Vaso-vagal Syncope (VVS) after Mydriatic Eyedrops in Cutaneous and Intracranial Vascular Deformations (Pascual-Castroviejo Syndrome Type II)

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Abstract

Purpose: To present a case of vaso-vagal syncope (VVS) after mydriatic eye drops in a patient with cutaneous hemangioma and intracerebral vascular abnormalities.

Case report: A 41-year-old woman presented to an ophthalmology clinic complaining of near vision disturbance. On examination, there was dilatation and telangiectasis of the conjunctival vessels on the temporal side of the right eye associated with right hemicranial and neck cutaneous involuted hemangioma ipsilateral to the conjunctival lesion. The patient experienced vaso-vagal syncope for approximately 15 minutes after mydriatic eye drops, 2 months prior to presentation. A magnetic resonance angiogram (MRA) showed right hypoplasia of the lateral and sigmoideus venous sinus ipsilateral to the external right hemangioma with predominantly compensatory drainage at the contralateral left system. At the neck vessels, the MRA showed dominant drainage through the lateral and jugular left sinus by the hypoplasia of the contralateral right system.

Conclusion: This case shows the association of telangiectasis of the conjunctiva, and cutaneous hemangiomas of the head and neck with anomalies involving the central nervous system (CNS) as described by Pascual-Castroviejo in 1978, which they called cutaneous hemangioma–vascular complex syndrome. The diagnosis was made after the patient experienced VVS after mydriatic eyedrops instillation.

Keywords: Vaso-vagal syncope; cutaneous hemangioma-vascular complex syndrome; mydriasis.

Introduction

We report a case of vaso-vagal syncope (VVS) in a 41-year-old woman with cutaneous hemangioma and intracerebral vascular abnormalities compatible with features previously described by Pascual-Castroviejo et al. We suspect, that the vaso-vagal syncope could be related to her clinical intracerebral venous insufficiency.

Case Report

A 41-year-old woman visited a general ophthalmologist for myodesopsia and near vision disturbances. She developed acute loss of consciousness for approximately 15 minutes while she was in the waiting area 20 minutes after the third sympathetic mydriatic eye drops. Both, the neurologist and the cardiologist of the medical center evaluated her and they concluded a
diagnosis of a vaso-vagal syncope. Two months later, the patient came to our ophthalmology clinic complaining of near vision difficulties. Her far vision was 20/20 in both eyes. On slit lamp examination, the only finding was dilatation and telangiectasis of the conjunctival vessels on the temporal side of the right eye (Figure 1-A). The intraocular pressure (IOP) was 14 mmHg, and the rest of the ocular examination including ocular fundus was normal. Left eye was completely normal. General examination showed a right hemicraneal and neck cutaneous involuted hemangioma ipsilateral to the conjunctival lesion in the right eye. The hemangioma was occult by the hair and the dress of the patient (Figure 1-B) and the lesion extended from the temporal right cranial side to the lower part of the neck (Figure 1-C and 1-D). However, her past medical history was negative.

Magnetic resonance angiography (MRA) showed right hypoplasia of the lateral and sigmoideus venous sinus ipsilateral to the external right hemangioma with predominantly compensatory drainage at the contralateral left system (Figures 2-A and 2-B). At the neck vessels, the MRA showed dominant drainage through the lateral and jugular left sinus by the hypoplasia of the contralateral right system (Figure 2-C).

Discussion

Cutaneous hemangiomas of the head and neck and their possible association with anomalies involving the central nervous system (CNS) were ignored until they were described by Pascual-Castroviejo in 1978. In 1996 they presented the clinical and imaging findings of 17 patients and suggested that this disorder, which they had called cutaneous hemangioma–vascular complex syndrome, should be included as a neurocutaneous syndrome. In 2005, they described vascular and nonvascular intracranial and extracranial anomalies associated with hemangiomas and vascular malformations of the face, neck, and/or chest in forty one patients. We describe a female patient with cutaneous hemangioma, conjunctival telangiectasis, and intracerebral vascular anomalies; specifically a right hypoplasia of the lateral and sigmoideus venous sinus ipsilateral to the external right hemangioma and at the neck vessels, lateral and jugular right sinus hypoplasia with dominant drainage through the contralateral system. Castroviejo et al suggest that spontaneous regression occurs in parallel in the cutaneous hemangiomas, and in the vascular pathology usually present internally at the same metameric level. They reported a 28 years old woman with marked decrease in caliber of the vessels of the ipsilateral side of the facial and neck hemangioma after 27 years, so they suggested that cutaneous, internal hemangiomas and vascular abnormalities could increase in size during the first months or years and then spontaneously regress.

Syncope in apparently healthy subjects is usually attributed to a vasovagal reaction and two cases on healthy patients who experienced VVS on applanation tonometry or instillation of dilating drops has been reported. However, the vaso-vagal nature of syncope remain unexplained and alpha-sympathomimetic agents are effective in preventing spontaneous episodes of vaso-vagal syncope during a short-term follow-up.

We report a VVS after instillation of dilating eye drops in a patient with cutaneous hemangioma and intracerebral vascular abnormalities compatible with features previously described by Pascual-Castroviejo et al. Vaso-vagal syncope may be a one-time occurrence due to sudden cessation of blood supply to the brain and is the most common
References

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In our patient, MRA shows the dominant venous drainage through the lateral and jugular left sinus by the hypoplasia of the contralateral right system. Intermittent increase in pressure of these engorged and congested veins and the abnormal drainage of the right system were probably an additional prerequisite for the occurrence of clinical symptoms and maybe the most likely cause of the development of syncope.

In summary, this case exemplifies that ophthalmologists need to be aware of recent advances in other fields in medicine. In addition, we need to demonstrate to other physicians the importance of ophthalmic consultation. To be aware of the ophthalmic manifestations of a series of systemic or neurologic diseases may save sight and even life itself in some cases. We suspect that the vaso-vagal syncope experienced by our patient, reflects her inability to activate these protective reflex mechanisms; a situation that could be exacerbated by her clinical intracerebral venous insufficiency.