Author’s response to reviews

Title: Subarachnoid haemorrhage due to intracranial vertebral artery dissection presenting with atypical cauda equina syndrome features: case report

Authors:

Lloyd Steele (lloyd.steele@nhs.net)
Muhammad Raza (muhammadhasan.raza@nhs.net)
Richard Perry (richard.perry3@nhs.net)
Neil Rane (neil.rane@nhs.net)
Sophie Camp (sophie.camp@nhs.net)

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Author’s response to reviews:

We thank the reviewers for their thorough review and suggestions, which have led to an improved manuscript. All of these have been responded to in a point-by-point fashion in a formatted form as a supplementary file. These responses are also listed below - unformatted (page and line numbers refer to the tracked version of the manuscript).

Editor Comment 1

Please make clear the advance your case report makes on the literature.

Response: The novelty of this case to our department, and in the literature, was its atypical presentation initially with spinal symptoms. We have highlighted this in the revised manuscript.

P2, L8

In this case we describe a rare presentation of SAH secondary to vertebral artery dissection (VAD) that initially presented with spinal symptoms, highlighting this as a consideration in patients presenting with sudden-onset spinal symptoms. Intracranial VAD with SAH is very rarely associated with spinal symptoms in the acute phase.[5, 6]

Reviewer 1:

Comment 1
“Facial palsy is an acknowledged presenting sign in SAH with or without VAD. When facial palsy occurs with other neurological deficits such as consciousness change, cerebellar signs or pyramidal signs, even no headache, SAH is one of the differential diagnosis. Therefore, the present case report is not a novel finding.”

Response: In light of this comment we have emphasised in the manuscript the novelty came from the initial spinal symptoms. As per reviewer 2 we have then commented on the subsequent development of more typical SAH features, including facial nerve palsy.

P2, L8

In this case we describe a rare presentation of SAH secondary to vertebral artery dissection (VAD) that initially presented with spinal symptoms, highlighting this as a consideration in patients presenting with sudden-onset spinal symptoms. Intracranial VAD with SAH is very rarely associated with spinal symptoms in the acute phase.[5, 6]

P7, L17

In our case, a convexal/subconvexal focus of blood affecting the parietal lobule was seen, as well as more generalised SAH, which could explain why the patient developed headache, facial nerve palsy, and confusion. In various case series of cSAH, reported features have included focal and transient motor and/or sensory symptoms (42-73%), including facial nerve palsy and pronator drift;[15, 16] headache (18-65%); and confusion (9.8%).[17-20] Although cSAH by definition spares the basal cistern and Sylvian fissures (which our case does not), previous descriptions of cSAH have been made when these regions have not been spared.[21-23] Alternative considerations for facial nerve palsy are compression by the aneurysm (which imaging did not suggest in our case) and vascular spasm disturbing the blood supply of the facial nucleus.[24, 25]

Comment 2

“There was no endovascular intervention or surgical treatment but only conservative treatment for this case and the follow-up time was only nine months. Management strategy and long-term outcome including rebleeding was not able to be clarified.”

Response: The treatment decision in this case was a difficult one. We have explained this decision in the manuscript, together with the ongoing management plan (See below).

P6, L3

The patient was discussed at the neurovascular multi-disciplinary team (MDT) meeting where conservative management of the VAD was advocated based on a delayed and atypical presentation, treatment risking vessel occlusion, and patient choice.
Imaging surveillance is to continue, with repeat MRA planned after a further 6 months.

Factors influencing our decision were the delayed and atypical presentation, the good neurological status on admission, patient aversion to operative management, and the feasibility of intervention - with the location of the fusiform aneurysm necessitating occlusion of the posterior inferior cerebellar artery. Flow diversion was also an off-label option, with limited data suggesting comparable outcomes in the long-term to conventional techniques, but with potentially greater short-term complications and the need for dual antiplatelet therapy.[30]

Comment 3

“The results of neurological examinations were confusing. peripheral nervous system examinations should comprise DTR and weakness patterns (e.g. distal/proximal, root distribution, etc). Several domains of neurological examinations were not mentioned. Right pyramidal or pronator drift involved both upper and lower limbs? Cerebellar signs? Plantar reflex? “

Response: Initially we restricted the examination findings to positive findings, but in this revised version we have expanded the examination.

His vital signs on admission revealed hypertension (blood pressure 160/92mmHg) and tachycardia (heart rate 103 beats per minute), but were otherwise within normal limits. Glasgow coma scale was 15. On examination there was a lower motor neurone pattern of left-sided facial weakness (House-Brackmann grade IV). The remainder of the cranial nerve examination was normal. Tone was normal and there was no ankle clonus. Pyramidal drift was present in the right upper limb, but muscle power was Medical Research Council grade 5/5 in all muscle groups. Deep tendon reflexes were elicited (normal) apart the ankle reflex, which was absent bilaterally. The plantar response was downgoing bilaterally. Light touch and pain sensation were reduced, but present, to T6 bilaterally - most notably across L4-S1 and S3-S4 (saddle anaesthesia) – and vibration sense was reduced to the ankle bilaterally. Gait was unsteady and the patient was unable to tandem walk. There was no limb ataxia. Anal tone was normal, there was no significant residual volume on urinary catheterisation, and the patient could feel a catheter tug.
“The authors speculated that the facial N palsy might have been due to the VA dissection and SAH. Curiously, the authors did not present MRI at the level of upper brainstem (i.e., around the facial N nucleus) in Figure 2: it may show the anatomical relationship between the facial N and the VA dissection. I suggest that the authors modify the Figure 2 by providing an MR image at the upper brainstem level.”

Response: We thank the review for this comment. The imaging has been reviewed and the facial nerve palsy did not appear to be secondary to a mass effect of the aneurysm. The neurino-interventional radiologist most involved in the case has reviewed the imaging and manuscript, and is currently preparing this figure. We have added the manuscript about the absence of direct aneurysmal compression (see below), and further information on the facial nerve palsy is addressed in comment 3.

P7 L22

Alternative considerations for facial nerve palsy are compression by the aneurysm (which imaging did not suggest in our case) and vascular spasm disturbing the blood supply of the facial nucleus.[24, 25]

Comment 2

“In Figure 1 (lumbar spine MRI), the presence of spinal SAH could not be seen. Meanwhile, the authors stated that diffuse FLAIR hyperintensity was seen on the cervicothoracic spine MRI. I suggest that the authors add a cervicothoracic spine MR image to Figure 1 to prove that diffuse SAH had actually been present in the spinal canal.”

Response: We thank the reviewer for identifying this area of confusion. An MRI of the brain an cervicothoracic MRI was performed at the same time as the MRI head, but the diffuse SAH was confined to the cerebrum. We have clarified this in the revised manuscript, and also commented on why the spinal SAH may not have been observed as based on the literature this would seem the most likely cause for this unusual case.

P5, L17

MRI of the brain showed diffuse subarachnoid fluid-attenuated inversion recovery (FLAIR) hyperintensity concerning for blood, including in the basal cistern and Sylvian fissures. There was also a focus of cortical/subcortical high signal in the left superior parietal lobule (Figure 2). MRI of the cervico-thoracic spine did not demonstrate SAH.

P6, L6
Very rarely, intracranial VAD has been associated with spinal extension of SAH.[5, 6] Although MRI in our case did not suggest a spinal SAH, in one case report MRI imaging of the spine was initially normal.[14] Moreover, the intensity of back pain in patients with SAH often correlates with the amount of blood in the lumbo-sacral subarachnoid space, [9] and the back pain had largely resolved by the time of admission.

Comment 3

“Regarding the clinical course, the facial N palsy was preceded by symptoms of the cauda equina injury. Considering the fact that the facial N is anatomically close to the enlarged VA, one might rather think that the facial N palsy would develop first when the VA dissection occurs. Why did the symptoms of the cauda equina injury develop prior to the facial N?”

Response: We thank the reviewer for this interesting comment. We have added a possible explanation for why facial nerve palsy developed after the spinal symptoms.

P7, L13

In a series of 8 patients with spinal SAH, 25% also had cortical/convexal SAH (cSAH).[13] The authors suggested that this was because of the redistribution of blood products in the spinal subarachnoid space. In our case, a convexal/subconvexal focus of blood affecting the parietal lobule was seen, as well as more generalised SAH, which could explain why the patient developed headache, facial nerve palsy, and confusion. In various case series of cSAH, reported features have included focal and transient motor and/or sensory symptoms (42-73%), including facial nerve palsy and pronator drift;[15, 16] headache (18-65%); and confusion (9.8%).[17-20]

Comment 4

“Ruptured VA dissections have a risk of rebleeding, and in many neurosurgical institutions, they will be treated with IVR to prevent rebleeding. While the authors described that [neurovascular multi-disciplinary team (MDT) meeting where conservative management of the VAD was advocated], I think it was lucky that the patient did not sustain rebleeding. I suggest the authors to explain for what reason conservative management had been considered safe in this case.”

Response: The treatment decision in this case was a difficult one. We have explained this decision in the manuscript, together with the ongoing management plan (See below).

P6, L3

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Comment 5

“The authors need to indicate whether consent for publication was granted by the patient (or by their family).”

The patient has consented to publication and has signed a consent form for publication of the case and related images which we can provide.