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CASE REPORT

Presenile dementia concomitant with Sjogren's syndrome: A Case Report

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ABSTRACT

Aside from the typical degenerative cause, what other etiologies should be considered for a dementia patient who was diagnosed with Alzheimer's disease at the age of 58? A female patient aged 65 has experienced a gradual deterioration in her speech over the course of the last seven years. This decline in speech has been accompanied by the presence of extensive tooth decay, referred to as "rampant caries," within her mouth, as well as the development of "vitiligo" on her skin. Aditionally, she exhibited an allergy to penicillin, along with an abnormal subset of lymphocytes (TBNK) and numerous foci of demyelination observed on her brain CT (limbic system). A final presenile dementia resulting from Sjogren's syndrome was determined, indicating a potential role of autoimmunity in the pathogenesis of presenile dementia when the patient was younger.

Keywords: Presenile dementia; Rheumatism; Autoimmunity; Pathogenesis

1. Introduction

The present case pertains to a 65-year-old female patient exhibiting prensenile dementia, who has reported a progressive decline in speech function over a period of seven years. Additionally, the patient presents with notable instances of "rampant caries" in her oral cavity and "vitiligo" on her skin. In addition, the patient exhibited an allergic reaction to penicillin, along with an atypical subpopulation of lymphocytes (TBNK), and multiple foci of demyelination demonstrated on her brain imaging (pointing to the limbic system). A final diagnosis of presenile dementia, complicated by Sjogren's syndrome, was determined in this particular case.

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2. Case Report

The patient, a 65-year-old woman who had retired from her job as a worker, was brought to the hospital due to a gradual decline in cognitive function that began at the age of 58. Initially, she possessed the ability to articulate her thoughts, albeit accompanied by a somewhat impaired memory. Subsequently, her condition progressed to include stiffness in her limbs, weight loss, and challenges in mobility. Upon admission to the hospital, she displayed evident signs of emaciation. During her stay, she encountered limb muscle atrophy, resulting in a near disappearance of her muscle mass on the Hegu-spot. Additionally, her hip muscles on both sides experienced atrophy. It is worth noting that she consumed a substantial amount of water on a daily basis.

As her condition continued to deteriorate, her daughter decided to bring her to our hospital for specialized care. Upon arrival, she was entrusted to the capable hands of Mr. Liang and Dr. Li, who would provide her with the necessary long-term treatment. The patient gradually reduced her verbal communication until eventually becoming completely silent and confined to bed throughout the day. Oral examination **(Figure 1)** indicated: the rampant caries, and the vitiligo seen on her hip skin. Her lymphocytic subset (TBNK) demonstrated: CD3 + CD4 + 1445 / μL (normal range: < 1247/ μL); CD3 + CD4 +(%) 55.2% (normal range: < 50%). Urine β-2 microglobulin showed 412 μg / L (normal range: < 300 μg / L). Urine β-2 microglobulin indicated 412 μg / L (normal range: < 300 μg / L). A CT scan of her brain **(Figure 2)** showed the multiple foci of demyelination; and the sarcoidosis also seen on her lung CT. With the information, a final diagnosis of presenile dementia, resulted from Sjogren's syndrome was determine for the patient.

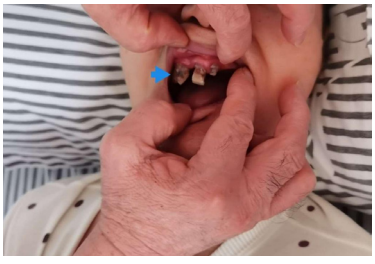

Figure 1- Patient's Oral Examination

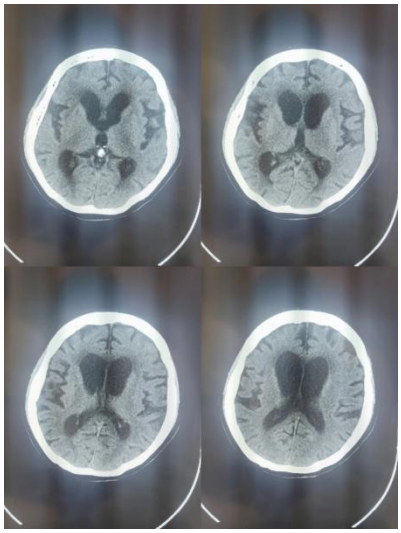

Figure 2 - CT scan of patient's brain

3. Discussion

Is it a "mere coincidence" or a result of "cause and effect" that this patient suffering from dementia also has a simultaneous autoimmune disease such as Sjogren's syndrome or rampant caries? This led her doctor to contemplate the matter extensively. Based on an extensive body of literature, it is widely acknowledged that a small subset of patients experiencing cognitive impairment exhibit the presence of sensitized T-lymphocytes and/or anti-nuclear antibodies (ANAs), which have been implicated in the "attack" on the central nervous system (CNS). The presence of "dopamine-producing cells" within the extrapyramidal system (EPS) and the hippocampus situated in the limbic system of the brain **(Figure 2)** contributes to the manifestation of symptoms associated with Parkinson's disease (PD) and Alzheimer's disease (AD) [1,2].

We made the assumption that the identification of "autoimmune marks" in the immunological profile of a patient with presenile dementia beforehand could be likened to discovering a "new therapeutic target" in the management of dementia. Hence, the incorporation of a small amount of "immunosuppressive ingredients" from Chinese herbal medicines nto the existing treatment regimen has the potential to enhance overall effectiveness.

The patient's allergic constitution to penicillin

had been previously established, along with the observation of "rampant caries" in her oral cavity **(Figure 1),** which pointed to the possibility of "Sjogren's syndrome" (SS). In addition, the patient suffering from "Vitiligo"on her skin, suggesting an autoimmune condition targeting melanocytes in the basal layer of the skin; Coupled with the abnormal TBNK levels, there is justification to consider the presence of "immune abnormalities" may be engaged in the pathogenesis and development of AD / PD in this setting. Which is also known as autoimmune dementia (AiD)[3]. It has been noted that the presence of "immune abnormalities" in patients with dementia is associated with a more rapid and severe decline in cognitive function.

From the moment symptoms appear, the patient should be unequivocally classified as "presenile". Over a span of seven years, the patient exhibited a lack of clear self-expression, necessitating reliance on her physical signs, laboratory findings, and imaging features to gather her medical history. Through investigating the etiology / pathogenesis of her dementia, we aim to utilize this as a chance to envisage an in-depth screening for the immunology in patients with presenile dementia (e.g., preventive detection and intervention). If abnormal immunology is verified, it is imperative to promptly initiate "secondary prevention" following the "primary diagnosis" of presenile dementia. In this context, "preventive interventions" would be carry out to slow down the advancement of the condition and enhance the patients' overall quality of life. The rheumatism department at our hospital plans to utilize this particular case study as a "research concept" to propose a research project, and we will endeavor to have a significant impact on it.

4. Conclusions

We have come to the conclusion that the occurrence of a relatively swift decline in cognitive abilities, such as the ability to speak, in a relatively young woman, particularly when accompanied by certain rheumatic conditions like Sjogren's syndrome, can be attributed to an autoimmune mechanism in the development of presenile dementia. This finding suggests an alternative explanation to the conventional degenerative theories in its pathogenesis.

Conflict of Interest

There is no conflict of interest.

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