

Case Report

Visual Pathway Disturbances in Rosai-Dorfman Disease: A Case Report

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Abstract

Rosai-Dorfman is a usually benign disease which is characterized by overproduction and accumulation of a specific type of white blood cell in the lymph nodes, most often those of the neck region. Different organs including the central nervous system may be affected in rare cases. The aim of the present manuscript is to report visual pathway disturbances measured in a case of Rosai-Dorfman with central nervous system involvement using electroretinography and visual evoked potential techniques.

Keywords:

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Introduction

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy is an uncommon benign lymphoproliferative disorder¹. The histologic hallmark of this disease is the finding of emperipolesis displayed by lesional histiocytes¹. The incidence of this disease is approximately 100 cases per year in the united state¹. The disease was first described in 1969 as a distinct lymphoproliferative disorder by pathologists Juan Rosai and Ronald Dorfman¹.

While this disease commonly affects lymph nodes, extranodal involvement of multiple organs has been reported including the central nervous system¹. However the involvement of central nervous system is very rare and is not well characterized².

It is known that the visual pathway is a part of central nervous system³. Visual pathway is a segment of visual system which starts from retina and end in the occipital region in brain⁴. Any pathological disturbances in this segment may be a sign of central nervous system deficiencies.

There are different methods to look for pathological abnormalities of retina and visual pathway including electroretinography (ERG) and visual evoked potential (VEP) techniques. Here we report a Rosai-Dorfman patient with central nervous system involvement undergoing ERG and VEP tests to look for the possible changes in her retinal and visual pathway function.

The present case report was approved by the ethics committee of Basir Eye Health Research Center, Tehran, Iran, and the patient gave written consent for this report.

Case Report

A 25 years female with established diagnosis of Rosai-Dorfman was referred to Basir Eye

Clinic, Tehran, Iran, with hand motion eye sight. The optical coherence tomography (OCT) and fluorescence angiography of the patient eyes revealed pathological changes in retinal layers. To further study the possible changes in her retinal pathway VEP and ERG tests were conducted for the patient.

The VEP, P₁₀₀ latency was increased in both eyes (125 and 130 msec in the right and left eye respectively). The normal range for VEP, P₁₀₀ latency is 100 msec. The value for VEP, P₁₀₀ peak amplitude was 2 mv for both eyes. The normal value for amplitude measured by our instrument is above 5 mv.

The ERG b-wave amplitude was 32 and 25 mv in the right and left eyes respectively. The normal value for ERG b-wave amplitude is more than 100 mv for our instrument. The respective latencies for the right and left eyes were 44 and 46 mv, which were in normal range (Normal range 40- 46 mv).

Discussion

A female patient with Rosai-Dorfman disease and poor vision was referred to our clinic. Her test results were abnormal in VEP and ERG examinations, indicating visual pathway and retinal disturbances in the patient.

Similar to our findings Nemir et al.,⁶ have reported a 4 year old patient with extranodal Rosai-Dorfman disease which involved the right optic nerve causing atrophy of the optic nerve papilla⁶. In their report visual evoked potential study of the right eye showed normal findings related to the prechiasmatic visual pathway but severe dysfunction of the right optic nerve⁶.

Conclusion

Retina and visual pathway of Rosai-Dorfman patients with central nervous system

involvement might be affected which can be diagnosed using electroretinography and visual evoked potential techniques.

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Footnotes and Financial Disclosures

Conflict of interest:

The authors have no conflict of interest with the subject matter of the present manuscript.