Poster Number 006. Case Report

SPONTANEOUS INTRACRANIAL HYPOTENSION: AN UNDERRECOGNIZED CAUSE OF DEBILITATING SYMPTOMS IN HYPERMOBILE CONNECTIVE TISSUE DISORDERS

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INTRODUCTION

Spontaneous intracranial hypotension (SIH) is associated with heritable disorders of connective tissue. SIH is an underrecognized cause of disabling headaches and multisystem dysfunction that must be included in the differential with other neurological and autonomic disorders.

Evaluation for SIH is fraught with difficulty due to specificity to varied etiologies. Etiologies include dural puncture from a bone spur, ruptured perineural cyst, dural ectasia, and, CSF venous fistula. Fistula likely represents 30% of causes of SIH but is the most difficult to diagnose.

Management of SIH is substantially different compared to other complications in this patient population.

CASE PRESENTATION

A 22-year old female with a history of migraine, joint hypermobility and Postural Tachycardia Syndrome (POTS), experienced a thunderclap headache while running. Her headache was positional in nature and was associated with photophobia, nystagmus, tinnitus, nausea, worsening dizziness, tachycardia, cognitive dysfunction and a sound of fluid rushing in the head with positional changes.

The patient was unable to spend prolonged periods of time upright, and, trialed more than 18 migraine medications without improvement. More aggressive treatment of the POTS did not improve headache.

Brain MRI found a pituitary cyst, and, a total spine MRI was unremarkable.

Hormone evaluation found a mildly elevated prolactin.

Lumbar puncture (LP) showed mildly elevated opening pressure and caused severe worsening of headache. However, the patient was not positioned properly. Following LP, lumbar epidural blood patch (EBP) resolved the headache for 3 days before returning to baseline.

One year after the onset, the patient had a cisternogram which showed questionable evidence of a CSF leak. Genetics confirmed the diagnosis of hEDS. A second lumbar EBP resolved the headache for 4 weeks, followed by a lumbar and thoracic EBP which provided 6 months of relief.

A CT myelogram was performed after recurrence of symptoms. No clear leak site was identified, but more than 20 perineural cysts were identified. High volume (46ml) multi-level

EBP was targeted to C7-T1, T6-T7, T10-11, and L2-3 and this provided 8 months of headache relief.

With recurrence of symptoms the patient then had 7 repeat single level EBPs, each with durable improvements. Three and a half years after the onset of headache, MR myelogram and lateral decubitus digital subtraction CT myelograms were performed due to suspected dural fistula at T8/9.
Laminectomy found two dural fistulas at T8/9 which were ligated with aneurysm clips and fibrin glue was applied.

Following surgery, the patient had a complete resolution of low-pressure headache and was able to return to full time school and regular exercise. She did experience episodes of rebound high pressure headaches that were treated with acetazolamide.

A syncopal event 11 months post-op caused a recurrence of severe positional headache, photophobia, vision changes, tinnitus, nausea, worsening dizziness, tachycardia, cognitive dysfunction and a sound of fluid rushing in the head with positional changes.

CONCLUSION
This case highlights the challenges with recognition, evaluation, and treatment of SIH. SIH is an important differential to consider in patients with heritable disorders of connective tissue.

REFERENCES

Kranz PG, Gray L, Malinzak MD, Amrhein TJ. Spontaneous Intracranial Hypotension: Pathogenesis, Diagnosis, and Treatment.Neuroimaging Clin N Am. 2019 Nov;29(4):581-594.

Farb RI, Nicholson PJ, Peng PW, Massicotte EM, Lay C, Krings T, terBrugge KG. Spontaneous Intracranial Hypotension: A Systematic Imaging Approach for CSF Leak Localization and Management Based on MRI and Digital Subtraction Myelography. AJNR Am J Neuroradiol. 2019 Apr;40(4):745-753.

Chan SM, Chodakiewitz YG, Maya MM, Schievink WI, Moser FG. Intracranial Hypotension and Cerebrospinal Fluid Leak. Neuroimaging Clin N Am. 2019 May;29(2):213-226.

DECLARATION OF INTEREST
None