

## Case Report

# Creutzfeldt-Jakob Disease after Dental Procedure along with the Initial Manifestations of Psychiatric Disorder: A Case Report

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### Abstract

**Introduction:** Creutzfeldt-Jakob disease (CJD) as a prion disease is an untreatable type of progressive neurodegenerative encephalopathy. Although no definitive case has been reported yet, here we report a case that given the history, course of symptoms, and recent dental practice, it is highly probable that it was caused by dental procedures.

**Case Report:** The patient was a 52-year-old woman who has had memory problems gradually with forgetting the names of family members since 6 weeks prior to the visit and shortly after the dental procedure. She experienced progressive visual hallucinations accompanied by a sharp decline in cognitive, verbal, and motor abilities in just a few weeks. Finally, the diagnosis of Creutzfeldt-Jakob was made for her based on the clinical history and typical brain MRI.

**Discussion:** Clinical evidence of this patient, along with positive brain MRI results, indicates the risk of prion transfer through dental procedures. Paying attention to the neurological aspects of psychiatric manifestations and increasing the awareness of dentists about how to deal with and act on the potential dangers of prion transfer is of paramount importance.

**Key words:** *Creutzfeldt-Jakob Disease; Dental Procedures; Psychiatric Manifestation*

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**C**reutzfeldt-Jakob disease (CJD) as a prion disease is a type of untreatable progressive neurodegenerative encephalopathy. The prevalence of the disease is estimated at about one and a half cases per million per year (1). The presence of numerous vacuoles in the neurons and loss of nerve tissue in the gray matter creates the classic sponge-like appearance of the brain responsible for its typical clinical symptoms (2). The iatrogenic form of the disease, with a prevalence of 1% among other types, has the lowest prevalence. The duration of the disease in iatrogenic cases is 6.3 months (2 to 25 months) and 56% of them die within 6 months (1, 3).

Most of the iatrogenic cases identified have been due to growth hormone or dura mater grafts.

Other sources of iatrogenic transmission of the disease include contaminated blood, corneal transplants, and neurosurgical instruments, and stereotactic EEG depth electrodes (4).

The transmission of the disease through dental instruments has been a matter of serious concern due to the widespread use of these devices. Although no definitive case has been reported yet, here we report a case that given the history, course of symptoms, and recent dental procedure, it is highly probable that it was caused by dental procedures.

### **Case Report**

The patient was a 52-year-old married woman and a housewife who had memory problems gradually with forgetting the names of family members since 6 weeks prior to the visit and shortly after the dental procedure. The patient gradually developed visual hallucinations in her home in the form of seeing dead people as well as seeing animals, which caused distress and fear in the patient.

Sometimes she would talk irrelevantly and was disoriented to time, place, and person. The patient's relationship with those around her has gradually declined. At that stage, the patient was also evaluated by a psychiatrist, and acute mood problems were raised and even the need for electrical convulsions was proposed. The patient was then advised to take antidepressant medication but did not take them.

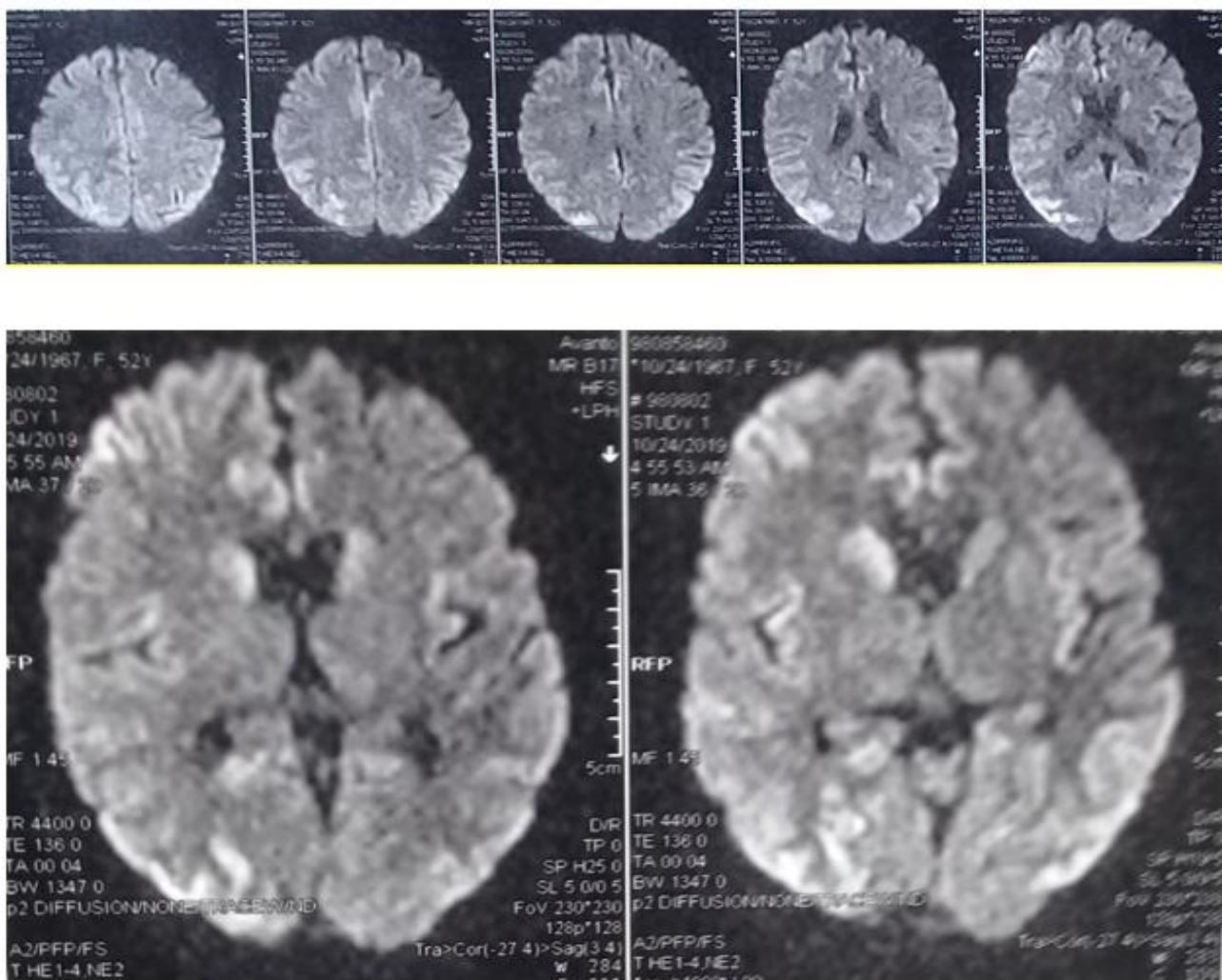
About 2 weeks later, the patient developed double urinary and fecal incontinence. Gradually the patient lost the ability to speak. The progressive course of symptoms was characterized by a complete lack of recognition of individuals and objects, a gradual inability to walk, and staying in bed all the time. Thus, the patient developed a lower lumbar bed sore within 2 weeks before recent hospitalization. Also, the patient gradually developed severe stiffness of the extremities and muscle jerking in the upper extremities.

On examination at the patient's arrival, the patient was in a stupor condition. She did not obey orders. The pupil reflex to light and fundoscopy was normal and symmetrical on both sides.

With painful stimulation, she moved her limbs briefly. Deep tendon reflexes of all 4 limbs were intensified and was +3. The plantar reflex was down. The limbs had severe and generalized stiffness. There was intermittent muscle jerking in both upper limbs.

The patient's parents were not relatives. The patient had no prior history of medication use, and there was no similar problem or any other neurologic problem in the patient's family.

Finally, the diagnosis of Creutzfeldt-Jakob was made based on the clinical history and clinical course of the disease and typical brain MRI (Figure 1). Other causes of rapid progressive dementia, such as vitamin B12 deficiency and brain infections (fungal, granulomatous, etc.), have been ruled out. Also, acute vascular disorders and metabolic and toxic problems were excluded in the patient. No signs of immunologic, demyelinating, or neurodegenerative disorders were found based on initial laboratory and imaging evaluations.



**Figure 1. Brain MRI-Diffusion Weighted Imaging (Hyperintensity and Hypersignality in Right Caudate Nucleus and Parieto- Occipital Cortex (Ribbon Sign)**

## Discussion

Prion diseases, which are degenerative disorders of the nervous system, are caused by transmissible particles that contain the pathogen isoform of the prion protein as a natural constituent of the cell membrane and are known as transmissible spongiform encephalopathy (TSEs)(5). The definite diagnosis of Creutzfeldt-Jakob disease is defined as using standard neuropathological techniques and/or immunocytochemically and/or Western blot confirmed protease-resistant prion protein and/or the presence of scrapie-associated fibrils. Probable cases, which are mainly considered in normal clinical conditions, are defined as follows:

(1) Cases who have neuropsychiatric disorder plus positive real time quaking-induced conversion (RT-QuIC) in cerebrospinal fluid (CSF) or other tissues; (2) rapidly progressive dementia; (3) and at least 2 out of the following 4 clinical features: (myoclonus, visual or cerebellar signs, pyramidal/extrapyramidal signs, akinetic mutism); (4) and a positive result on at least one of the following laboratory tests: (a typical EEG (periodic sharp wave complexes) during an illness of

any duration; (5) a positive 14-3-3 protein in CSF assay in patients with a disease duration of less than 2 years; (6) a high signal in caudate/putamen on magnetic resonance imaging (MRI) brain scan, or at least 2 cortical regions (temporal, parietal, occipital) either on diffusion-weighted imaging (DWI) or fluid attenuated inversion recovery (FLAIR) and without routine investigations indicating an alternative diagnosis (6).

Identifying cases of accidental transmission of CJD through medical or surgical procedures has raised concerns about secondary transmission of the disease. Also, evidence of the presence of pathogens in lymphoreticular tissue in some populations and based on some studies has raised concerns about widespread subclinical infection in some populations (7).

Reports of CJD transmission through medical devices (neurosurgery instruments, brain electrophysiology electrodes, human pituitary hormone, corneal transplantation, etc.), although declining in recent years, are alarming in number. There have also been reports of cases of CJD through blood transfusions (8).

Brown et al emphasized concerns in this regard, with the possibility of continued transmission of CJD through infected instruments. They emphasized the attention of patients with rapidly progressive dementia or cerebellar symptoms and the use of appropriate diagnostic methods for their early diagnosis (9).

Thomas et al indicated CJD is a neurodegenerative prion disease, which is transmissible by infected neurosurgery instruments previously used in affected individuals (10).

Although there are various reports on the duration of the iatrogenic CJD (iCJD) incubation period, the duration of 1 to 2.3 years has been suggested as the minimum time (11). However, in some iCJD animal models, incubation time of about 170 days is also mentioned (12).

Laboratory studies show that standard methods of sterilization and disinfectant may not be sufficient to completely eliminate contamination from prion-contaminated instruments. In this unfortunate event, the devices may pass on the prion to others (4).

Concerned about the transmission of the disease through dental procedures, there are unclear mechanisms for such a situation through the transmission of prions following those procedures. Adherence to the highest standard is required to control possible early infection and to use appropriate decontamination procedures (13).

In the study of a number of patients with suspected CJD variants in the UK, autopsies were performed from different areas to check for the presence of prion protein (PrP). They had positive results in the brain, trigeminal ganglia, and tonsils. Despite the lack of PrP in most dental and oral tissues, its presence in some of these areas seems to be a concern (14). This complicates the issue of PrP transfer due to the possibility of secondary involvement of related lymphatic tissues and the risk of damage to them at different stages of dental procedures. There has been no reported case of definitive dental transmission of prion disease, but since it is theoretically possible to transmit it through medical instrument, sufficient information from all patients before any dental procedures on the medical and family history of prion disease should be provided (15). Given the experiences of the possibility of transmitting the disease in animal models through dental procedures, it seems possible to endanger the transmission of the disease by endodontic instruments if there is no effective prion decontamination (16).

Based on existing experience, prion proteins are resistant to many disinfection and sterilization techniques. Inactivation of prions is not affected by such methods as irradiation, boiling, drying heat, and such chemicals as formalin, beta-propylactone, and alcohol.

Due to the potential risk of prion transfer during dental procedures, disposable dental instruments should be used for people who are likely to develop the disease.

Also, in cases where it is not possible to use disposable forms of equipment, they must first be cleaned by physical methods, and the equipment must be kept in hot 1N sodium hydroxide solution for 1 hour. Then, they should be placed under vacuum in an autoclave at 134 to 138 °C for 18 to 20 minutes (17).

In the presented case, we witnessed the clinical manifestations of rapidly progressive dementia, myoclonus jerking, akinetic mutism, and rigidity. Given the patient's history and lack of risky conditions prior to dental procedures in the field of prion diseases accompanied with lack of positive family history, progressive symptoms following dental procedure and the result of brain MRI[6], there is a high probability of diagnosis of Creutzfeldt-Jakob disease in the field of prion transfer during dental procedure. The neuropsychiatric manifestations that the patient has experienced have occurred without any previous history of psychiatric disorder. In addition, the patient had no history of previous neurological disorders. She had no history of epilepsy or cerebrovascular problems. There was also no family history of psychiatric or neurological disorders.

This report aims to warn about the need to obtain a history before dental procedures on the risk of developing CJD, observing the principles underlying the use of common equipment and strict adherence to the principles of sterilization based on the above.

Health policy-makers' attention to the necessity of education in this field for the target groups of dentists and related professionals seems to be necessary.

Also, as was seen in this patient, at a stage of illness, psychiatric symptoms such as visual hallucinations and decreased communication with others were the predominant aspect of the disease. This emphasizes the need to pay attention to the neurological aspects of psychiatric manifestations.

## Conflict of Interest

The authors declared no conflict of interest.

All identifying information about the case has been removed/ altered.

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