INTRA-DIALYTIC INTRACRANIAL PRESSURE MONITORING IN A PATIENT WITH IDIOPATHIC INTRACRANIAL HYPERTENSION AND DIALYSIS DISEQUILIBRIUM SYNDROME

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INTRODUCTION

Dialysis disequilibrium syndrome (DDS) is a rare neurological manifestation of intermittent haemodialysis (HD), thought to be due to promotion of cerebral oedema and characterised by symptoms of raised intracranial pressure (ICP).

We present a 25 year old female with focal segmental glomerulosclerosis (FSGS) in end-stage renal disease (ESRD). She commenced HD at The James Cook University Hospital at the age of 18 years.

Following investigations for loss of vision, a diagnosis of IIH was made and a lumbo-peritoneal (LP) shunt was placed at age 20 years in order to preserve eyesight. She continued to have headache during HD and we became concerned her shunt had blocked; however opening pressure upon LP was 16cm H2O.

The headache was attributed to low CSF pressure, exacerbated by fluid shifts during HD. Cessation of HD led to relief of headache.

As she was due to be transplanted three months later, she did not wish for LP shunt revision.

Idiopathic Intracranial Hypertension

Idiopathic Intracranial Hypertension (IIH) is rare, with an incidence of 0.9 -1.5 cases/100,000. Obese women of reproductive age are most at risk, but IIH can affect people of any age or weight. At James Cook, we perform 6-10 shunts annually for IIH, covering a population of 1.2million.

Symptoms of IIH include headache, transient visual obscurations, intracranial noises, photopsia, retrobulbar pain, diplopia and potentially sustained visual loss, as in our case.

Diagnosis requires exclusion of other neurology, elevated ICP (opening pressure >25cmH20), normal CSF composition and normal neuroimaging.

Differential diagnosis includes space-occupying lesion, venous outflow obstruction, obstructive hydrocephalus, decreased CSF absorption, increased CSF production and malignant hypertension.

Pathophysiology includes intracranial venous hypertension but it is unclear if this is a cause or effect of excess IIH. Cerebral oedema has also been postulated, but there is no MRI or pathologic evidence to support this. The theory regarding those with central obesity involves raised intra-abdominal pressure, pleural pressures, cardiac filling and central venous pressure.

Uraemia is listed as a rare cause of IIH but this did not apply to our patient who had excellent compliance and URRs.

Dialysis-Associated Headache

Headache is one of the commonest side effects of HD and there are a multitude of potential causes.

These include: blood pressure change, medications, electrolyte disturbance, (e.g. Sodium), caffeine withdrawal, intraocular pressure change, stress or anxiety, as well as other headache syndromes.

Given we had systematically excluded these in our complex patient, we began to suspect her symptoms were due to a variant of DDS.

METHOD

Our patient's initial valve-less LP shunt system was revised upon sustaining persistent low pressure headache to a shunt with a 5:35cm gravitational valve. Unfortunately FSGS recurred in her transplant after 2 years and she recommenced HD. The HD-associated headache returned and she became quite distressed by this. She was referred urgently back to our neurosurgical team.

ICP monitoring was undertaken while on the HD unit, to determine if the headache was due to intracranial pressure change. We expected to discover low pressure headache occurring during HD, due to her functioning LP shunt.

RESULTS

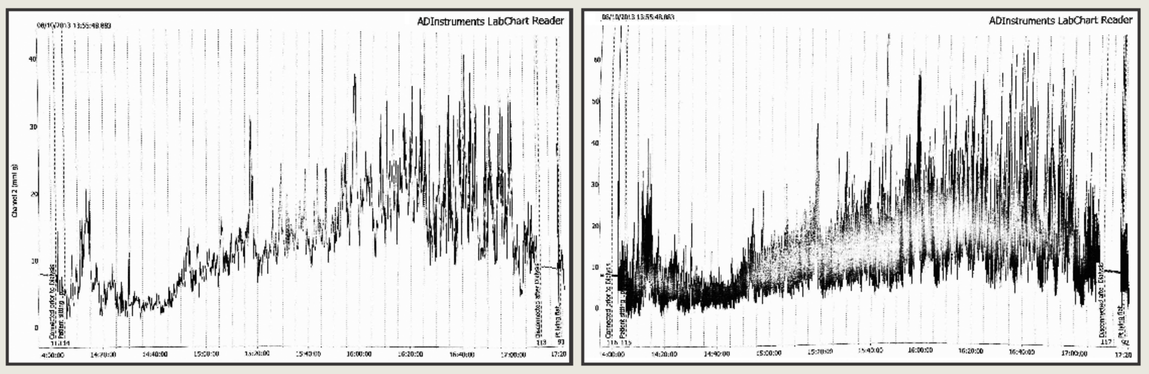

Figure 1: This Intracranial Pressure (ICP) monitor readout shows escalating pressure readings throughout an afternoon session of Haemodialysis.

Converse to our expectation, ICP tracing during HD revealed a progressive rise in ICP, from a baseline below 5mmHg, culminating at over 20mmHg, with spikes above 40mmHg. This ICP rise correlated with reported symptoms of headache during HD and was the opposite to what we would have expected with IIH.

Due to the gravitational valve in her LP shunt, CSF drainage via the shunt occurs at a higher opening pressure when the patient is upright. She was advised to dialyse in a recumbent position, thus reducing shunt valve opening pressure. This increased CSF drainage, reducing ICP during HD and relieving headache.

We concluded DDS was the cause of these unexpected findings and therefore that DDS was the underlying cause of our patient's headache.

DISCUSSION

This patient is not morbidly obese so it may be that her renal condition has precipitated her IIH. The diagnosis of IIH came after she started HD; an LP shunt was inserted not long before transplantation. This patient's IIH did not fully resolve when her renal function normalised with transplantation – at that time she had a functioning shunt in place.

She suffered low pressure headaches improved by the insertion of a gravitational valve pre-transplant. The renal transplant stopped her headaches because she did not require dialysis; however, we do not have any evidence to confirm that her IIH resolved during that time period. Indeed, had it resolved and she still had a functioning shunt, she would have probably developed chronic low pressure headaches. When she restarted HD, her elevated pressure was better controlled in a recumbent position, whereas normally, ICP is increased in recumbency.

We believe our results support the theory of raised ICP during HD in patients with DDS. Cerebral oedema is a likely causative factor for raised ICP but is not definitively proven by ICP measurement alone. It is feasible that an osmotic shift of water occurring as a result of rapid clearance of small molecules during HD could cause cerebral oedema and raised ICP.

In IIH, where the patient is already at risk of raised ICP and has reduced cerebral compliance, relatively small pressure changes caused by cerebral oedema may be enough to make the patient symptomatic.

CONCLUSION

Our results confirm evidence of raised ICP during HD in support of DDS, in a patient with previously diagnosed IIH. A gravitational valve in her LP shunt gave us a unique opportunity to lower her ICP during HD with positional change alone. This greatly improved her symptoms.

ICP monitoring in an HD patient is rare; we recommend it is considered for those with a neurosurgical history who sustain unexplained and disabling headache on HD.

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