis and at relapse. The authors report a unique case of chiasmatic low-grade astrocytoma presenting with signs and symptoms related to the metastatic site rather than the primary site. In this respect, the possibility of appearance of symptoms and signs related to leptomeningeal dissemination preceding the signs and symptoms belonging to the primary site should be considered in this type of benign tumours.

Keywords Chiasmatic low-grade glioma · Leptomeningeal dissemination · Spinal metastasis

Introduction

Astrocytomas of the optic nerves and chiasm account for 7% of central nervous system tumours in children [11]. The more aggressive hypothalamic-chiasmal gliomas of the juvenile type are histologically benign, and only in extremely rare cases has histological malignancy ensued [2, 9, 12]. Since the chiasmal gliomas may extend upward into the hypothalamus and hypothalamic gliomas may extend downward to involve the optic chiasm and tract, chiasmal and hypothalamic gliomas may be indistinguishable from one another radiologically and surgically [10].

Young children aged less than 5 years with chiasmatic-hypothalamic glioma have been known to suffer an aggressive course, and they show higher morbidity and mortality rates than older patients [3, 9]. Sacral intradural spinal metastasis of childhood chiasmatic-hypothalamic

low-grade astrocytomas through leptomeningeal dissemination is extremely rare in the literature [1, 2, 4–6]. Leptomeningeal seeding of low-grade chiasmatic gliomas has been documented in several series, both at the time of diagnosis and at relapse [1, 2, 4, 6, 7]. However, no cases have been documented with presenting signs and symptoms appropriate to the metastatic site, rather than the primary site of the tumour.

In this paper we report a unique case of low-grade chiasmatic glioma first presenting with signs and symptoms of sacral intradural spinal metastases.

Case report

In October 1997, an 8-year-old boy presented to the Department of Neurosurgery in Cerrahpaşa Medical Faculty of Istanbul University with a 2-month history of difficulty in walking and low back pain radiating to his left leg. On admission neurological examination revealed only paraparesis, especially in proximal muscle groups, sad-

diotherapy. Whole-brain radiation therapy and whole-spinal radiation in 20 fractions were given.

Discussion

A small percentage of paediatric low-grade gliomas, in contrast to their benign characteristics, manifest widespread dissemination either at presentation or later [1, 2, 4–6]. Although cerebrospinal metastases of low-grade astrocytomas are considered rare, they have been found to metastasize, especially through leptomeningeal dissemination [1, 2, 4, 6, 7, 9].

Chiasmatic and hypothalamic astrocytomas are lesions arising in close proximity to the ventricles and basal cisterns, and they show a predilection for CSF seeding. Tumour spread here almost certainly occurs on the basis of CSF dissemination. Biological factors that may contribute to dissemination include tumour consistency and degree of intracellular adhesiveness [7, 8].

It is likely that patterns of adhesion molecule production, protease secretion, and growth factor pathway activation within the tumour may play a part in determining the ability of free-floating tumour cells to become adherent to and to multiply on ependymal and leptomeningeal surfaces [1, 7, 8].

To our knowledge, leptomeningeal spinal metastases of low-grade chiasmatic-hypothalamic gliomas have previously been reported in coming up to 12 cases [1, 2, 4, 5, 6, 7]. In only 4 of these was the timing of the leptomeningeal dissemination and manifestation of the primary tumour synchronous. The onset of symptoms and signs secondary to spinal metastasis in the remaining 8 cases was recognized in periods of 4 months to 8 years [1, 2, 4, 5, 6, 7].

Clinically, hypothalamic gliomas present primarily with hypothalamic dysfunction, and optic gliomas with visual loss. This may be the most useful point of difference, since the distinction between a large chiasmatic glioma and a hypothalamic glioma may be arbitrary.

Our patient presented with difficulty in walking and low back pain radiating to his left leg. He had no visual field defects and no diencephalic syndrome, and visual acuity was also normal. A two-stage operation was planned and the pterional approach was chosen for the subtotal removal of this chiasmatic lesion. The sacral lesion was subtotally excised in the second stage of the operation, with the knowledge that this could be a spinal metastasis of a low-grade astrocytoma through leptomeningeal dissemination.

As we mentioned above, in the literature only 4 cases of chiasmatic-hypothalamic low-grade gliomas showed simultaneous signs and symptoms of primary and of metastatic sites. This is a unique case, since all the presenting signs and symptoms arose from the metastatic site, rather than the primary site of the tumour.

Thorough screening of the neuraxis with MRI is mandatory in children with low-grade astrocytomas who are noted to have multiple leptomeningeal or periventricular deposits on their initial cranial neuroimaging studies. However, routine imaging evaluation of the spinal axis probably is not needed in patients with unifocal intracranial disease and no symptoms that can be referred to the spine [7]. On the other hand, all cases presenting with only signs and symptoms belonging to the metastatic site due to leptomeningeal dissemination should have their entire neuraxis evaluated with MRI scans.

In conclusion, spinal metastasis of chiasmatic low-grade gliomas are rare, although the number of cases reported may increase in the future with the frequent use of MRI, which is the modality of choice for detecting and delineating the extent of tumour dissemination. These patients can present with signs and symptoms secondary to the metastatic site as a result of leptomeningeal dissemination, and these may precede other signs and symptoms belonging to the primary site.

This possibility should be remembered in the follow-up of children with low-grade chiasmatic-hypothalamic gliomas.

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