

Brain hypoperfusion on Tc-99m-ethylene dicycysteine diethyl ester single-photon emission computed tomography in Hashimoto's encephalopathy

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ABSTRACT

We present a 17-year-old female, previously diagnosed of autoimmune hyperthyroidism who had an acute neurological episode and presented high antithyroid antibodies titers, cerebral spinal fluid and electroencephalogram changes. Tc-99m ethylene dicycysteine diethyl ester brain single-photon emission computed tomography (SPECT) showed global and patchy hypoperfusion. With glucocorticoid therapy, clinical symptoms disappeared; there was a decrease in antithyroid antibody levels and repeat brain SPECT revealed improvement of perfusion.

Keywords: Antithyroid antibodies, brain single-photon emission computed tomography, Hashimoto's encephalopathy, hypoperfusion

INTRODUCTION

Hashimoto's encephalopathy (HE) is a rare neurological syndrome in patients with serologic evidence of autoimmune thyroid disease. Antithyroid antibodies are always increased. As clinical presentation is variable, diagnosis is made by excluding other causes of encephalopathy.

We describe the case of a young female previously diagnosed of autoimmune hyperthyroidism who had an acute neurological episode and presented high antithyroid antibodies titers at that moment. We report the findings on Tc-99m ethylene dicycysteine diethyl ester (ECD) brain single-photon emission computed tomography (SPECT) during the exacerbation and the recovery phases of encephalopathy.

CASE REPORT

We present a 17-year-old female who had an acute episode

of left-sided weakness and hypoesthesia and dysphasia for around 15 min. It was associated with progressive right frontal headache and vomiting. As past medical history, she had recently been diagnosed of autoimmune hyperthyroidism, but she had undergone no treatment. Laboratory evaluation showed high free T₄ (FT₄) (26 pg/ml, normal 8-18) and remarkably elevated serum levels of antithyroperoxidase (3,228 UI/ml, normal < 60) and antithyroglobuline antibodies (>22,200 UI/ml, normal < 280). Since clinical features were unspecific, it was necessary to exclude causes of encephalopathy (infectious, metabolic, toxic, vascular, neoplastic, paraneoplastic, etc.). Both brain computed tomography (CT) scan and magnetic resonance imaging (MRI) showed normal findings. Patient was noted to have elevated cerebral spinal fluid (CSF) protein as well as diffuse slowing on electroencephalogram (EEG). A brain SPECT 45 min after injection of 68.8 MBq (18.6 mCi) of Tc-99m ECD was performed. Projection data were obtained on a gamma camera millennium VG (GE, Medical Systems, USA) in a 128 × 128 matrix using zoom of 1.5, 64 views, 20 s/view. SPECT images showed diffuse and patchy defects of the cortical uptake [Figure 1].

With all these findings, HE was diagnosed and steroid therapy was initiated. Clinical symptoms disappeared and there was a decrease in antithyroid antibody levels. Treatment for hyperthyroidism was started with antithyroid drug and titers of FT₄ normalized. A repeat brain SPECT was performed 3 months later with the injection of 66.6 MBq (18.0 mCi) of

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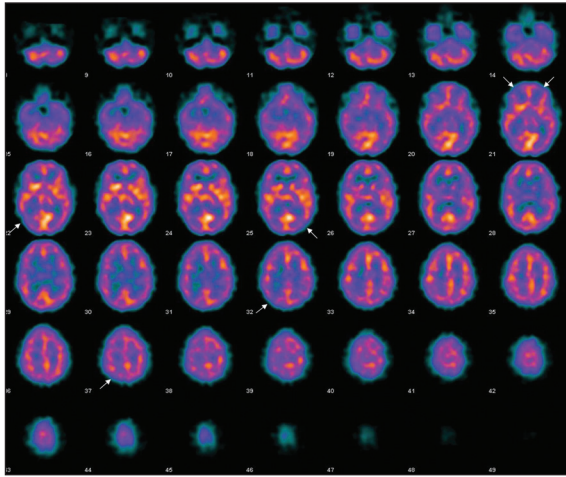


Figure 1: We show transaxial brain single-photon emission computed tomography images with irregular and patchy decreased cortical tracer uptake of both hemispheres (arrows)

Tc-99m ECD using a dual-head gamma camera (Elsint Helix, GE Medical Systems, USA). The data acquisition parameters were 64×64 matrix using $\times 2$ zoom, 64 views, 20 s/view. SPECT images showed small uptake defects, but demonstrating significant improvement of perfusion respect from the previous study [Figure 2].

After 3 years of follow-up, patient remains free from relapse or recurrence of the neurological symptoms.

DISCUSSION

HE is a rare neurological syndrome in patients with serologic evidence of autoimmune thyroid disease. Antithyroid antibodies are always increased, although titers do not correlate to the degree of encephalopathy.^[1,2] Clinical presentation is variable, from stroke-like episodes to progressive cognitive impairment.^[2,3] Most of the cases diagnosed of HE are affected by Hashimoto's thyroiditis and less frequently, by other autoimmune thyroid diseases, chiefly Graves' disease,^[4] that is the reason why the choice of a comprehensive definition like "encephalopathy associated with autoimmune thyroid disease" seems more appropriate than HE. Thyroid function is usually clinical and biochemically normal^[5] although its status varies greatly.^[1-3] Brain CT scan and MRI present unspecific findings in adults and they are usually normal in pediatric patients; although, very few cases have been described in children. Elevated CSF protein as well as diffuse slowing on EEG, are common findings too.^[3,5,6]

The pattern of brain perfusion on SPECT of the majority of patients is diffuse and patchy defects, with areas of reduced cortical uptake not related to a specific region,^[6-8] although focal hypoperfusion has also been reported.^[2,9] There is no correlation among SPECT hypoperfusion, neurological or thyroid clinical status and antibody titers, but frontal hypoperfusion has been described in euthyroid patients without encephalopathy.^[10]

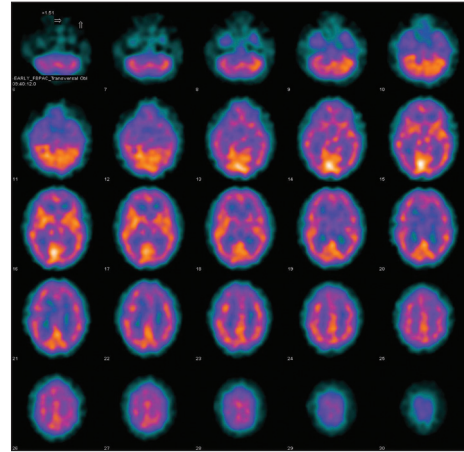


Figure 2: On selected transaxial brain single-photon emission computed tomography images, a comparative improvement of perfusion respect from the previous study is shown; although, small perfusion defects in brain cortex are still present

Corticosteroid is the treatment of choice, because of its effectiveness in the reduction of the neurological symptoms^[3] and gradual improvement of brain perfusion on SPECT.^[6,7] The pathogenesis of this encephalopathy is still unknown and poorly defined by autoimmune mechanisms as disruption of cerebral microvasculature secondary to autoantibody or immune complex deposition that could alter vascular reactivity and be responsible for global and/or focal hypoperfusion on brain SPECT.^[3,6]

In this case, we present brain perfusion defects on SPECT contributed to diagnosis of the neurological syndrome. It was the only diagnostic imaging technique with positive findings and it allowed initiation of the steroid treatment and her clinical improvement.

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