Author's response to reviews

Title: Aqueductal Developmental Venous Anomaly as an Unusual Cause of Congenital Hydrocephalus: Case Report and Review of the Literature

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Author's response to reviews: see over
Dear Editor,

Thank you for your notes on our submission, “Aqueductal Developmental Venous Anomaly as an Unusual Cause of Congenital Hydrocephalus: Case Report and Review of the Literature” (MS 1880386470576246). We have addressed the reviewers’ comments to the best of our ability. Please find below our responses:

Reviewer #1:

1) “The proposed DVA is actually in the quadrigeminal cistern and the vein of Galen is significantly more dilated than would be expected.”

After re-reviewing the MRI of the brain with contrast with our neuroradiologist, we have found that the DVA is intraventricular (inside the posterior 3rd ventricle) in the region of the aqueduct rather than the quadrigeminal cistern. Additionally, the vein of Galen is normal size for the patient’s age.

2) “…follow up is needed to rule out the possibility of arteriovenous malformation or arteriovenous fistula.”

We closely examined the MIR of the brain with contrast with our neuroradiologist and found that the MRI does not suggest any vascular malformation. T2-weighted images do not demonstrate any abnormal signal flow voids. Additionally, we do not feel that more invasive investigation with cerebral angiography and all its attendant risks is warranted in this case.

Reviewer #2:

1) “It is not so unique an occurrence…with already several cases described in the literature.”

In our report, we reviewed 10 other cases of hydrocephalus previously described in the literature. In our opinion, what makes our case unusual and worthy of publication is that our patient would be the youngest reported case in the literature with hydrocephalus occurring in the perinatal period from DVA.

2) “…the MR with contrast images presented, on these grounds, could just be the documentation of a venous congestion…”

We along with our neuroradiologist would have expected venous congestion from hydrocephalus to be a global phenomenon rather than an affliction just affecting a single vein at the region of the cerebral aqueduct. After careful re-examination of the MRI of the brain with
contrast, we saw no other dilated or congested veins.

3) “...to confirm a venous anomaly an MR angiography image at least should be added.”

We do not have an MRA or MRV to add to our case report. We did not initially suspect a venous anomaly as a possible cause for hydrocephalus, so we did not order an MRA/MRV upfront. Moreover, we do not feel that the risks justify putting the infant through general anesthesia again to obtain an imaging study that would confirm what is already seen on MRI of the brain with contrast – a DVA causing obstructive hydrocephalus at the level of the origin of the cerebral aqueduct.

Thank you for considering our submission. We hope that our comments provide clarity and facilitate understanding of our manuscript. Please let us know if there are any other questions or concerns.

Sincerely,

Andrew Jea

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