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Creutzfeldt-Jakob Disease Transmitted by Dura mater Graft

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We report a 41-year-old woman who underwent a craniotomy and removal of a left clinoidal meningioma invading the cavernous sinus in May 1989. A graft of cadaveric dura mater (lyodura) was used as plastia, covering part of the left temporal lobe. A small tumoral fragment remaining was treated with radiotherapy. She stayed asymptomatic until October 1992 (41 months after surgery) when she began a rapidly progressive clinical deterioration (ataxia, dysarthria, pyramidalism, myoclonus and generalized slowing on the EEG). Two months later, a brain tissue frontal biopsy (fig. 1) led to the diagnosis of spongiform encephalopathy, thus suggesting Creutzfeldt-Jakob disease (CJD). The patient went into a progressive coma dying in February 1993.

The first description of a transmissible, person-to-person form of CJD was reported in 1974 in a patient who had received a corneal transplant [1]. Thadani et al. [2] published the first report of CJD transmitted by a cadaveric dura mater graft in 1988. Since then, 14 more cases [3–5] including ours have been reported (table 1). All of them, except 1, related to a commercial dura mater (Lyodura) made by Brown Melsungen Company of Germany.

CJD can be transmitted through particles containing an abnormal form of a prion protein. These infectious particles are highly resistant to most of the common sterilization procedures. The Committee on Health Care Issues of the American Neurological Association [6] recommended 1-hour exposure to 1 molar NaOH, or steam autoclaving for 1 h at 132 ° C as the standard sterilization procedures for CJD tissue or contaminated material. After May 1987, according

Fig. 1. Spongiform change, neuron loss, and astrocytosis in the brain biopsy. HE. x 160.

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Table 1. Dura mater graft cases

Age onset
Sex
Latency
Original surgery
Initial symptoms

Thadani et al, 1988 [2] Nisbet et al, 1989 Masullo et al., 1989 Miyashita et al., 1991 Willison et al., 1991 Pocchieri et al, 1992 Esmonde, 1993 Esmonde, 1993 Martinez-Lage et al., 1993 [4] Martínez-Lage et al, 1993 [4] Martinez-Lage et al, 1993 [4] Martinez-Lage et al, 1993 [4] Lancetta, 1994 Yamada et al., 1994 [5] Present case, 1995				
28	F	19 months	temporal cholesteatoma	unsteady gait, slurred speech
25	M	31 months	head injury	dementia
27	M	44 months	fibrose dysplasia	confusion, gait disturbance
26	F	33 months	cerebellar hemangioblastoma	unsteady gait
32	F	47 months	syringomyelia	dementia
32	F	10 years	ethmoidal meningocele	ataxia
51	F	84 months	syringomyelia	mood change
44	F	96 months	meningioma	visual symptoms
19	M	16 months	cerebellar astrocytoma	ataxia
57	F	43 months	syringomyelia	ataxia
18	M	79 months	cerebellar astrocytoma	ataxia
34	M	105 months	syringomyelia	dizziness and slurred speech
28	F	60 months	Arnold-Chiari	cerebellar dysfunction
31	F	56 months	pituitary adenoma	slurred speech, unsteady gait
41	F	42 months	meningioma	instability

Cadaver lyodura was used in all cases. Lyodura lot No. 2105 used. Noncommercial lyodura used.

to Braun Melsungen AG, their procedures of collection and processing of dura mater were revised in order to reduce the risk of CJD transmission, including the manufacturing process of exposure for 1 h to 1 molar NaOH. However, some authors [7, 8] have emphasized that complete inactivation of the agent producing CJD cannot be obtained, and that such treatment merely prolongs the incubation period. The epidemic proportion of this group of patients with CJD transmitted by dura mater graft could be larger if we think of the potential transmission of the disease to patients with severe head injuries and malignant brain tumors who, due to the short period of survival after surgery, do not surpass the theoretical incubation period of CJD. These groups of patients are the major working load of any neurosurgical department. Brown et al. [9] have shown in a recent work that allelic homozygosity at polymorphic codon 129 was present in 92% of the patients with CJD studied. This supports the thesis that the genotype of codon 129 enhances the susceptibility to CJD iatrogenic infections. We agree with others [10] that despite the assurance on the safety of the product treated with 1 molar NaOH, this homograft source should be abandoned, and we advocate preferential use of temporalis fascia, fascia lata, pericranium or synthetic substitutes.

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