A 20-year-old-female presented to the emergency department (ED) with a chief complaint of a persistent dull headache associated with a 7 mm dilated, non-reactive right pupil, and occasional blurry vision for 4 days. The patient had a past medical history significant for Noonan’s syndrome (NS). In the ED the patient’s physical examination demonstrated normal vital signs, chronic right divergent strabismus (Figure 1), a dilated right 7 mm pupil (Figure 2), and intact extraocular muscles without visual field cuts or other acute neurological deficits. Computed tomography angiography of the brain demonstrated the diagnosis (Figure 3).

Oculomotor nerve palsy (ONP) in patients with posterior communicating artery aneurysms (PCoA) is known clinically. Direct compression of the oculomotor nerve by the aneurysm itself is considered to be the mechanism causing ONP in patients without subarachnoid hemorrhage. NS is an autosomal-dominant condition of multiple congenital abnormalities, regarded as a type of dwarfism with a reported incidence of between 1 in 1,000 and 1 in 2,500 live births. These abnormalities include craniofacial anomalies, such as ocular hypertelorism, low-set ears, low posterior hairline and webbed neck; shield-shaped chest; cubitus valgus; cryptorchidism; and congenital heart defects. Interestingly, well documented cases of patients with NS and valvular and non-cerebral vascular lesions may show a preponderance for underlying defects in vascular architecture, i.e. Ehlers-Danlos syndrome. Only 4 cases of intracranial aneurysms have been previously reported in individuals with NS. Finally, careful attention must be paid to all patients, not just those with NS, who present with a new onset dilated pupil; and should raise one’s suspicion of an underlying PCoA (Figure 4).
Artery Aneurysm with Noonan’s Syndrome

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