bony. Desmoplastic fibromas may occur at any age range, although its higher incidence is observed at the first three decades of life\textsuperscript{(1–3,6)}. Despite conflicting data, it seems there is no predilection for sex\textsuperscript{(2,6)}. Local recurrence is frequently observed in cases where complete resection is not. Clinically, the patients are either asymptomatic or may present with pain, edema, joint effusion and pathological fracture\textsuperscript{(1–6)}. The differential diagnosis should consider rhabdomyosarcoma, fibrosarcoma, giant cell tumor, among others. Despite the imaging methods usefulness in the lesion delimitation, the diagnosis is histopathological.

At MRI, most lesions present with iso/hypointensity on T\textsubscript{1}-weighted images and low signal intensity on T\textsubscript{2}-weighted images\textsuperscript{(1–3,6)}, but there are reports of lesions with hypersignal on T\textsubscript{2}-weighted images\textsuperscript{(1–3,6)}. The enhancement may be variable, and according to some authors, such variation may be a result of the cellular content of the lesion\textsuperscript{(3,4)}. In the present case, there was homogeneous iso/hypointensity on T\textsubscript{1}-weighted images and subtle hypersignal on T\textsubscript{2}-weighted images, with foci of low signal intensity. After gadolinium injection, marked contrast enhancement, with noticeable perineural dissemination through the third division of the trigeminal nerve were observed. Such aspects on T\textsubscript{2}-weighted sequences, and the presence of perineural dissemination are not commonly observed as compared with the typical imaging pattern described at MRI.

Reports on diffusion in desmoplastic fibromas were not found in the literature. In the present case, areas of diffusion restriction were not observed. Recent studies highlight the use of diffusion-weighted imaging in the evaluation of head and neck lesions, showing that apparent diffusion coefficient $< 1.22 \times 10^{-3} \text{mm}^2/\text{s}$ are suggestive of malignancy\textsuperscript{(7)}. In the present case, the value for apparent diffusion coefficient was $1.45 \times 10^{-3} \text{mm}^2/\text{s}$, corroborating the previously described findings.

The authors conclude that the diagnosis of desmoplastic fibromas should be considered in patients under the age of 30 presenting with tumor particularly located in the mandible, and that such a hypothesis cannot be ruled out in case of less noticeable foci of hypointensity on T\textsubscript{2}-weighted images.

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**Dear Editor,**

A 72-year-old woman with rapidly progressive dementia, behavioral changes and apraxia of gait for seven months, extrapyramidal signs and diffuse myoclonus. Electroencephalography demonstrated periodic electric activity with high amplitude acute phase waves diffusely distributed over the cortex. The cerebrospinal fluid was normal. Magnetic resonance imaging (MRI) was performed (Figure 1).

The association of clinical, radiological, electroencephalographic or cerebrospinal fluid findings (presence of 14-3-3 brain protein in diseased patient for less than two years – absent in this case),

### Creutzfeldt-Jakob dementia

*Demência por doença de Creutzfeldt-Jakob*

**Dear Editor,**

A 72-year-old woman with rapidly progressive dementia, behavioral changes and apraxia of gait for seven months, extrapyramidal signs and diffuse myoclonus. Electroencephalography demonstrated periodic electric activity with high amplitude acute phase waves diffusely distributed over the cortex. The cerebrospinal fluid was normal. Magnetic resonance imaging (MRI) was performed (Figure 1).

The association of clinical, radiological, electroencephalographic or cerebrospinal fluid findings (presence of 14-3-3 brain protein in diseased patient for less than two years – absent in this case),

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**Figure 1.** A: Axial magnetic resonance imaging of the skull demonstrating foci of hypersignal at diffusion-weighted sequences in the heads of the caudate nuclei, putamina, thalami and medial occipitotemporal gyri. B: At the ADC mapping, the low signal intensity in the same region confirms the diffusion restriction.

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allows for a probable diagnosis of Creutzfeldt-Jakob dementia (CJD). The differential diagnosis is made with other diseases associated with dementias, as follows: a) Alzheimer’s disease, that does not present with alterations observed at diffusion-weighted images; b) vascular dementia, with multiple infarcts, but with abnormalities at diffusion-weighted images only in cases of recent infarcts, and without diffuse cortical involvement; c) other encephalopathies that present alterations restricted to the cortex at diffusion-weighted images (such as MELAS – mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes – a genetic metabolic disease occurring in younger patients); venous hypertensive encephalopathy and chronic herpes simplex encephalitis.

CJD is a subacute spongiform encephalopathy that presents with rapidly progressive dementia, and is the most frequent among rare prion diseases. Myoclonus, pyramidal, extrapyramidal and cerebellar signs are associated. Psychiatric symptoms are observed at the first six months; and progressive immobility, cortical blindness, dysphagia and mutism constitute late signs of the disease. Death generally occurs one year after the symptoms onset, since there is no therapy to prevent a fatal outcome. The disease is subdivided into sporadic (85% of cases), familial, iatrogenic and a less common, recently described variant related to epidemic bovine spongiform encephalopathy.

Like in other acute encephalopathies, the 14-3-3 brain protein may be present in the cerebrospinal fluid. Encephalography may demonstrate periodic activity composed of three-phase high-frequency waves over attenuated background activity.

At MRI, hyperintense signal is observed in the basal ganglia, putamen and later in the cerebral cortex on T2-weighted and FLAIR sequences. Such alterations suggest CJD, more than any other dementia disorder. At early stages of the disease, however, conventional imaging studies may present normal results. Diffusion-weighted sequences may favor an early diagnosis, demonstrating abnormal hyperintense foci even before the appearance of electroencephalographic alterations, and should be performed in case of suspicion of CJD. Diffusion restriction is observed, probably in association with the cytotoxic edema secondary to spongiform degeneration and to accumulation of abnormal cytoplasmic vacuoles. As the disease progresses, global atrophy is observed and, in general, this is the only alteration depicted at computed tomography.

Histopathological analysis (Figure 2A) demonstrated spongiform alterations with variable intensity in the neuropil, markedly in the caudate nucleus, putamen, and in the region CA1 of the hippocampus, moderately in the frontal and temporal cortices, and slightly in the parietal and occipital cortices. Immunohistochemical analysis demonstrated gliosis (Figure 2B). Such results are compatible with a definite diagnosis of CJD. The lesions distribution, the absence of similar cases in the family, and the absence of a known infectious source are compatible with the sporadic presentation of the disease. Such a diagnosis is relevant for controlling the transmission and to rule out treatable causes of dementia.

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Figure 2. A: Section of the caudate nucleus demonstrating abundant small, optically empty vacuoles in the gray matter. The characteristic architecture of the caudate nucleus is highlighted by parallel white matter tracts through the gray matter (400×). B: GFAP immunohistochemistry demonstrating reactive astrocytes, a finding compatible with spongiform encephalopathy (400×).

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