used in symptomatic epilepsy, SAM automated analysis appears to offer improved detection of irritable zones and beneficial volumetric and frequency descriptions compared to conventional dipole modeling.[2]

Our findings are consistent with those of other investigations,[12] which have provided evidence suggesting that large tubers correlate better with the EEG foci than small tubers; our SAM(g2) findings revealed that the epileptic activity in this patient arose from a large tuber and surrounding tissue. However, a large tuber involving the right frontal area failed to exhibit epileptogenicity. Frontal tubers have been reported to have the weakest correlation with epileptogenic foci.[13]

SAM(g2) analysis was a useful tool for localizing the epileptogenic tuber in this patient. This suggests that the combination of the brain anatomical data provided by MRI with the functional images of MEG and SAM(g2) analysis may help identify epileptogenic zones, and differentiate between epileptogenic and nonepileptogenic tubers in patients suffering from tuberous sclerosis. This may be particularly important when evaluating intractable epileptic patients for surgical treatment. Further application of SAM(g2) in large samples of patients with tuberous sclerosis and other focal epilepsies with multiple brain structural lesions may confirm this argument.

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Primary intracranial low-grade fibromyxoid sarcoma (Evans tumor)

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Abstract

Low-grade fibromyxoid sarcoma was first described in 1987 as a rare soft tissue neoplasm characterized by a bland and deceptively benign histological appearance but with aggressive behavior. A 20-year-old male patient presented with a recent history of headache and seizure. A right frontal mass was detected on MRI and he was operated upon to remove the intracranial mass. Histological examination revealed mildly atypical fibroblastic cells embedded within a myxoid matrix. Nuclear atypia and pleomorphism were minimal, and necro-

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sis was not present. The lesion was diagnosed as a low-grade fibromyxoid sarcoma. Although primary intracranial low-grade fibromyxoid sarcoma has characteristic histological features, clinical and radiological correlation is necessary to make the correct diagnosis.

Keywords: Low-grade fibromyxoid sarcoma; Primary intracranial; Spindle cell tumor

1. Introduction

Primary intracranial sarcomas are very rare. Paulus et al.[1] reported 19 primary sarcomas out of a total of approximately 25,000 brain tumors (<0.1%). Low-grade fibromyxoid sarcoma (LGFMS) is a rare sarcoma with a deceptively benign histological appearance. However, these tumors are associated with a high local recurrence rate, and distant metastasis, mostly to the lungs, may be seen. Described for the first time in 1987 by Evans, this particular neoplasm has only been recognized widely during recent years.[2,3] LGFMS typically presents as a deep intramuscular soft tissue mass in the lower extremities or trunk. LGFMS often presents non-distinctive cytological and histological features. This tumor is most commonly seen in young adults.[4]

To our knowledge, only a single case of primary intracranial LGFMS has been previously reported.[1] Treatment of this malignant tumor consists of surgical resection and it is regarded as being refractory to the radiotherapeutic and chemotherapeutic regimens currently used for soft tissue sarcomas.[5] Here, we present a case of primary intracranial LGFMS, emphasizing its clinical, radiological and histological features.

2. Case report

A 20-year-old male patient was referred to our clinic with a 2-month history of progressive headache and seizure. On admission, the results of a neurological examination were normal. There was no relevant family history and no past history of significant disease existed. Blood chemistry and complete blood count values were within normal limits and a chest X-ray revealed no abnormality. Cranial MRI revealed a huge contrast-enhancing mass in the right frontal region (Fig. 1). The patient underwent right frontal craniotomy and a well-circumscribed mass, which was attached firmly to the falx, was totally excised. Intra-operative frozen section examination of the surgical specimen revealed nothing contributory. The postoperative course was uneventful.

Grossly, the tumor was an irregular tan-gray mass of $3.5 \times 2.5 \times 1.5$ cm. On the cut surface, focal areas of glistening grayish-white substance were admixed with poorly demarcated firm areas. Neither necrosis nor hemorrhage was present. Microscopic examination revealed that the tumor was composed of bland spindle-shaped cells with indistinct pale eosinophilic cytoplasm and small hyperchromatic oval nuclei. The cellularity was low to moderate. In the more cellular areas, the cells showed nuclear pleomorphism with high levels of mitotic activity (Fig. 2A,B). The tumor cells were embedded in a variably fibrous or myxoid stroma that tended to alternate in different areas of the tumor. A prominent network of branching capillary-size blood vessels was also seen. Reactive gliosis was observed in the adjacent brain parenchyma. Immunohistochemical analysis of the tumor cells revealed diffuse expression of vimentin, but the cells did not express glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), smooth muscle actin or S-100 protein. The histopathologic and immunohistochemical findings were compatible with a diagnosis of low-grade fibromyxoid sarcoma.

Detailed examinations such as abdominal and thoracic CT scans, abdominal ultrasonography, whole bodys cintigraphy and X-rays were performed in an attempt to find a possible primary origin. No other tumor was found and the lesion was accepted as primary. Four months later, the patient complained of headache and an MRI examination revealed a mass in the left frontal region without any recurrence at the primary site (Fig. 3). The patient underwent left frontal craniotomy and the tumor was excised totally with the adjacent falx. Radiotherapy and chemotherapeutic regimens were started after the second operation. Intracranial recurrences continued to develop for

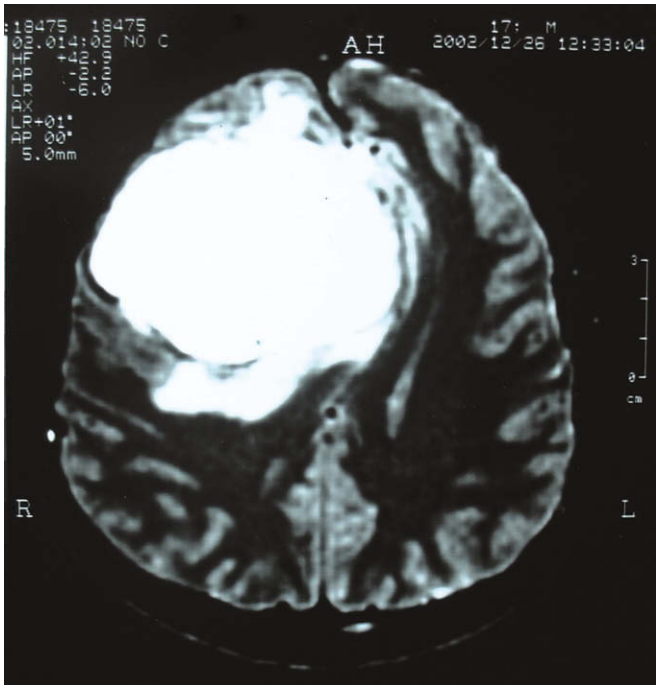

Fig. 1. T_2-weighted axial MRI showing a huge right frontal mass and midline shift.