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CE - LETTER TO THE EDITOR

Central nervous system hemangiopericytoma with bone and lung metastases: a case report

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Dear Editor,

A 69-year-old woman, who had undergone surgical resection of a brain tumor diagnosed as meningioma 7 years previously, came to our attention for persistent headache and a weight loss of about 5 kg during the last month. Physical examination was normal; in particular no neurological deficiencies were found. Blood tests revealed normocytic anemia only. A gadolinium-enhanced brain magnetic resonance showed an intracranial lesion of about 3 × 3 cm close to the meningeal falx, which was suggestive of meningioma recurrence (Fig. 1a). Therefore, surgical excision of the meningeal lesion was planned. On a pre-operative chest X-ray a pulmonary nodular lesion was found in the left lower lobe. Thoracic computed tomography showed pulmonary bilateral nodular masses and bone abnormalities of ribs and dorsal vertebrae. A gadolinium-enhanced magnetic resonance of dorsal and lumbar column revealed a T10 nodular vertebral-epidural lesion with spinal compression (Fig. 1b, c). On positron emission tomography, areas of increased tracer uptake suggestive of metastatic malignant lesions in these sites were observed (Fig. 1d, e). A bone biopsy of T10 vertebral body was performed: spindle-shaped cells arranged around blood vessels, positive for vimentin and negative for epithelial antigens on immunohistochemical analysis, were detected, suggesting the diagnosis of hemangiopericytoma (HPC). Then, the patient underwent surgical excision of the spinal and intracranial meningeal lesions, whose histological and immunohistochemical analysis confirmed the diagnosis of HPC. The bioptic specimen of the meningeal intracranial lesion which had been resected 7 years previously was re-examined: a similar immunohistochemical pattern was observed, suggesting recurrence of a previous misdiagnosed HPC with bone and lung metastasis. Chemotherapeutic treatment with adriamycin was performed with decrease of pulmonary lesions diameter and stability of bone metastases on a 3-month computed tomography re-evaluation.

HPCs are rare tumors which arise from pericytes around blood vessels and capillaries and can be located anywhere in both central nervous system (CNS) and extra-CNS sites. However, most HPCs are found in the musculoskeletal system and skin, while CNS ones are rare, and account for 2–4 % of all meningeal tumors and <1 % of all CNS tumors. In the CNS, this tumor is more commonly found in the cranial cavity, whereas spinal involvement is rare. HPCs were first described by Stout and Murray in 1942. Initially, they were classified as a variant of meningiomas; however, their similarity to peripheral soft-tissue tumors led to a separate classification by the World Health Organization. HPCs can resemble meningiomas on imaging and even microscopically, leading, like in our case, to frequent misdiagnosis. However, unlike usual benign meningiomas, which rarely metastasize extracranially, meningeal HPC is a locally aggressive, malignant tumor, with a high rate of local recurrence and distant metastases, which may occur several years after initial treatment. Microscopically, these tumors, which are derived from the mesenchymal external layer of blood vessels basement membrane, could be distinguished from meningiomas by vimentin staining, which is specific to mesenchymal tissue; moreover, HPCs are negative for epithelial membrane antigens, which are typically positive in meningiomas [1]. Surgery is the primary treatment of choice for meningeal HPCs. The excision should be as complete as possible to reduce the chances of

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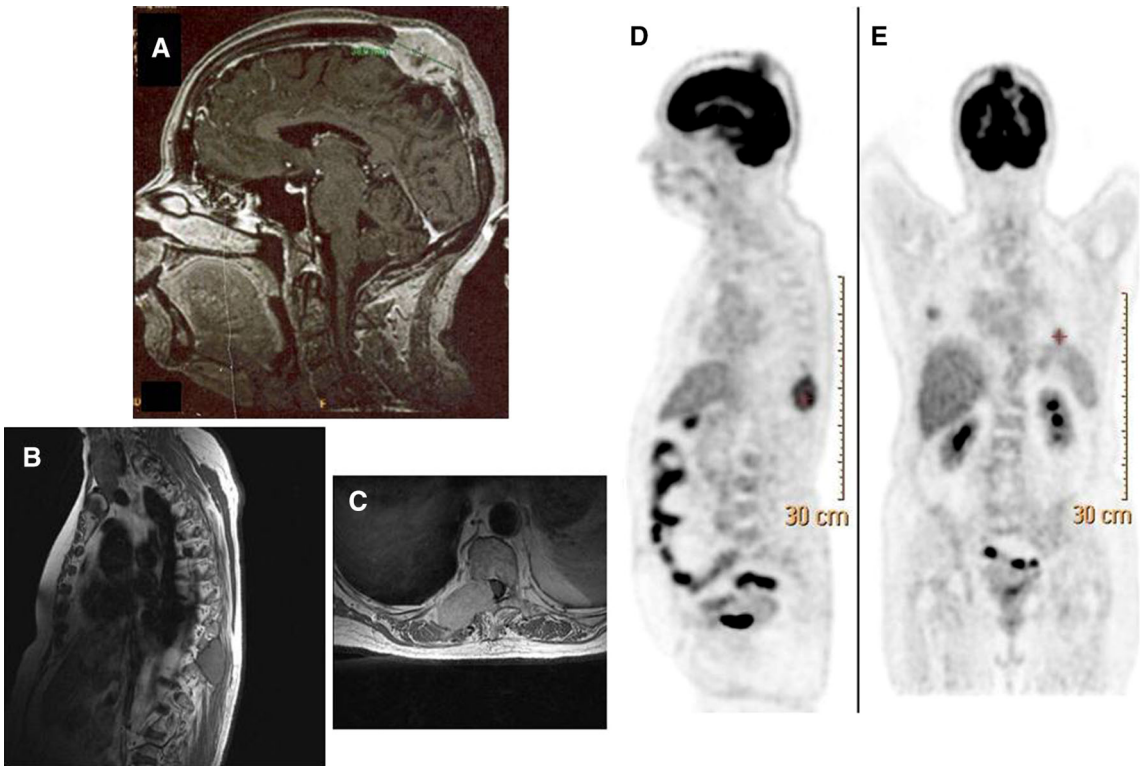

Fig. 1 (**a**) Sagittal image of a gadolinium-enhanced brain magnetic resonance showing an intracranial lesion of about 3 × 3 cm close to the meningeal falx. Sagittal (**b**) and axial images (**c**) of a gadolinium-enhanced magnetic resonance of dorsal and lumbar column demonstrating a T10 nodular vertebral-epidural lesion with spinal compression. Positron emission tomography sagittal (**d**) and coronal (**e**) scans showing areas of increased tracer uptake corresponding to the intracranial, spinal, pulmonary and fifth right rib lesions

recurrence or metastasis. Because the tumors exhibit high vascularization, the excision may lead to substantial blood loss. Therefore, some authors suggested that preoperative embolization may be of benefit for total resections by reducing bleeding [2]. Given the high risk of local recurrence in patients with both total and subtotal tumor resections, adjuvant radiotherapy is often applied, although no reduction in the rate of metastasis or survival benefit has been demonstrated [3]. Stereotactic radiosurgery has also been used for patients with a high surgical risk [4]. There is no standard treatment for patients with advanced metastatic disease. Indeed, various chemotherapeutic agents have been used, but no consensus exists on the most appropriate drug, dose, scheme of administration, or response criteria. Anthracycline, such as adriamycin, alone or in combination, appear to be the most effective agents, and are largely used in first chemotherapy line. Based on the fact that expression of vascular endothelial growth factor receptor (VEGFR) and platelet-derived growth factor receptor (PDGFR) has been detected by immunohistochemistry in the tumoral cells of HPC, anti-angiogenic agents, such as interferon-α, thalidomide, and tyrosine kinase inhibitors (sunitinib, sorafenib and dasatinib) have been used as first-line or successive treatments in patients with advanced disease, achieving different results in terms of response and disease stability [5]. Given the high rate of local recurrence and distant metastases, which often occur many years after the initial treatment, careful long-term follow-up is necessary for all HPC patients.

Conflict of interest None.

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