CREUTZFELDT-JAKOB DISEASE IN VENEZUELA

A CASE REPORT

*ALEJANDRO J. CARABALLO H.**

SUMMARY — A case of Creutzfeldt-Jakob disease (CJD) in a 32 year old man is presented. The clinical picture included a rapid progressive dementia associated with ataxia, global aphasia, myoclonus and pyramidal signs, death ocurred after 13 months. The diagnosis of CJD was confirmed by CT and neuropathological studies. This is the first report of CJD occurring in Venezuela.

Doença de Creutzfeldt-Jakob na Venezuela: registro de caso.

RESUMO — Registro de caso de doença de Creutzfeldt-Jakob (DCJ) em um paciente com 32 anos de idade. O quadro clínico abrangia demência rapidamente progressiva associada a ataxia, afasia global, miocloniais e sinais piramidais. A morte ocorreu após 13 meses. O diagnóstico foi confirmado por dados dia TC e do estudo neurcpatológico. Este é o primeiro relato de DCJ na Venezuela.

Creutzfeld-Jakob disease (CJD), a rare disorder, is attributed to a slow virus infection. Clinico-pathological aspects of the first case of CJD observed in Venezuela aie **reported in this paper.**

CASE REPORT

NAM, a 32-year-old heavy machinery operator, according to references from relatives, presents since 2 months ago a frame characterized by: gradual loss of recent memory, absent-mindedness concentration, irritability and incompetence to carry out everyday actions. One month later, he is admitted into Caracas University Hospital where a cardiopulmonary evaluation and laboratory routine were performed, with results within normal values. A CT scan of the brain was performed showing a moderate increase of the silvian fissures (Fig. 1*). He is discharged against medical opinion. Two months later, he starts to show changes in his behavior, developing paranoid and delirious ideas, with visual and auditory hallucinations, lacking introspection. As the disease progresses, he loses all his faculties, not being able to recognize the members of his family. He presents dysphagia, aphasia and incontinence of urine, being admitted at the Maracaibo Psychiatric Hospital where he stayed for 15 days showing no general changes. Seven months after onset of the disease he presents progressive and insidious ataxic march, spasticity and generalized myoclonia. His physical condition deteriorates gradually until he loses his capacity to move by himself staying permanently in bed.

Eleven months after onset of the disease, he is sent to the Southern General Hospital in Maracaibo. He was a non-drinker and non-smoker. There was no history of tuberculosis, hypertension, diabetes mellitus and no positive family-history of any neurological disease.

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Physical examination: admitted in bed, stupored state, fever of 39t>C, blood pressure of 100/70mmHg. The addominal and cardiopulmonary examination showed no abnormalities. Presence of decubitus scars in the gluteal region, and lower limbs. Neurologic examination: hypertonic, spastic, muscle masses atrophied, abulic, uttering guttunal sounds, motor sensible aphasia, hyperephlexic, abdominal reflexes abolished, Hoffmann bilateral negative, Babinski bilateral positive and positive succedaneious, suction and prehension reflexes present, palmomentuial reflex present, positive clonus in hands, knees and feet. Complementary examinations: blood count and usual biochemical examinations normal; a second CT scan of the brain revealed a marked increase in the cortical grooves and silvian fissures, lateral ventricles and the third ventricle show lan increase in volume (Fig. lb), and an image compatible with a subdural collection may be seen on the right frontoparietal region (Fig. lc). The patient died from intercurrent pulmonary disease 13 months after onset of clinical frame.

Neuropathology — Encephalon weighting 1050 g, presenting a marked diffuse cortical atrophy of the convolutions. Coronal sections showed marked thinning of the cortical ribbon

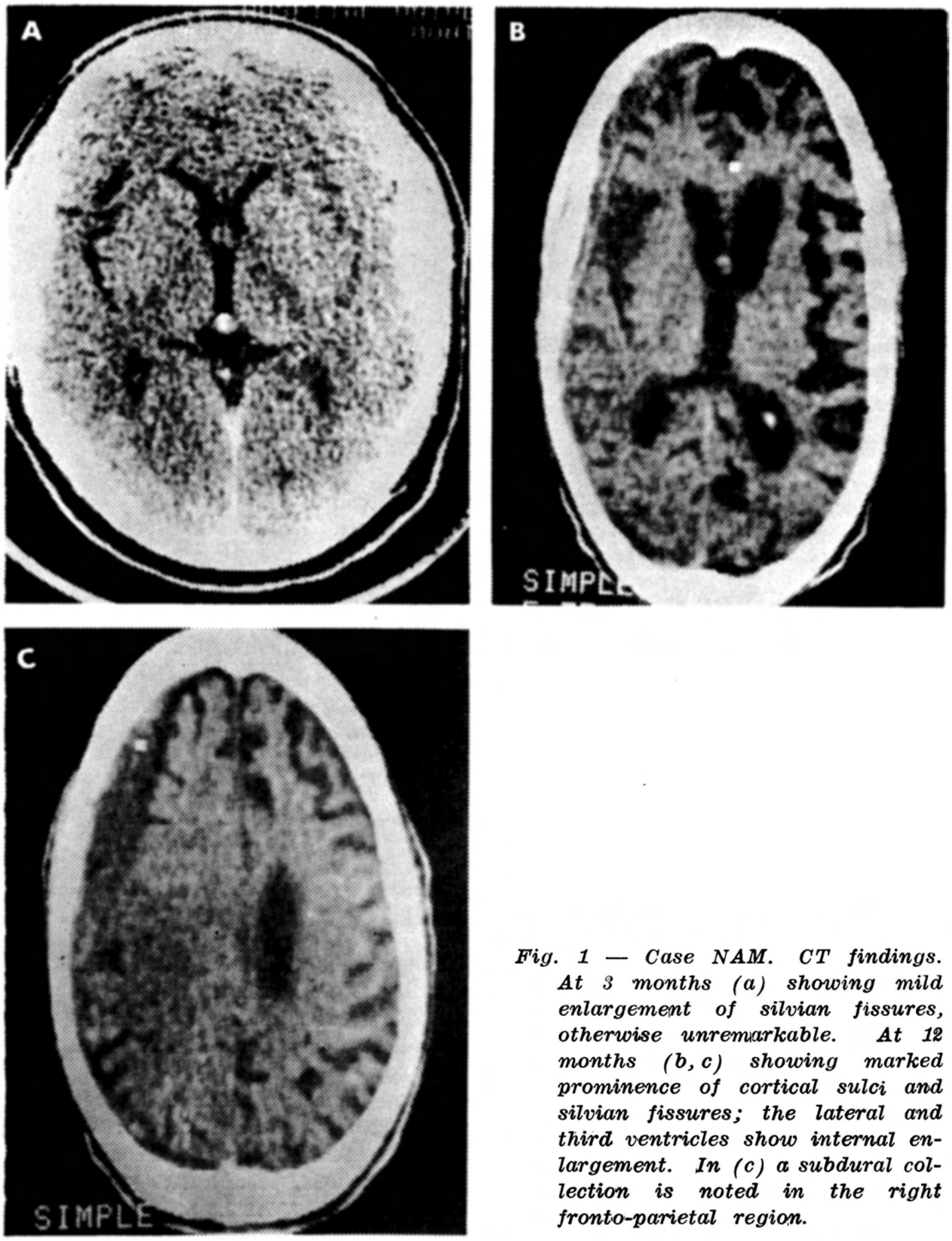

Fig. 1 — Case NAM. CT findings. At 3 months (a) showing mild enlargement of silvian fissures, otherwise unremarkable. At 12 months (b, c) showing marked prominence of cortical sulci and silvian fissures; the lateral and third ventricles show internal enlargement. In (c) a subdural collection is noted in the right fronto-parietal region.

which wias granular in appearance and dilatation of the ventricles. The basal ganglia, particulary the caudate nuclei were atrophic. Microscopy revealed the characteristic spongiform change, neuronal loss and gliosis of the cerebral cortex (Fig. 2).

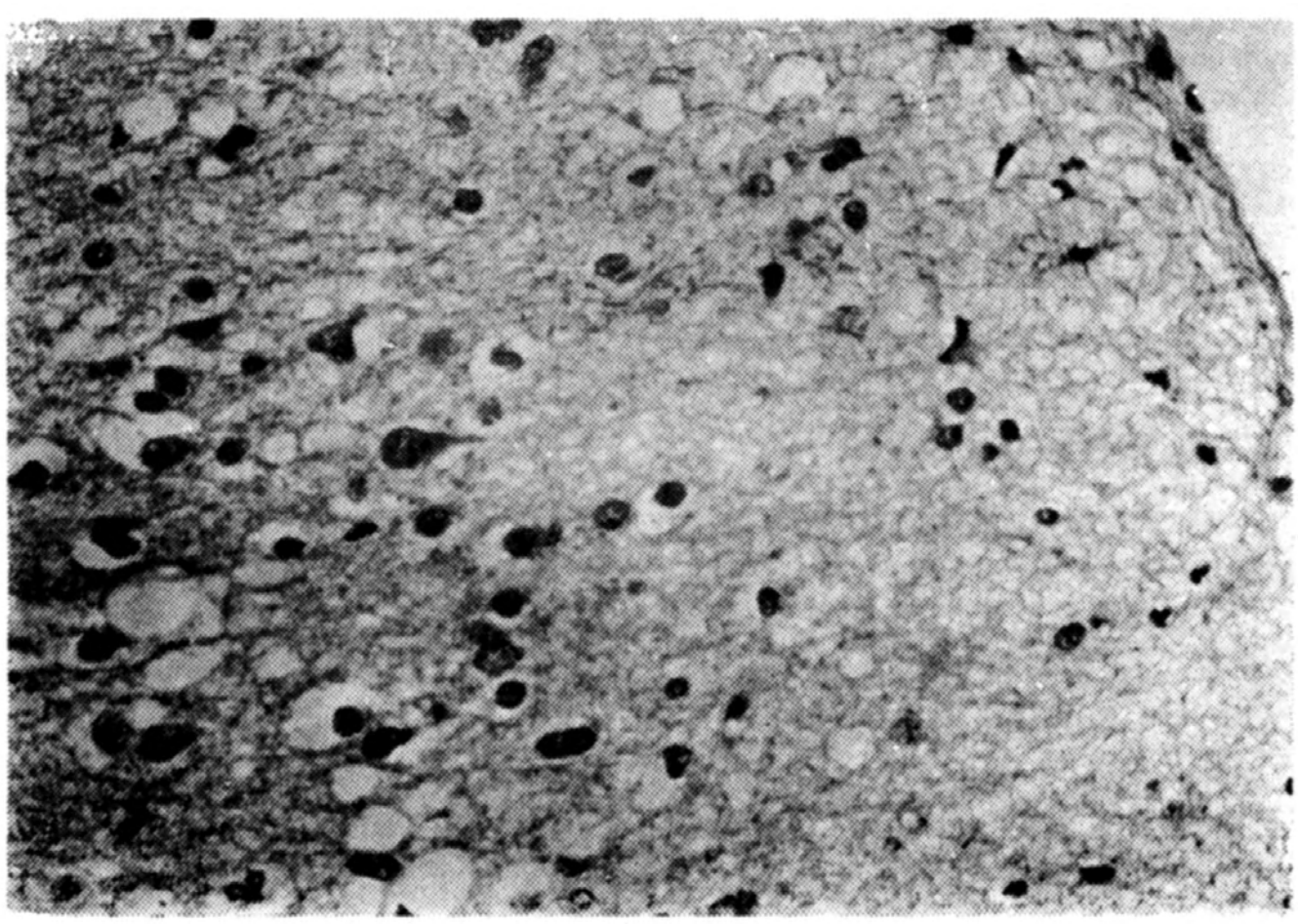

Fig. 2 — Case NAM. Neuropathology. Cerebral cortex: characteristic spongiform changes. HE stain (×150).

COMMENTS

'1 he recent reports of young patients with CJD after human growth hormone therapy (prepared from pools of pituitary glands obtained at autopsy) is alarming, and probably new cases will be described 3,6,8,13.

CJD is a well-known clinico-pathological entity 2,4,11,14-17 -with a worldwide distribution, and an incidence of about one per million 10. Argentina[12], Brazil 1.9 and Chile 5 are some South American countries where the disease has been recognized. We present the clnico-pathological aspects of the first case of CJD in Venezuela.

Clinical picture of the patient included rapid progressive dementia associated with global aphasia, ataxia, myoclonus and pyramidal signs. Diagnosis was confirmed uy ti and neuropathological findings.

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