A CASE OF RECURRING AMAUROSIS FUGAX

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WW a 43 year old Caucasian Hospital Manager, first presented in July 08 complaining of transient mono-ocular visual loss in the right eye. She described sudden onset loss of vision – ‘like a curtain coming across the vision’. She then proceeded to have similar symptoms in the left eye. There was no history of a subsequent headache. The episode would between 30 seconds and 10 minutes and could occur up to 10 times a day. At times, the attack was also associated with some left arm numbness. She was initially reviewed by Dr Vu [Stroke Consultant] who diagnosed Amaurosis Fugax and started her on standard anti-platelet therapy [combination of Aspirin and Dipyridamole]. She had very little in terms of vascular risk factors [nil hypertensive, minimal alcohol and a life-long non-smoker with a fasting cholesterol 3.12]. Her PMH history consisted of Gilberts’ Syndrome and endometriosis. She also suffered a DVT following a hysterectomy. Her initial investigations of a CT head and carotid duplex were normal.

Despite anti-platelet therapy she continued to have attacks of loss of vision and they would flit from left to right eye. All in all she had up-to a hundred subsequent attacks. A MRA of her carotids done on the 17th of July demonstrated bilateral 80% stenosis at the origin of both internal carotid arteries with an intact vertebral circulation. Due to the frequency of her ‘attacks’, she was urgently reviewed by a Vascular surgeon on the 24th of July who observed bilateral retinal infarcts on fundoscopy. He proceeded to perform a carotid duplex which did not demonstrate any plaques bilaterally but did reveal high velocity [120] on the right (suggestive of at-least 70% stenosis) and normal velocity [less than 80] on the left. Due to the severity of her symptoms she was admitted to hospital and proceeded to undergo an endarterectomy of the Right Carotid.

However her endarterectomy (done the next day- 25th July), revealed no stenosis and a normal looking carotid artery. A subsequent CT angiogram of the Aortic Arch and Carotids [28th of July] revealed no abnormality in the brain or carotid

circulation. She was investigated extensively at Broomfield Hospital. Trans-thoracic and a bubble Echo were normal. She had a positive ANA test with negative ANCA, double-stranded DNA and ENA. A full vasculitic screen was normal [including Thrombophilia, Lupus anti-coagulant and Factor V Leiden]. Bloods and an urine collection for catecholamines were also unremarkable. She was referred to the Royal London hospital for a tertiary neurological assessment.

The neurologists at the Royal London repeated the majority of the above described tests with similar negative findings. A repeat MRA was normal. After much discussion, it was thought that WWs symptoms may possibly be attributed to carotid artery vaso-spasm [similar to the spasming of the coronary arteries in Prinz-metal angina]. She was initiated on Verapamil 120mg daily with surprising results. Her symptoms completely resolved and she experienced no reoccurrence as long as she complied with the verapamil.

Discussion

The idea of cerebral blood vessel ‘spasm’ leading to arterial insufficiency and thus neurological symptoms is not a new concept. From its inception by Raynaud as a possible cause for gangrene in limb extremities (Raynaud 1862), it was expounded as a possible cause for Transient Ischaemic Attacks [TIAs] as early as the 1900s by (Osler 1906). However as time passed, thromboembolism and arterial plaques became central tenements towards the treatment of Cardiac and Cerebro-vascular disease. However a search of the literature has revealed a recent renaissance for the notion of artery spasm in the inducement of neurological symptoms.

The largest case series of its kind has just been published by (Ducros, et al. 2007), recounting the diagnosis and presentation of the Reversible Cerebral Vasoconstriction Syndromes(RCVS) involving 67 patients. In his review of the subject, (Calabrese, et al. 2007) defines RCVS as a ‘group of disorders characterized by prolonged but reversible vasoconstriction of the cerebral arteries, usually associated with acute-onset, severe recurrent

headaches with or without additional neurologic signs and symptoms'. The RCVS syndromes cover other eponymous appellations like Call-Fleming syndrome, Migraine Vasospasm, Post-partum angiopathy and drug-induced cerebral angiopathy, to mention a few. Its patho-physiology is not well understood but is thought to be primarily due to sudden and unwelcome alteration of cerebral vascular tone. It is common among young females (Chen, et al. 2006a, Calabrese, et al. 2007) and may be provoked by factors like pregnancy (Singhal and Bernstein 2005), medications or through substance abuse (Calabrese, et al. 2007). Its diagnosis at present involves a process of exclusion and demonstration of a 'reversible obstruction' in the cerebral vasculature by modern day imaging techniques such as CT angiograms or MRAs.

Amaurosis fugax is a clinical manifestation of retinal hypoperfusion. With regards to isolated Amaurosis Fugax [without headaches as in WW's case], the most common cause is generally attributed to thrombo-embolism from the heart or the carotid but there have been a number of case reports describing vasospasm [of the ophthalmic or carotid artery] as the primary cause of symptoms. (Petzold, Islam and Plant 2003), (Heckmann, et al. 2003), (Burger, et al. 1991) and (Bernard 1999) have illustrated though fundoscopy, retinal photography and laser doppler flowmetry the existence of vasospasm in patients with transient monocular blindness. (Winterkorn, et al. 1993) went a step further and gathered 9 patients with recurrent attacks of amaurosis fugax despite anti-platelet therapy and demonstrated complete symptom resolution in all patients through the use of calcium-channel blockers [nimodipine or verapamil]. He also established the return of symptomatic attacks once the treatment was withdrawn.

Calcium-channel blockers help stabilise vasomotor tone in arterial vessels by acting on vessel walls to promote vasodilation. They have been shown to be effective in the treatment of Raynauds (Rodeheffer, et al. 1983), coronary artery spasm (Hillis and Braunwald 1978) and cerebral artery spasm post sub-arachnoid haemorrhage (Pickard, et al. 1989). An alternative possible treatment modality was highlighted by (Yokohama, et al. 2006) who successfully used a stellate ganglion block to produce immediate relief

of vasospastic symptoms in a young 35-year old patient. However this technique has not proved to be as effective in preventing further attacks.

Thus, vasospasm should always be born in mind when considering cases of recurring TIAs or migraine-associated with neurology that seem resistant to treatment. Modern-day imaging has advanced so much so that today it is possible to demonstrate the vasospasm of arteries. Increased detection and a higher profile, will lead to successful treatment and resolution of symptoms in certain cases. Patients seem to respond well to treatment in the form of calcium-channel blockers and possible simple life-style adjustment [such as altering medications or stopping drug dependency].

However, vasospasm with transient neurology may well be misdiagnosed as a stroke that was successfully thrombolysed and it adds to the expanding list of stroke mimics that pose a clinical conundrum to the physician in the acute setting-particularly in the light of the short window for thrombolysis and clot removal. Careful history taking [repetitive nature of symptoms over-time] and examination along-with better and more advanced imaging at stroke outset seem to be the only solution to the riddle of distinguishing vasospasm from thrombo-embolism.

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