Intermittent visual loss and exophthalmos due to the Blue rubber bleb nevus syndrome

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PURPOSE: To describe the occurrence of intermittent visual loss in a patient with orbital hemangioma due to the blue rubber bleb nevus syndrome.

METHODS: Case report. RESULTS: A 70-year-old woman complained of intermittent blurred vision combined with pressure behind her left eye occurring after head bending or Valsalva maneuvers. Examination revealed 2 mm of enophthalmos on the left. After downward bending of the head the eye became exophthalmic by 12 mm and visual acuity decreased to finger counting. Neuro-ophthalmologic examination was otherwise normal. Magnetic resonance imaging revealed an intracanal hemangioma. Bluish, soft, rubber-like, compressible and slowly refilling hemangiomas on the inferior lip, sublingually, on the hand, and vagina led to the diagnosis of BRBNS.

CONCLUSIONS: Intermittent visual loss may be the presenting symptom of an orbital hemangioma in BRBNS. Recognition of this rare condition is important because of its potentially life-threatening complications.

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