VEP Pediatric Case Study

Retinoblastoma Detected After Failed Visual Evoked Potential (VEP) Test in a Pediatric Office

Presentation

A six month old female patient was given the Enfant[®] Pediatric VEP Vision Test during a routine wellbaby visit at Willows Pediatric Group in Westport, CT. She had a normal physical exam, including normal red reflex, no leukocoria, no eso- or exotropia, PERRL, EOMI, and normal fix and follow. There was no family history of vision problems, she appeared in good health and was attentive at the time of the exam. The VEP Vision Test indicated a statistically significant difference in visual function between the right and left eyes, and the child was referred by pediatrician Jeffrey A. Owens, MD to a pediatric ophthalmologist.



Assessment

The patient did well with the initial ophthalmic exam, but upon dilation, lesions were observed. The pediatric ophthalmologist's diagnosis was suspected bilateral retinoblastoma. The pediatric ophthalmologist referred the patient to an ophthalmic oncologist at Yale University. During the visit with the ocular oncologist, the patient was able to follow with both eyes; she had normal external aspect of both eyes, and normal anterior segments.

The patient was examined under anesthesia. A funduscopic exam of the right eye revealed a 6x6x3 mm lesion superonasal to the optic nerve, ½ disc diameters away from the optic nerve with 90% calcification. Two other lesions were found in the right eye, one equatorial lesion of 3x3x1.5 mm with 90% of calcification and another equatorial lesion of 5x5x2.5 mm without calcification. Funduscopic exam of the left eye also revealed a yuxtapapillary lesion of 4x4x2.5 obscuring approximately 30% of the optic nerve with 10% of calcification, together with another lesion of 8x8x4 mm superotemporal to the macula not involving the fovea with 5% of calcification.



Diagnosis and Treatment

The patient was diagnosed with bilateral retinoblastoma. The oncologist explained to the parents that retinoblastoma is a rare and serious cancer, but has a high survival rate if treatment begins early. He also discussed genetic counseling and the utility of genetic testing, and the need for family members to be examined.

The decision was made to treat the patient with systemic chemotherapy and local intra-arterial chemotherapy. The intra-arterial chemotherapy consisted of a catheterism of the ophthalmic artery and local delivery of the drug. The tumors were also treated with a transpupillary thermotherapy (TTT) laser using an indirect ophthalmoscope. Photographs and ultrasound were taken to document the progression of the disease. The patient was scheduled to return to the ophthalmic oncologist regularly for 6 months to assess for new tumor growth or regrowth, and the need for continued treatment.

According to her oncologist, the patient continues to do well 2 ½ months post-diagnosis. The retina is completely in place. All the tumors are regressing and the fovea remains anatomically normal. This is a good response and the tumors might be pulled away and this child may well end up with good vision in both eyes. This is becoming a success.

Conclusion

Dr. Owens commented, "If this patient had not had an Enfant[®] VEP vision test, it is unclear how many months, or even years, it would have taken for her disease to present clinically, at which point there may have been significant distant metastases and a much poorer prognosis. At this time, she has a 97% likelihood of survival." The patient's mother said, "I feel in this case the VEP test saved her life. I'm grateful we were at a pediatrician who had the VEP because if not, we wouldn't know right now. We would be years down the line thinking about removing her eye or maybe dealing with brain cancer."

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