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LETTER TO THE EDITOR

Primary dedifferentiated liposarcoma of the dura mater: case report

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Dear Editor,

We report the case of a 62-year-old man who was referred to our institution in June 2008 for a depression of the temporoparietal cranial vault on the right side. This deformation had been known for 10 years with no occurrence of neurological symptoms.

Lateral X-ray of the cranium showed an extensive zone of osteolysis of the temporoparietal area on the right side. A brain computed tomography (CT) showed an osteolytic lesion of the parietal bone, involving the inner table and extending to the diploe and the outer table with presence of fat. Brain magnetic resonance imaging (MRI) demonstrated a T2 hypersignal tumor mass. The lateral part of the lesion displayed a T1 hypersignal, and a low signal on the fat-saturated, T1-weighted sequence, signaling the existence of fat within the tumor. No brain edema or brain displacement, dural thickening or abnormal enhancement was observable.

The surgical resection was justified by the recent progression of the lytic process as shown on sequential plain radiographs. Peroperatively the dura mater was involved by a yellowish compact tumor mass. After the resection, no evidence of leptomeningeal and subpial infiltration was observed. While complete removal was achieved, closure and reconstruction were obtained by a duraplasty (Goretex) and a methylmetacrylate cranioplasty. The size of the resection specimen was 5 cm. The patient had microscopic complete resection. Microscopically, with HES (hematoxylin-eosin-saffron) coloration ×100, the tumor was composed of a proliferation of spindled cells arranged in a myxoid or fibrous background. Tumorous cells were mildly atypical and showed few mitoses corresponding to the low-grade dedifferentiated component. Very focally, one could observe well adipose tissue with eccentric hyperchromatic nuclei and vacuolated cytoplasm corresponding to the well-differentiated component of liposarcoma. Few areas of reactive osteogenesis were present as the tumor infiltrated bone. According to immunohistochemistry, the spindle cells were negative for α-smooth actin, CD34, PS100, CD68 and EMA. Focal nuclear positivity was seen with anti-CDK4 antibodies in well-differentiated adipocytes and in some spindle cells. MDM2 showed more widespread nuclear staining in spindle cells. Amplifications of the oncogenes CDK4 and MDM2 were detected by FISH (fluorescent in situ hybridization) performed on representative paraffin sections (Fig. 1). The final diagnosis was consistent with a primary low-grade dedifferentiated liposarcoma of the dura. Full staging imaging with CT did not show any additional tumor mass.

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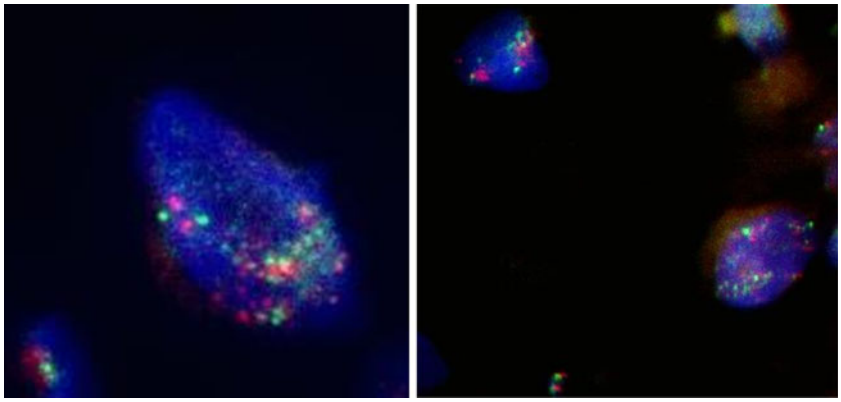

Fig. 1 FISH (fluorescence in situ hybridization): MDM2 amplification (*green*) and CDK4 amplification (*red*)

No adjuvant treatment was undertaken. Head CT and MRI performed at 46 months after surgery did not reveal local recurrence, and the thoracic CT scan did not show pulmonary metastases.

Compared to primary mesenchymal tumors, malignant intracranial neoplasms such as liposarcomas are rare [2, 6, 9, 10]. Since 1970 only three other cases of dural liposarcomas have been reported in the literature. The most frequent location of dedifferentiated liposarcomas is in the retroperitoneal area [3, 7].

We report here the first case of dedifferentiated primitive liposarcoma of the dura that has been documented with immunohistochemical and genetic data. Immunohistochemistry displayed staining of the CDK4 and MDM2 proteins, and FISH amplification confirmed overexpression of CDK4 and MDM2, which is commonly found in dedifferentiated liposarcomas and well-differentiated liposarcomas. Amplification of both genes is absent in benign lesions [1] and in pleomorphic liposarcomas [5].

The differential diagnosis includes the reactive process, such as fasciitis with fat involvement, fibromatosis or spindle cell lipoma, and most other spindle cell/pleomorphic undifferentiated tumors [1]. However, making the diagnosis of dedifferentiated liposarcoma is difficult and time consuming.

Surgical removal is the treatment of choice for liposarcoma. The quality of resection is the main predictor of local recurrence and of the overall survival [8]. Biomolecular confirmation of malignant tumor is an argument for a reoperation in case of invasion of the resection margins. Dedifferentiation is correlated with poor outcome. Unlike well-differentiated liposarcoma, both low- and high-grade dedifferentiated liposarcomas are subject to metastasize in 15 % of cases [4]. Dedifferentiated liposarcomas are characterized by high local recurrence even decades after of the initial diagnosis. Therefore, we recommended long-term monitoring of these patients. In the present case research on MDM2 and CDK4 amplifications has changed the monitoring because of the risk of local recurrence and metastatic dissemination. Almost 4 years after the resection, there has been no recurrence of the tumor.

Conflicts of interest None.

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