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AUTHOR'S REPLY

Multiple desmoplastic infantile gliomas—therapeutic challenges

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Dear Editor:

We appreciate the interest of Rojas-Medina and Moro in our recently published manuscript [1, 2]. They raise some important issues which definitely will require further clarification in the future. Indeed, desmoplastic glioma might not be such a benign lesion when we consider the clinical course in some instances which have been reported. This is even more true for those patients who presented with multiple lesions, either combined cranial and spinal lesions or with multiple cranial lesions only. While in some patients tumors remain stable or may even regress, there is a rapid progression of the tumor in others, and as demonstrated repeatedly, the patient may also succumb within a few months.

Another important issue is whether or not there is a primary lesion initially with subsequent secondary lesions or whether these tumors may indeed manifest in multifocal locations. There are arguments for both. We presume that the spinal tumors rather present seeding of the supratentorial tumors via CSF. Conversely, however, with regard to the nature of these tumors and their maldevelopmental features, simultaneous appearance at multiple cerebral sites is possible.

Furthermore, we would like to emphasize that according to recent observations, desmoplastic infantile gliomas often carry BRAF V600E mutations. Koelsche et al. [3] observed BRAF mutations in 3 out of 18 patients which was the case also in the patient published by us [1]. The availability of selective BRAF V600E inhibitors like vemurafenib or dabrafenib may open new therapeutical options for these patients. Remarkably, patients with anaplastic gangliogliomas and pleomorphic xanthoastrocytomas with BRAF V600E mutations may respond to such treatment [4–6].

Due to the rarity of these tumors, available information has been derived mainly from case reports or from small case series. Hopefully, this tumor entity will deserve more attention in the future and data from different centers may be pooled to obtain further information about the true nature of these unusual tumors.

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Compliance with ethical standards

Conflict of interest There is no conflict of interest.

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