Rare disease

Extraventricular neurocytomas

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Summary

Central neurocytomas are uncommon, benign tumours of the central nervous system. These tumours have been recognised outside the limits of the cerebral ventricles and, in these instances, have been called as 'extraventricular neurocytomas'. The authors described a case of frontal neurocytoma and discuss the current knowledge on these infrequent neoplasms.

BACKGROUND

Central neurocytomas (CN) corresponding to WHO grade II, are uncommon, benign tumours of the central nervous system usually associated with favourable outcome. Traditionally, these lesions were reckoned as intraventricular oligodendrogliomas or ependymomas until detailed immunocytochemical analysis of their neuronal phenotype was clarified. Currently, these tumours have been well recognised outside the limits of the cerebral ventricles and, in these instances, have been called as 'extraventricular neurocytomas' (EVNs). CN comprises only 0.1–0.5% of all brain neoplasms.¹ EVNs are even rarer. The first report dates from 1989 and they affect predominantly children and young adults (median 34 years), with no gender predilection. Since then, few studies have reported additional cases of EVNs. Many controversies exist regarding EVNs: origin, clinical course, recurrence, need for adjuvant therapy and final outcomes. The authors described a case of a patient harbouring a frontal neurocytoma and discuss the current knowledge on these infrequent neoplasms.

CASE PRESENTATION

A 26-year-old female patient sought medical attention due to multiple episodes of seizures.

INVESTIGATIONS

MRI revealed a frontal lesion that intensely enhances after gadolinium injection, without relation with the ventricles (figure 1). No perilesional oedema was noticed. As the lesion presented an apparent dural base, the preoperative diagnosis was meningioma.

TREATMENT

Patient underwent a right frontal craniotomy. The lesion presented an ill-defined plane of dissection with the adjacent brain and no relation with the dura. However, it was totally removed with negligible blood loss. Pathological examination displayed a neurocytoma (figure 2).

OUTCOME AND FOLLOW-UP

Postoperative course was uneventful and no adjuvant therapy was proposed. After 2 years of clinical follow-up,

patient presented significant improvement in the number of seizures and MRI showed no recurrence.

DISCUSSION

EVNs are rare primary tumours with no apparent connection with the cerebral ventricles. Reports have demonstrated involvement of cerebral hemispheres (commonly frontal and parietal lobes), thalamus, cerebellum, pons, amygdala, pineal gland, retina and spinal cord.¹ Neurocytomas have also been reported arising from unusual sites, such as an ovarian teratoma in the pelvic cavity.¹ Due to the small number of reported cases most of the current knowledge about EVNs derives from studies on more common CN, however, whether or not these data can be extrapolated to EVNs remains uncertain.

According to the original description of Hassoun *et al*², it has been accepted that CN arises from the nuclei of the septum pellucidum. Other locations have been suggested, such as the third ventricle and the subependymal plate of the lateral ventricles, as possible sites of origin. It remains controversial as to whether neurocytomas arise from cells committed to neuronal phenotype or from bipotential cells in the periventricular matrix.² In vitro studies have demonstrated that CN have reminiscent features of bipotential precursor cells, which may present glial and neuronal differentiation. Neurocytomas have now been reported in almost every subcompartment of the craniospinal axis.¹ Finding neurocytomas in extraventricular locations may require revisiting the current theory that subependymal progenitor cells are the cells of origin for these tumours.

In contrast to CN, typical EVNs display a wider morphological spectrum encompassing arrangement of neurocytes in sheets, clusters, ribbons or rosettes, with neuropil dispersed either in broad zones (neuropil islands) between these cell arrangements or localised within the rosettes.^{1–3} Neurocytes most often demonstrate finely granular, slightly eosinophilic cytoplasm, occasionally with artefactual vacuolations resembling oligodendroglioma.¹ Nuclei of these tumours are strikingly monotonous and rounded, with finely speckled chromatin, distinct nuclear rims and one to three small nucleoli. Ganglionic differentiation is more common in EVNs and has been observed in 66% cases in one series.³

Figure 1 MRI revealed a frontal lesion that intensely enhances after gadolinium injection, without relation with the ventricles.

Figure 2 Pathological examination displayed a neurocytoma.

EVNs have been more often described as pathological reports. So that, clinical presentation, role of radical removal, need for adjuvant therapy and final outcome are still understudied. EVNs generally manifest through mass effects in the form of seizures and hemiparesis. Brat *et al*⁶ described a series of cerebral EVNs that presented with seizures, diplopia, headache and vomiting. Overall, fewer than 100 local recurrences have been reported to date, most of which were associated with atypical neurocytomas and EVNs.¹ Neurocytomas are thought to represent the benign end of the spectrum of neuronal tumours that are generally amenable to surgical excision, with an overall favourable outcome.¹ Even residual tumours are noted to behave in a biologically indolent fashion. Thus, the majority of EVNs is well differentiated and appears unlikely to recur after gross total resection. Subtotal resection, atypical histologic features and high cell proliferation rates may correlate with recurrence.¹

Alternative/adjuvant modalities in the form of radiation therapy (RT) have been pursued, especially in a setting of subtotal resection and/or recurrence. However, considering the benign nature of the tumour and risks of radiation toxicity, routine prophylactic radiotherapy should be avoided in cases with complete resection and in cases that are not atypical. Owing to the paucity of cases and lack of adequate follow-up, role of RT as primary treatment modality after biopsy remains controversial and in our opinion is not recommended for typical tumours.

Chemotherapy has been found beneficial in recurrent CN that cannot be resected but have been irradiated without success. It has usually constituted part of a multimodality

therapeutic regimen as an acceptable alternative to potential long-term side effects of radiotherapy in younger patients.

Learning points

- In summary, EVNs are very rare lesions and most of our understanding derives from pathological studies on CN. However, they may represent distinct entities with different natural history and outcomes. Overall, EVNs are benign lesions that are probably curable through gross total resection. Radiotherapy and chemotherapy should be reserved for very specific cases and do not play an important role in the routine treatment. Since most of studies are from pathological reports, the publication of clinical cases is paramount to clarify the clinical course, the role of total resection and the need for adjuvant therapy.

Competing interests None.

Patient consent Obtained.

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