

## CASE REPORT

### Two Cases Of Variant Creutzfeldt-Jakob Disease (vCJD) Referred To The Department Of Community Mental Health, Aldershot Garrison In 2003

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#### ABSTRACT

In the year 2003 the Department of Community Mental Health (DCMH) at Aldershot Garrison received referrals of two soldiers, a sergeant and a lance corporal, who presented with a complex picture of neurological and psychiatric symptoms. Both had been investigated very thoroughly by neurologists who, owing to the mainly negative results of their investigations, were unable to make a diagnosis. Of the two patients one had also been assessed as a psychiatric in-patient in a civilian hospital and had been referred to the Aldershot DCMH for continuing care. The other had been transferred, after investigations, to the Defence Services Medical Rehabilitation Centre (DSMRC) at Headley Court but, failing to make progress, was also referred for psychiatric assessment. Both patients were obviously unwell but the nature of their illnesses remained obscure. Within a few months both had died and the diagnosis of one of the human transmissible spongiform encephalopathies (TSEs), vCJD, was made at autopsy in one of them, but both were registered by the National CJD Surveillance Unit (3) with a diagnosis of vCJD. The circumstances were so unusual and prompted the writing of this paper by one of the psychiatrists involved.

#### Introduction

The Transmissible Spongiform Encephalopathies (TSEs) (1) are a family of degenerative brain disorders which affect mammals including man and other primates, cattle as mad cow disease or bovine spongiform encephalopathy (BSE), sheep and goats (scrapie), members of the deer family, felines, mink, rodents etc. Widespread neuronal loss creates a spongiform degeneration together with astrogliosis and amyloid plaques. The disease can be transmitted from one individual to another and from one species to another by a process of direct contamination. Hereditary forms do also occur indicating a genetic aetiology in some cases. The transmitting agent has been identified as an abnormal variety of a normal tissue protein called the prion protein (PrP). It is found attached to all tissue cell surfaces but it predominates in the

brain. The normal PrP has a rapid metabolic turnover, is readily denatured by proteinase enzymes, is soluble in aqueous fluids and has a genetically determined amino-acid sequence. The abnormal PrP has an identical amino-acid sequence but a different folded structure. It is not broken down by proteases, is insoluble in aqueous or alkaline fluids and tends to aggregate. Because of its resistance to bio-degradation it is called PrP<sup>Res</sup>. It may also be labelled according to the host animal and called PrP<sup>Sc</sup>, PrP<sup>BSE</sup>, PrP<sup>CJD</sup> and so on. Neither the mechanism by which PrP<sup>Res</sup> becomes a tissue toxin, nor the mode of its creation has been explained. It is known that in some way PrP<sup>Res</sup> interacts with natural PrP to achieve toxicity. Experiments on mice which have been bred so that PrP is absent from their bodies, do not show neuro-degeneration when infected with PrP<sup>Res</sup> even though the latter persists in the body.

The PrP gene is located within a single exon of the short arm of chromosome 20 (2), and occurs in five variants controlled by codon 129 of the gene. PrP consists of either methionine or valine or both amino-acids and may be homozygous (MM or VV) or heterozygous (MV). It is known that vCJD has mainly occurred in individuals who are homozygous (MM) but the explanation for this is elusive. The disease is monitored by the national CJD Surveillance Unit and is reported on annually by the Department of Infectious and Tropical Diseases, London School of Hygiene and Tropical Medicine. (3).

The human forms of TSE include Gerstmann-Sträusler Syndrome, Fatal Familial Insomnia, Kuru, Sporadic CJD (sCJD) and variant CJD (vCJD) (4). Prion diseases were first identified in 1922 and the familial form Gerstmann-Sträusler Syndrome in 1936. In 1957 a neuro-degenerative disorder was seen in an isolated New Guinea tribe, the Fore Tribe, who practiced familial cannibalism, causing the disease to be transmitted to susceptible relatives. It was called Kuru, the trembling sickness, by the natives. Researchers noted similarities between kuru and scrapie. In 1966 kuru was successfully transmitted to monkeys after an incubation period of 2 years. The search for an infectious agent, such as a virus, failed but was narrowed down in 1967 to the prion protein. In humans the commonest CJD is the sporadic form.

## Incidence

sCJD is the basic form of the disease and has a world wide incidence of about 1 per million per annum, although minor regional differences are noted. It is distributed equally between the sexes and presents in individuals over the age of 40 years, mainly in the age group from 60 to 75 years, after a long incubation period. Once the symptoms appear it progresses to a fatal conclusion within about 5 months. The median age of death is 64 to 67 years. In 1996 it became evident in the United Kingdom (UK) that a new variant had emerged which affected younger people in the 16 to 40 year age bracket. It also ran a fatal course but with a duration of about 12 months. The median age of death is 28 years. This is vCJD.

vCJD is connected with the development in the UK of cattle suffering from BSE, which appeared in 1986 and reached its zenith in 1992, when 1,200 infected cattle were recorded per week. (5, 6,). vCJD was first diagnosed in 1992. The incidence assumed epidemic proportions and peaked in the year 2000 when 28 deaths were recorded. Between January 1997 and June 2005 156 deaths occurred in the UK. The rate in all other countries was much lower as was the BSE infection rate. It has been confirmed that vCJD was caused by the consumption of beef products from infected cattle. Later it was also confirmed that iatrogenic infections arose as the result of organ transplantations from infected individuals, blood transfusions, infusions of human growth hormones, and the use in surgery of instruments contaminated with PrP<sup>(Res)</sup> which had resisted standard sterilization procedures. PrP<sup>(Res)</sup> is very tough physically, it resists antiseptics and strong alkalines. In an autoclave it needs to be heated to 134°C for at least an hour for its destruction.

The susceptible UK population, aged 16 to 40 years, amounts to about 35 million, giving a maximum annual incidence of about 0.85 per million. The Armed Forces population falls into this susceptible bracket but its number is relatively small. Over the course of the 13 years since vCJD first appeared there have only been a handful of deaths amongst military personnel; of these two were seen in DCMH Aldershot in 2003!

## Clinical Features

During the course of vCJD evidence of dementia presents in all cases. In 66% of all cases the presentation is of psychological dysfunction and eventually affects 98% of all cases. Patients' relatives bear the brunt of their instability and intolerance. Depression is seen commonly and associated with impaired sleep and appetite, apathy and loss of concentration, symptoms of anxiety and episodes of panic, obsessive features and impaired libido. Sensory changes tend to occur at an early stage. There are complaints

of numbness, hypersensitivity to physical stimuli, burning sensations or cold sensations and pain of an aching quality. Dysaesthesia of one form or another, including complaints of impaired vision, affects 70% of patients and indicates thalamic degeneration. Cerebellar signs of inco-ordination, ataxia, myoclonus, tremor, and dystonia tend to present after 4 to 6 months and affect 98% of sufferers. This cluster of symptoms is unique to vCJD. In due course the signs of dementia become obvious and the final stages prior to death include dysphagia, incontinence and akinetic mutism; evidence of psychosis is rare.

## Investigations

Initially patients may be referred for psychiatric opinion but emotional disorder and neurological symptoms tend to follow each other with little delay. It is probable that the neurologist will be the first specialist to be consulted but whenever psychiatric referral takes place, and the symptoms suggest the possibility of psychosis or psycho-neurosis, it will be essential to consider whether all other evidence to be gleaned from the presenting history, developmental history, pre-morbid history, family history and current life circumstances are in accord with psychopathology. It is most unlikely that a person who has had stable developmental circumstances and has revealed a positive attitude towards accepting and overcoming challenges is going to retreat into hysterical conversion disorders, or psycho-neuroses or other forms of uncharacteristic behaviour. An individual who is unwell, and who has been exhaustively investigated for organic disorder with negative findings, does not necessarily have a psychiatric disorder.

The neurologist may be faced by an unusual cluster of sensory, motor and behavioural symptoms which are not explained by examination and laboratory findings. Steps will have to be taken to exclude all those conditions which can damage CNS function such as poisons, infections, space occupying lesions, metabolic disorders and trauma. Degenerative disorders such as Alzheimer's disease, Parkinson's disease, Pick's disease, dementia with Lewy bodies, Huntington's disease and auto-immune disorders amongst others will need to be excluded. The commonest differential diagnoses are Alzheimer's Disease and dementia with Lewy bodies. Evidence may not be forthcoming from laboratory tests, radiography, electro-encephalography (EEG), and somato-sensory evoked potentials. Cerebral scanning with magnetic resonance imaging (MRI), positron emission tomography (PET) and single photon emission computed tomography (SPECT) may all prove to be normal. It is possible for all tests to be negative until the illness has reached a terminal stage. In vCJD the EEG may well reveal a background of slow activity, absent alpha waves and period-

ic sharp waves, but these are only found in 70% of cases. CSF examination should reveal no cellular content. The CSF protein may disclose an abnormal factor, the 14-3-3 neuronal protein, which occurs associated with neuronal damage but is found in only 50% of cases. Recent refinements in MRI scanning technique may disclose symmetrical hyper-intense signals in the posterior thalamic region, called the pulvinar sign or FLAIR (10), which occurs in 92% of proven cases of vCJD. It is easily missed but may also be seen in the cortex, globus pallidus and peri-aqueductal grey matter. The pulvinar sign is very specific for that disease.

Absolute confirmation of vCJD occurs if the abnormal PrP<sup>Res</sup> can be found in tonsillar or CNS biopsy specimens from living subjects, but negative findings are common and clinical complications may occur. Biopsy is not to be recommended for patients suffering from an untreatable disease. The diagnosis depends upon the typical symptomatology, and the presence of the pulvinar sign on MRI scanning to clinch the matter; which may be supported by an abnormal EEG, and by the 14-3-3 factor in the CSF.

Laboratory investigations of CSF samples and biopsy materials from living subjects, and from autopsy specimens are constantly being refined. The essential problem is the search for PrP<sup>Res</sup>. Samples are treated with proteases to eliminate the normal PrP. However, specificity remains uncertain in living tissues. The National CJD Surveillance Unit (NCJDSU), which is located at the Western General Hospital, Edinburgh EH4 2XU, has facilities to perform all the relevant investigations and post-mortem studies; and provides advice concerning examinations and management of all cases. The unit can be contacted by telephone on 0131 5372128.

## Case Histories

**Case 1.** The first case was that of an unmarried 25 year old male Army chef who had got his professional qualifications prior to enlistment in June 1999. He was referred by the consultant neurologist at the Defence Services Medical Rehabilitation Centre (DSMRC) and seen on 23rd January 2003. His disciplinary record was good and he had completed military training without any problems. In September 1999 he was posted to a unit in British Forces Germany where he settled in well. In February 2001 he and other members of his unit were detached for a six month tour of duty in Cyprus. In June of that year, and having some time to spare, he volunteered to be a member of a squad of 32 soldiers providing assistance to the local civilian police who were protecting a police station from a group of about 200 rioters. During that action he became separated from his team by four rioters who knocked him to the ground and punched and kicked him. He curled up on his left side thus receiv-

ing the brunt of the assault on the right side of his head and body. His memory of the event is vague, subjective to some extent but possibly influenced by what he was told. He was rescued before long and he recalls recovering full consciousness a little later in the military hospital. Examination confirmed bruises scattered over the right side of his head and body but there were no fractures and he was not detained in hospital for observation. He returned to his duties without delay. A month later he completed his tour in Cyprus and rejoined his parent unit in Germany. He appeared to be well and proceeded on a normal period of four weeks, post-operational leave which he decided to spend at home in England with his family. The holiday was not a success. When his parents were interviewed by the psychiatrist (the author of this paper) they said that he had become uncharacteristically irritable and surly. He was apathetic and did little else except watch television, which was out of character. He did not complain to them of any physical symptoms and they observed nothing other than his irascible attitude. At last, patience exhausted, they urged him to leave and return to his unit. In his account during psychiatric assessment he said that he had begun to experience aching pain along the right side of his body together with feelings of numbness, some burning discomfort, and some apparent muscular weakness also on the right side. He had not suffered from these symptoms before going home. On his return to his unit in Germany he also experienced back pain and he reported sick and was medically examined and reassured that there was nothing abnormal to be found. He continued to perform his duties as a chef and his condition did not immediately deteriorate. By the middle of 2002 he seemed to be well enough to proceed on a military exercise to Oman, but he had begun to experience some twitching of his right arm and leg. Once more he reported sick. He was evacuated back to his unit for further examination and from there was transferred to the Royal Centre for Defence Medicine (RCDM) at Birmingham, where he arrived in August 2002 and remained under neurological investigation for four months. He continued to complain of pain and dysaesthesia. His behaviour was co-operative but he refused to allow his parents to be informed about his circumstances. His brain and spinal cord were scanned thoroughly. A wide range of blood tests were normal as was the examination of his CSF. He developed a relationship with one of the nurses and he expressed a desire to be returned to duty but was obviously not well. At one stage he complained of urinary retention and claimed to have catheterised himself. It was decided to transfer him to the DSMRC under the care of the consultant neurologist where he was seen in November 2002 and was admitted for in-

patient rehabilitation. He also had a full examination by a urologist who found no abnormality on video-urodynamic examination. The staff at the DSMRC found his behaviour to be variable and unpredictable. He exhibited a staggering gait which appeared to be more noticeable when he was observed or with other patients. His symptoms seemed to change day by day. He was neither aggressive nor uncooperative but the staff wondered whether he was faking, however he expressed a desire to return to work! When seen in out-patients he appeared relaxed and co-operative but his manner was so relaxed that the term "belle indifference" came to mind. He was able to give a history of current and past events, with no evidence of disordered speech or verbal content. He described no hallucinatory experiences and expressed no bizarre ideas, nor feelings of passivity or omniscience. However, his staggering gait was grossly evident and his ideas of a return to duty were obviously unrealistic. An anomaly was noted in that he said he had been reared in Willesden, London, whereas his parents stated it was Stanmore, London. Both he and his parents described problems during his childhood connected with the breakdown of his mother's marriage during his infancy, and her subsequent re-marriage whilst still in her teens, and the birth of two half-sisters only two and three years younger than him. He denied any memory of problems in his sibling relationships but his parents confided that there had been a lot of sibling rivalry. The psychiatrist was influenced by the negative results of physical examinations together with the evidence of some problems in childhood, and the history of the assault in Cyprus. He made an initial diagnosis of hysterical conversion disorder and planned to offer desensitisation by abreaction under hypnosis, together with further emotional exploration.

At the next consultation he co-operated well with hypnotic induction and reached a light trance. He described the assault readily but he disclosed absolutely no emotional release. The possibility of a conversion disorder was abandoned, together with ideas of post-traumatic stress disorder. The next step was to interview his parents, who were fully co-operative. They were perplexed by his behaviour, but were concerned and made a good impression. They both worked well with each other during the examination. They confirmed the early history of the breakdown of Mother's first marriage but the second was going well. They described his early displays of sibling jealousy and persisting ambivalence in his response to them, but he was not a naughty boy even though he made unreliable statements and related better to non-family members. More particularly, his school attendance had been good and he conformed. From the age of ten years he had a passion for the Sea Cadets and

retained his membership till the end of his schooling. In GCSEs, he obtained 5 'C' grades and then completed Sixth Form, achieving a Diploma in Health and Social Care. Thereafter he got a job as a pub barman for a year, coped well with rowdy elements and then moved on to train as a chef, obtained his qualifications and enlisted into the Army. This is not the history of an inadequate or nervous personality. He revealed persistence, aim, social ability, conformance and achievements. A psychological explanation for his symptoms had become unlikely. He did not impress as being so clever or so devious as to fake an illness; furthermore, he disclosed no possible motives for wanting to do so. The question of his intellectual function needed to be considered and he attended again for some basic psychometric screening. Again he responded willingly and a start was made on the vocabulary section of the Shipley Hartford Assessment Scale (SHAS). It became obvious within a few minutes that he was completely at sea. He was nervous and hesitant from the start, made only five correct responses in the first six word definitions and then went to pieces and seemed to rely on guess-work. After the thirteenth question the test was abandoned. Arrangements were made for his transfer to the Duchess of Kent's Psychiatric Hospital, Catterick Garrison for further observation and tests. Some time thereafter he was transferred to a civilian hospital where he soon died. The diagnosis of vCJD was confirmed at autopsy. The interval between the onset of symptoms and death amounted to about twelve months.

**Case 2.** On the 27th June 2003 a 32 year old male physical training instructor (PTI), who had been married for eleven years and had two children aged 12 and 10 years, was transferred from psychiatric in-patient care in a civilian hospital to Aldershot Garrison for on-going out-patient management in accordance with standard Ministry of Defence procedure. A diagnosis had been made of affective disorder, with symptoms of depression, anxiety and conversion disorder. He was assessed by a consultant psychiatrist at the DCMH together with two community psychiatric nurses who attempted to set up a therapeutic programme. He was living in a married quarter with his wife and children. Within four weeks, by the 24th July 2003, it had become clear that there was no progress and that matters at home were becoming fraught. His wife had endured some nine months of stress. His mood and temper had been very unstable from the start of his illness. Accordingly, he too was transferred to the Duchess of Kent's Psychiatric Hospital for further assessment. He too deteriorated rapidly and died in a civilian hospital some twelve months after the commencement of his symptoms. There was no autopsy but a

diagnosis of vCJD was accepted by The National CJD Surveillance Unit (3).

His symptoms had presented themselves with a sensation of pain in his right big toe which he noticed a short time after he had had an operation to stabilize his right ankle. The operation was performed in September 2002 and recovery had been uneventful, (he had had a similar operation on his left ankle some ten years earlier which had been entirely successful). He became uncharacteristically irritable and harsh towards his children. It seems possible that the change of mood had preceded the pain in his toe, which was about a month after the operation; but there was little chronological difference between the two. His irritability was very troublesome and by the end of December 2002 there had been a period of marital separation followed by an attempt to use Relate counselling. At first no importance was attached to his toe pain which would have been regarded as due, in some way, to the operation, but pain and associated dysaesthesia began to spread up both legs and advanced over his body, hands, face, lips and tongue. His speech became slurred. He experienced difficulties with motor co-ordination and balance, sleep became disturbed, he felt depressed and was tearful at times, his concentration became affected and he had difficulty with reading and memory. The advance of his symptoms led to referral to a consultant neurologist and admission on the 17th March 2003 for in-patient neurological assessment. By that time he had serious lower limb and mild upper limb ataxia but muscular power remained normal, as did his tendon reflexes and intellectual function. Some mild dysfunction of his touch and temperature perceptions were noted and somato-sensory evoked potentials (SSEPs) were a little delayed in both upper limbs and his left leg, but it was difficult to explain these in the context of the motor dysfunction. Chest X-ray, full blood count, ESR, clotting screening, Vitamin B12, blood sugar, serum electrolytes and calcium, and tests of renal, hepatic and thyroid function were all normal. Likewise, anti-body screening for Wasserman Reaction (WR), anti-nuclear factor (ANF), and anti-neutrophil cytoplasmic antibodies (ANCA) and anti-cardiolipins (ACL) were all normal. The consultant neurologist also consulted a colleague but both were stumped. In the MRI scans of this patient and the Army chef there was no mention of the "pulvinar sign". It is possible that that sign was not known to scanners at the time, or was missed. Unable to establish an organic aetiology they referred the patient for psychiatric assessment and he was admitted to a civilian psychiatric hospital on 8th April 2003.

He was seen by two psychiatrists. They were impressed by the negative investigations, the evident symptoms of depression and a history that both the patient's father

and paternal grandfather had had one episode of depression in middle age. If this was a psychiatric disorder it was difficult to account for the gross motor malfunctions but both could be viewed as hysterical conversion disorder within the setting of a disintegrating marriage. There was, in fact, no past history of maladaptive behaviour or emotional instability but there seemed to be no alternative explanation. His mother and stepfather had both been interviewed by the neurologist and a psychiatrist. He had had a stable childhood with normal development, good health, conforming behaviour, and had been physically active and competitive. He had enlisted into the Army as a boy soldier aged 16 years and had trained as a PTI from the start. He had performed well and gained promotion to sergeant. He had never had any traumatic experience and had been regarded as a well-motivated soldier. There is no recorded evidence of mental dysfunction prior to the onset of this illness. A diagnosis of conversion hysteria was at odds with the facts. The depressive and anxiety aspects of his behaviour might readily be understood as a reaction to his illness and its effects upon his career and family well-being, within the setting of an individual with some hereditary tendency to experience affective symptoms, but his projected irritability on to his wife and children is not in accord with the idea that he was worried about their welfare. Following his admission to the civilian psychiatric hospital he was prescribed a tranquillizer, some night sedation with Zopiclone and medication with the SSRI anti-depressant Sertraline. He remained as an in-patient with breaks as an out-patient for two and a half months and he participated in group therapy. At first there was an improvement in his mental state and neurological symptoms which led to his referral to the DCMH at Aldershot for out-patient rehabilitation, but his condition deteriorated and continued to deteriorate to his death when the correct diagnosis was established.

## Discussion

The incidence of vCJD in the UK reached a peak in the year 1999/2000, giving a rate of 1.08 per million a year, and has since fallen to about 1 per two million per year. The chance that any medical specialist will be faced with a case of vCJD during the course of his or her career is small. Although the symptomology and clinical course of vCJD is singular and characteristic, its rarity tends to pre-empt the making of a correct pre-death diagnosis unless one's mind has been alerted. The two cases described in this paper were, between them, assessed by at least four consultant neurologists, together with other medical specialists, both as in-patients and out-patients. Both were referred on for psychiatric assessment and were seen by one psychiatrist in the first case and three in the

second. At no time was the possible diagnosis of vCJD even mooted. The neurologists were perplexed by the negative results of a range of examinations and tests. The psychiatrists did no better and, acting on the basis of the negative test findings, assumed that there had to be a psychiatric explanation which they were obliged to find. None of the psychiatrists got as far as rejecting a psychiatric aetiology. In both cases the psychiatric diagnosis was a rather vague one of a mixed affective disorder with features of hysterical conversion disorder. Both patients were in quite a frightening situation suffering unpleasant symptoms which were persistent and which threatened their job security and careers. Their anxiety and depressed mood could be explained by worry. Of course there was more to it than that. They both presented with a most unusual cluster of symptoms, which, if not due to some organic cause, suggested a high degree of constitutional emotional instability. In the case of the PTI there were no pre-morbid features to justify such a notion. Even in the case of the chef, a critical examination of his formative years did not reveal a lack of positive features: he was not passive, dependent or timorous, even though there had been problems in his home environment likely to make him feel ambivalent about trust in close inter-personal relationships. There was the assault he experienced in Cyprus. That was certainly a frightening and credible cause for the development of post-traumatic stress reaction (PTSR), which could lead to the mood change observed by his parents some five weeks after the event. The diagnosis of post traumatic stress disorder, (PTSD) requires a history of being exposed to a traumatic event which involved an actual threat of death or serious injury and, in addition, the symptoms of distressing and intrusive recollections of the event, recurring distressing dreams of the event, flash-backs, intense emotional distress when exposed to internal or external cues reminiscent of the trauma, and behaviour designed to avoid any sort of re-exposure, either in thought or deed, to the circumstances which were associated with the trauma. In the chef's case he had experienced an event which could have been significant but he described none of the subsequent reactions. He did not show any reluctance to return to his unit in Germany. He continued to perform his duties so efficiently that, several months later he was considered fit enough to proceed on an overseas training exercise. There is no history of the appearance of maladaptive behaviour such as increasing alcohol consumption or tobacco intake, no attendance unreliability at his place of work, no change in his attitude towards his comrades, no infringement of military rules. He did not describe intrusive recollections or nightmares or feelings of panic. He did describe the symptoms of pain

and dysaesthesia and motor weakness, which are not indicative of PTSD, but could conceivably be explained by conversion hysteria. Conversion hysteria is usually mono-symptomatic not poly-symptomatic and indicates a significant emotional conflict within the patient which he or she feels unable to resolve by some form of voluntary action, or in which the symptoms offer some sort of advantage such as concessions, or care, or transfer of obligations to others. The mechanism of conversion hysteria is subconscious. Presenting symptoms of paralysis, or deafness, or blindness are not consciously calculated to achieve some advantage to the patient. Nevertheless, the border between conversion disorder and factitious disorder (faking) is thin. Either way, a careful examination of the patient should exclude the existence of organic dysfunction and reveal the source of the conflict, which can be confirmed when an effective clinical approach releases pent-up emotions and unveils a conflict which can then be appropriately addressed. In this case a careful assessment eliminated conversion disorder and factitious disorder and PTSD, which left the conundrum; what was wrong with him? The answer, very clearly, was that his disorder was not psychogenic and had to be organic, but in the mind of the psychiatrist this had already been ruled out by the neurological examination results. The psychiatrist resolved his dilemma by transferring it to his colleagues in an in-patient setting.

The case of the PTI was dogged by fewer false trails. He was an experienced and competent soldier and had earned his promotion to substantive sergeant. He had disclosed no past history of neurotic reactions. He had a good job with a guaranteed pension to ensure his ability to support his family and maintain his self-esteem. The formulation of a neurotic affective disorder was based on the evidence of recent marital disharmony, which could conceivably lead to loss of morale in a person with a genetic pre-disposition to develop an affective disorder, based upon a history of affective disorder experienced by his father and paternal grandfather. In fact the marital breakdown was temporary, his wife continued to support him and they occupied an Army married quarter right up to the time of his death. The evidence indicates that his mood change coincided with the other symptoms of his illness. The catastrophic clinical picture simply did not conform with his personality and nature which had never been dysphoric or irresponsible. The psychiatrists, as in the first case, found themselves driven into a corner because of their regard for the thoroughness and competence of the neurological investigations. They were influenced by his obvious depression, but one needs to distinguish between the effects of unhappiness or worry and a genuine affective illness. He must have

been very worried, not about his marriage but about the nature of his illness which threatened his career, his self-esteem and his ability to support his family. The clinical notes do not describe any negative self-concepts such as worthlessness, uselessness, or being a handicap to the welfare of his family, or having negative career prospects and negative self-esteem, which are characteristic of depressive illness. Indeed he showed a willingness to co-operate with all the medical recommendations and was clearly keen to do everything in his power to promote his recovery, including the taking of prescribed medication and participation in group psychotherapy.

### Conclusions

vCJD is a rare condition and the diagnosis is elusive. Only the presence of the pulvinar sign on MRI scanning is diagnostically reliable but not more than 92%. The most reliable evidence is the clinical presentation which may then be supported by the pulvinar sign, the presence of the 14-3-3 factor in the CSF, and the EEG findings of a loss of the normal rhythms, and the presence of scattered bi-phasic or tri-phasic sharp wave complexes with a frequency of 1 to 2 per second. With respect to the psychiatric assessment it is well known that a diagnosis of conversion disorder should be made with caution and with evidence of a clear cut stress factor, for which the patient, under mental conflict, can see no acceptable solution. A diagnosis of factitious disorder or conscious symptom faking must be related to a clear-cut advantage to the patient. Neurologists and psychiatrists are bound to consult each other about complex diagnostic issues and psychiatrists must be wary of making formulations which only approximate to the clinical evidence. Any neurologist who encounters a patient who is diagnostically elusive would be criticised if no psychiatric opinion was requested, even if a diagnosis of CJD was suspected. The psychiatrist's opinion must be entirely objective and also sceptical, thorough, realistic and as near foolproof as possible, in line with the Diagnostic and Statistical Manual of Mental Disorder - IV (DSM-IV), and the International Classification of Diseases 10th edition (ICD-10).

The neurologists were backed into a corner by the persistent negative results to virtually all their very thorough investigations. If they had not then referred the patients for psychiatric assessment they stood to be criticised. The psychiatrists found themselves confronted by a most unusual clinical picture but were persuaded that the diagnosis had to be psychiatric and non-organic; there were some circumstances which supported such a view. Close scrutiny, after the event, reveals that the full facts did not support a primary psychiatric illness. The fact is that none of us even cast doubt upon a psychiatric aetiology.

The neurologists relied correctly upon objective evidence, which was lacking. The psychiatrists should have been equally objective, but the basis for their opinions had to rely less upon clinical investigations and more upon the accumulated influence of many factors. When clinical features are bizarre and contradictory, and careful assessment reveals no ulterior motives for faking illness, and no prior history of significant maladaptive behaviour, it becomes necessary for a psychiatrist to reject a psychiatric diagnosis. We are unlikely to see another case of vCJD but diagnostic difficulties arise in other conditions including sCJD.

Finally, there is the problem of what to tell the patient and relatives once the diagnosis has been made? A diagnosis of CJD is unlikely to be made in less than six months from the appearance of symptoms. By then the disease is well advanced and there may have been more cognitive damage than is evident from talking to the patient. It is possible that the patient may never try to pin down a doctor for a diagnosis and will accept a caring attitude and palliative help. Relatives need to be told the hard facts. They do not have long to make all the necessary arrangements when death is imminent. They need to understand that the illness is not due to any fault on the part of the patient or themselves. They need to appreciate the rarity of the condition and fatal prognosis. Helpful advice can be had by contacting the National CJD Surveillance Unit, The Western General Hospital, Edinburgh EH4 2XU, which must be informed anyway to notify the National CJD Surveillance Programme.

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