Beware of a diagnosis of TIA in a young patient

Sarim Kamal Omar MBBS, MRCPI, Mazen Rizeq FRCP

Cavernomas are abnormal vascular lesions consisting of many small lobules, like a bunch of small berries. They may be asymptomatic or they may present with seizures, in around 60% of cases, or symptoms similar to a transient ischaemic attack (TIA) or stroke. Here, the authors describe a case of a 30-year-old woman who presented with TIA-like symptoms for which treatment was started. Imaging revealed a spinal cord cavernoma, after which the course of management was changed and the patient referred to the neurosurgeons.

Presentation
A 30-year-old lady, an ex-smoker, with a background history of anxiety and asthma, presented to A&E with a sudden onset of headache, neck pain, right shoulder pain which radiated to the right arm along with numbness, and heaviness of the same side. She denied any chest pain or shortness of breath. The symptoms virtually resolved in about 20 minutes. Interestingly, she had a similar but more severe episode on the left side ten years earlier and was investigated for a TIA but no definite diagnosis was made.

On admission she looked well and observations were normal. CNS examination showed slightly reduced power of 4-5 / 5 in the right upper limb, plantars were downgoing and there were no cerebellar signs. Bloods, ECG, chest X-ray and CT brain were normal. She was started on aspirin 300mg once daily by the admitting team and was referred to the stroke team for further assessment.

The next day she developed further numbness on the same side and became significantly ataxic and was seen by the stroke team at the request of the admitting team. The original diagnosis of TIA was queried given her young age and the previous episodes; hence the possibility of either a demyelinating disease, a posterior circulation lesion, a possible dissection or stenosis in the vertebral artery was entertained. Hence an urgent MRI to include the cervical spine was arranged. Interestingly enough, this showed a haemorrhage in a cavity within the right side of the cervical cord at C2/C3, lots of surrounding high T2-weighted signal in the cord with mild contrast enhancement around the edges (see Figures 1 and 2).

A diagnosis of spinal cord cavernoma was made and the acute symptoms were thought to be most probably due to a bleed.
Case notes  TIA diagnosis

(Please note the patient received aspirin 300mg on two occasions.)

In light of the MRI findings, aspirin was stopped immediately and the patient’s case was discussed with the neurosurgeon, who suggested managing the patient expectantly, while plans for possible stereotactic radiosurgery would be considered after the MDT meeting. At the time of preparing this case, we understand that the patient has had a dynamic magnetic resonance angiogram (MRA) followed by a focused spinal angiogram. The latter has excluded any arteriovenous shunting in this area.

At a follow-up clinic the patient remained well with no recurrence of her symptoms. She has decided to undergo the stereotactic radiosurgery, and has regular follow-ups with the neurosurgical team.

Discussion

A cavernous malformation, also known as a cavernous haemangioma or cavernoma, is an abnormal vascular lesion consisting of many small lobules, like a bunch of small berries. These contain blood products in different stages of evolution. The ‘sinusoidal’ compartments are enclosed by abnormally thin and quite fragile endothelialised walls. Unlike an arteriovenous malformation, there is no large feeding artery and no large draining vein in a cavernoma. However, there frequently is an associated venous angioma.

The prevalence of cavernous malformations is between 0.1% and 0.5%; they are about as common as brain arteriovenous malformations, and far less common than brain aneurysms.

Most cavernomas are found in the supratentorial region of the brain hemispheres, but up to 1 in 4 or 5 are found in the posterior fossa / infratentorial, especially in the pons region of the brainstem. Much less commonly, a cavernoma may be found in the spinal cord, as was the case for our patient, but this may be more likely to occur in patients with a family history of cavernous malformations.

Cavernomas may be asymptomatic, or may present with seizures (60%) or, as in our patient, with TIA- / stroke-like symptoms with progressive neurological impairment or ‘deficits’ (50%). Some can present with hydrocephalus or raised intracranial pressure depending on their size and location. It is uncommon for cavernomas to cause sudden catastrophic or devastating neurological injury, but the progressive brain or spinal cord injury associated with cavernomas may be severely disabling as time goes on.

The risk of haemorrhage is between 0.5 and 1% per year. The re-haemorrhage rate varies in the literature, but is between 4 and 10% per year.

Cavernomas are best detected using MRI cerebral angiography.

The treatment is surgery. There is no effective radiation treatment. The one exception is a possible cavernoma variant that occurs in the venous sinuses (intrasinus cavernoma) which has been reported to be susceptible to radiation (eg gamma knife or stereotactic radiosurgery). Cavernomas can develop in regions of brain that have previously been exposed to radiation.

In general, a cavernoma that is enlarging radiologically and / or symptomatic should be considered for surgery. Even those located in the brainstem or spinal cord or in other highly ‘eloquent’ areas (areas of the cortex which if removed, could lead to a loss of linguistic ability or sensory processing or can even lead to a degree of paralysis or complete paralysis at times), should be considered for removal if symptomatic and if relatively safely accessible surgically.

In a recent study published by Kharkar, et al. 14 patients were included from 1989 to 2002. The mean age at presentation was 42 years. Four lesions (29%) were located in the cervical region and 10 lesions (71%) were present in the thoracolumbar spinal cord. Seventy-one per cent were managed conservatively. Four patients (29%) were treated surgically. The mean duration of symptoms before presentation was 33 months. The mean duration of follow-up from the time of presentation was 42 months. In this series, patients harbouring a symptomatic spinal cord cavernous malformation did not have significant permanent neurological decline during the follow-up period when treated with the conservative approach of observation. This is a similar situation to the case described above as, at the time of writing, our patient was still under active follow-up with no neurological deterioration, yet surgery was still planned and considered.

Three cases of surgically verified intramedullary cavernous angiomas (cavernoma) of the spinal cord were reported in 1995, by Gordon, et al. The most common presenting symptom was pain, which in all cases preceded weakness. In two of the cases the typical progression of sudden paroxysmal worsening of symptoms accompanied by pain was noted. This was thought to be related to bleeding in the lesion. In the case we have described, we believe that the two doses of aspirin given to the patient led to a bleed in the cavernoma itself and this was clearly reflected in her new neurology findings the day after and the spontaneous resolution of symptoms within a few days.
Prescribing Information

Matoride XL 18mg, 36mg & 54mg
Prolonged Release Tablets
(methylphenidate hydrochloride)

Please consult the Summary of Product Characteristics before prescribing

Indication: As part of a comprehensive treatment programme for Attention Deficit Hyperactivity Disorder (ADHD) in children aged 6 years of age and over when remedial measures alone prove insufficient. Must be under the supervision of a specialist in childhood behavioural disorders. Dosage & Administration: Tablet for oral administration, taken once daily in the morning with or without food. Must be swallowed whole with the aid of liquids, and must not be chewed, divided, or crushed. Pre-treatment screening: baseline evaluation of patient’s cardiovascular status (blood pressure and heart rate), concurrent medication, past and present co-morbid medical and psychiatric disorders or symptoms, family history of sudden/unexplained death and pre-treatment height and weight on a growth chart. Dose titration: Start at the lowest possible dose. Dose adjustment in 18mg increments at approximately weekly intervals. Patients currently using methylphenidate. Consider lower doses of short-acting methylphenidate formulations. Patients Currently Using Methylphenidate: Recommended dose conversion from 5, 10, 15 mg methylphenidate three times a day is 18, 36, 54mg Matoride XL daily, respectively. Discontinue if improvement not observed after dosage adjustment over one month. Contraindications: Hypersensitivity to the active substance or to any of the excipients: glaucoma; phaeochromocytoma; during, or within 14 days of discontinuing treatment with MAOIs, hydroxyphenyl or thyroxine; diagnosis/ history of severe depression, anxiety, or anorexia, neuropsychiatric disorders, suicidal tendencies, psychic symptoms, severe mood disorders, mania, schizophrenia, psychopathology/ borderline personality disorder, diagnosis/ history of severe and episodic (Type I) Bipolar (affective) Disorder (if not well-controlled); pre-existing cardiovascular disorders; pre-existing cerebrovascular disorders, cerebral aneurysm, vascular abnormalities including vasculitis or stroke. Precautions: Long-term use (more than 12 months): ongoing monitoring (at each dose adjustment and then at least every 6 months) for: cardiovascular status (blood pressure and pulse); neurological signs and symptoms (for those with cerebrovascular disorders and additional risk factors); psychiatric/neurological conditions (including suicidal ideation, possible precipitation of a mixed/manic episode in patients with comorbid bipolar disorder; epilepsy as may lower convulsive threshold); growth (height, weight and appetite). De-challenge recommended at least once yearly. In new to methylphenidate: Where medical conditions compromised by increases in blood pressure / heart rate. Not recommended in known structural cardiac abnormalities, cardiomyopathy, serious heart rhythm abnormalities, or other serious cardiac problems due to symptomatic effects. Potential for abuse, misuse or diversion in drug or alcohol dependency. Not to be taken by patients with rare hereditary problems of galactose intolerance, the Lapp lactase deficiency or glucose-galactose malabsorption. Should not be used in patients with pre-existing severe GI narrowing (pathologic or iatrogenic) or in dysphagia or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools. No experience in renal or hepatic insufficiency. If leukopenia, thrombocytopenia, or significant swallowing difficulties. The tablet shell may be noticed in stools.

A case was described in the British Journal of Neurosurgery where a 31-year-old male presented with a one year history of left leg weakness and right leg sensory changes. Myelography revealed a probable intramedullary lesion at T4 and CT confirmed its intramedullary nature. At operation, an almost black, firm mass was found projecting from the dorsal aspect of the cord and was totally excised. Histological examination revealed it to be a cavernous angioma.

In a recent paper published by Papageorgiou, et al. from Greece, a 67-year-old woman presented with walking difficulties with acute onset of pure right leg monoparesis with moderate spasticity. The initial diagnosis was lacunar stroke, but the brain MRI revealed a right temporal cavernoma, not associated with her monoparesis. The consequent spinal MRI revealed an intramedullary lesion at the T1 level, consistent with a cavernoma.

In a French study of a group of spinal cord cavernomas, published by Labaigue, et al. initial symptoms were progressive (60.37%) or of acute myelopathy (37.7%). Clinical symptoms were related to spinal cord compression (50.94%) and haemorrhage (41.50%). Using McCormick classification, 22 patients were autonomous (grades 1-2), 12 handicapped (grade 3), and 3 bedridden (grade 4) at the end of the follow-up. This study defined clinical and MR patterns of spinal cavernomas. Surgery lasting improved more than half of the patients.

In a recent paper from Finland, published by Kivelev, et al. from the Department of Neurosurgery at Helsinki University Central Hospital, a unique case of an intradural extramedullary spinal cavernoma had a successful surgical removal following a bleed in the cavernoma itself. Interestingly, this case was also diagnosed as a TIA and aspirin was given at the time of admission, in a similar way to our patient.

In summary, we have described a patient who presented with TIA-like symptoms and was started treatment in-line with recommended guidelines, but imaging revealed a spinal cord cavernoma, after which the course of management changed and patient was also referred to the neurosurgeons for further management, in addition to being followed up by us.

Case notes

TIA diagnosis

No conflicts of interest were declared.

Acknowledgments

The authors would like to thank Dr P McMillan for his expert reporting of the MRI scans and to Dr R. Nahas for his help in formulating the MRI scans into this paper.

References


Adverse events should also be reported to Sandoz Ltd, 01276 698020 or uk.drugsafety@sandoz.com.