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WHAT'S YOUR DIAGNOSIS

What is your Diagnosis? Hydrocephalus in a patient with hemophilia A



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A 17-year-old male with a past history of hemophilia A presented to the emergency department with headaches, dizziness and an unsteady gait following a recent fall. He had intercurrent hemophilia arthropathy, and his medications were tramadol and anti-hemophilic factor VIII. A CT scan revealed hydrocephalus with a patent aqueduct of Sylvius, a normal fourth ventricle and an irregular cystic enlargement in the region of the third ventricle as well as a dilatation of the lateral ventricles (Fig. 1). There were no intracranial hemorrhages noted. Ophthalmology was consulted to rule out papilledema. Ophthalmic examination revealed an alert and oriented young man with bilateral visual acuities of 20/25, Ishihara colour vision of 15/15 OU, normal pupillary reactions without relative afferent defects and normal anterior segments. Mild

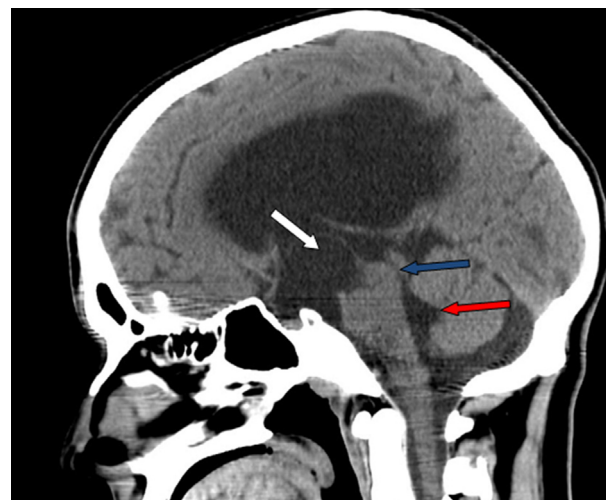


Figure 1 Sagittal CT scan of the brain showing an irregular sellar and suprasellar cystic lesion (white arrow), dilated lateral ventricles and normal fourth ventricle (red arrow) and a patent aqueduct of Sylvius (blue arrow).

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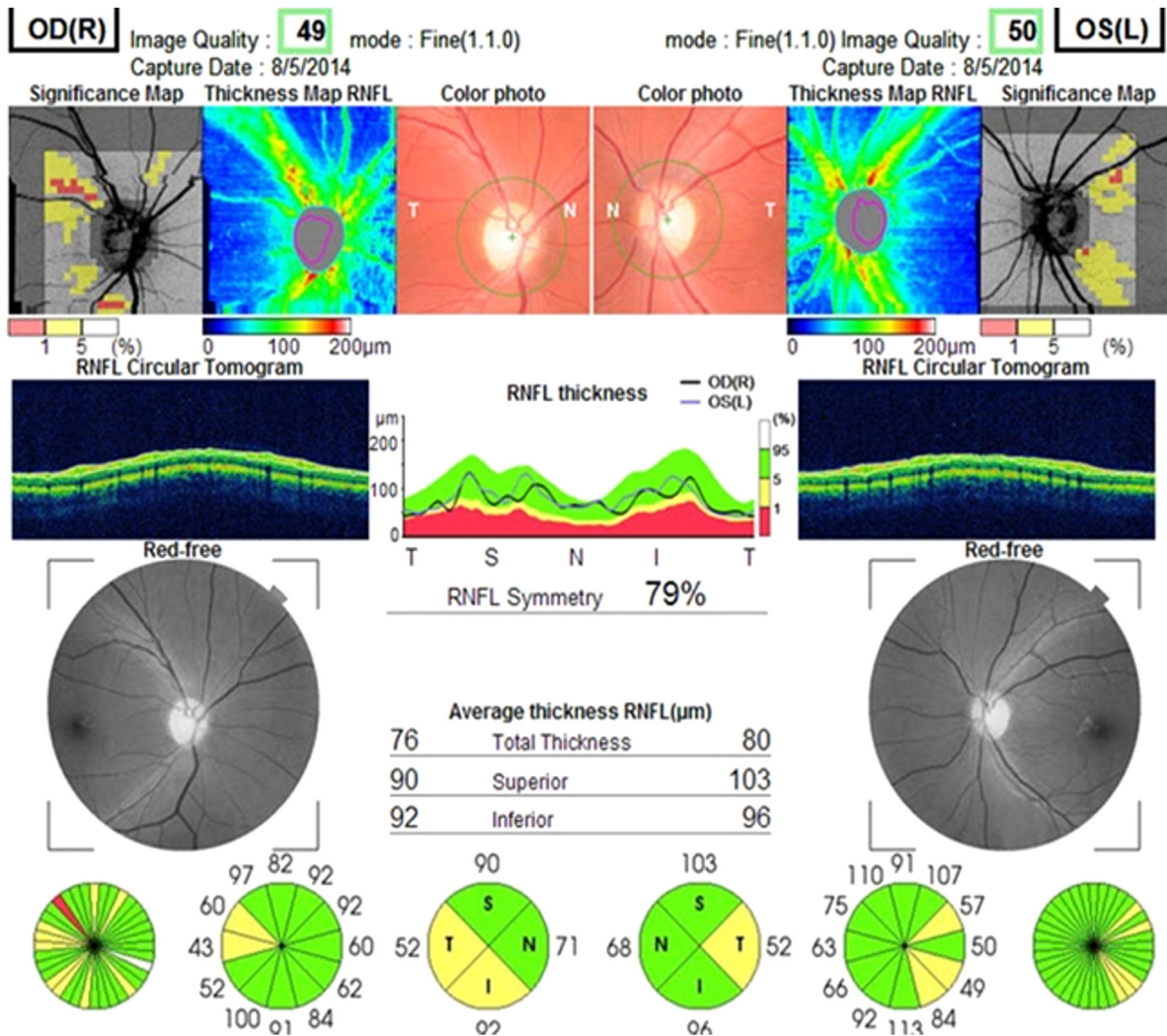


Figure 2 Fundus disc photos and OCT: Mild temporal disc pallor and temporal optic nerve atrophy with thin retinal nerve fibre layers, which were likely the result of chiasmal compression. OCT shows a linear map of retinal nerve fibre thickness with the lines dipping into and below the yellow zones (thin RNFL) in the temporal regions.

bilateral temporal optic nerve pallor was noted on dilated ophthalmoscopy. There were no signs of current or previous papilledema. Optical coherence tomography (OCT) of the optic nerves supported the clinical findings, displaying modest segmental temporal retinal nerve fibre layer (RNFL) thinning (Fig. 2) with an average thicknesses of 76 µm and 80 µm in the right and left nerves,

respectively (normal ~100 µm) [1]. The Humphrey visual fields were consistently unreliable. The neurological exam was completely normal. What is this cystic lesion and what other investigations would you order?

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