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Case Report

Anaesthesia Section

Laparoscopic Adrenalectomy in a Patient of Von Hippel Lindau Syndrome with Ventriculo-Peritoneal Shunt-Anaesthetic management

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ABSTRACT

Von Hippel Lindau (VHL) syndrome has a predilection to manifest multiple haemangioblastomas in the retina and central nervous system. We report a rare case of raised intracranial pressure during bilateral laparoscopic adrenalectomy for pheochromocytoma in a patient of VHL syndrome who had a ventriculoperitoneal shunt.

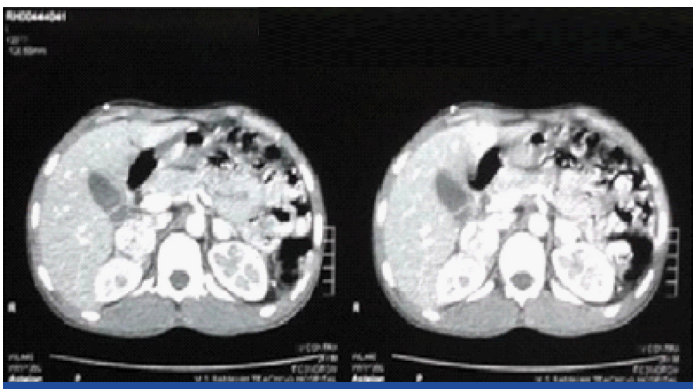
Keywords: Complication, Intraoperative, Laparoscopic surgery, Pheochromocytoma, Ventriculo-peritoneal shunt, Von hippel lindau syndrome

CASE REPORT

A 24-year-old, 55 kg male presented with reduced vision, episodes of headache associated with nausea and vomiting since 45 days. Contrast enhanced CT scan of the brain showed a non-enhancing hypodense space occupying lesion (38x31 mm) on the left side of the cerebellar hemisphere. MRI brain and spine revealed multiple craniospinal lesions in the left cerebellar hemisphere, medulla, and C7 spinal level with syrinx extending from C2 to D10 and compression of the 4th ventricle with dilatation of the 3rd and both lateral ventricles. A diagnosis of multiple haemangioblastomas was made.

In view of worsening cerebellar signs and the mass effect on brain he underwent emergency VP shunt procedure. The patient had a GCS of eight postoperatively, a repeat CT brain revealed extradural haemorrhage on the operated side which was evacuated with a craniotomy, following which patient's sensorium improved. Intraoperative hypertension (systolic of 220 mmHg) was observed during both surgeries requiring control with nitroglycerin infusion.

Evaluation for persistently high blood pressure with ultrasonogram of the abdomen revealed bilateral adrenal masses right > left (63 x 36mm > 31 x 21mm). The suspicion of bilateral pheochromocytoma, was confirmed by CT abdomen [Table/Fig-1]. 24h estimation of urinary normetanephrines revealed an elevated value (3,645µg/day).



[Table/Fig-1]: CT scan - Abdomen & pelvis - plain & contrast: bilateral bulky adrenals with heterogenous enhancement noted on contrast study

In view of the presence of multiple craniospinal haemangioblastomas and bilateral pheochromocytoma a diagnosis of Von Hippel Lindau (VHL) syndrome was made. The plan was to treat the craniospinal haemangioblastomas subsequent to bilateral adrenalectomy.

Persistently, elevated blood pressure was managed with a combination of Tab. Prazosin 2.5 mg tid and T. Atenolol 25 mg bd before surgery. Preoperatively he had a regular pulse of 80 bpm and blood pressure of 118/78 mmHg. Routine haematological, biochemical values, thyroxin, parathormone and cortisol levels were within normal limits. Chest X-ray, serum electrolytes, calcium and phosphorus were also normal. The 12 lead electrocardiogram and echocardiogram was normal.

Patient was accepted for bilateral laparoscopic adrenalectomy under ASA PS III under combined General and Epidural Anaesthesia. A 16G intravenous access was secured. An epidural catheter was sited at T12-L1 interspace and the catheter was fixed at 15 cms with tip probably at T6 level. The right radial artery and left subclavian vein were cannulated for invasive pressure monitoring. Patient was induced with Inj. midazolam 1mg, Inj. fentanyl 100 µg Inj. propofol 100mg IV and paralysed with Inj. Vecuronium 5mg IV. One minute prior to intubation Inj. Lidocaine 75mg was given IV to minimise response to intubation. Following intubation with 8.0 mm cuffed ETT anaesthesia was maintained with Oxygen-Air mixture with Isoflurane.

Once pneumoperitoneum was created the surgeon was requested to confirm the patency of the VP shunt by demonstrating the drainage of the CSF into the peritoneal cavity. The right sided tumour was removed first which was uneventful. The vitals were stable with a heart rate of 82bpm and BP of 130/80 mmHg. During resection of the left sided tumour there was a sudden surge in the BP to 210/110mm Hg with a HR of 42/min and no fresh ECG changes were noted. The combination of severe hypertension and bradycardia during pneumoperitoneum in the presence of a VP shunt, an increase in ICP was suspected. The surgeon was requested to stop the surgery and deflate the pneumoperitoneum. Inj. Frusemide 40mg stat and Inj. Mannitol(20%) 100ml IV was administered over the next twenty minutes to reduce the ICP and the vitals to normal range. The surgery was restarted and lasted 4½ hours. Patient was hemodynamically stable throughout the procedure. At the end of

the surgery the BP was 130/72mm Hg and HR of 87/min. Total fluid input was 4000mL, urine output was 2200mL and estimated blood loss was 400mL.

The patient was monitored in the ICU for 48h. Postoperative pain relief was provided with epidural infusion of 0.125% bupivacaine and fentanyl (2μ/mL) at 5mL/hr with Inj. Paracetamol 0.5g IV as rescue analgesic. Further course was uneventful.

DISCUSSION

VHL is a rare autosomal dominant disorder with a prevalence of about 1 in 50,000. Patients with retinal lesions present in their second decade whilst those with spinal cord and/or brain hemangioblastomas generally become symptomatic in their fourth decade. Hypernephroma, renal cell carcinoma, pheochromocytoma, cysts of kidney, pancreas, epididymis and liver can occur. Although benign, the tumors can cause pressure symptoms on surrounding structures or due to haemorrhage [1,2].

Management of anesthesia in patients with VHL should exclude the possible presence of pheochromocytomas. On search of literature we could find only two case reports describing the anaesthetic management of pheochromocytoma in patients with VHL [3,4]. In patients who are usually normotensive or have mild hypertension, the diagnosis is likely to be missed in the preoperative screening [1,2].

The perioperative mortality rate during elective resection of pheochromocytoma is 0% - 3%, and in undiagnosed or poorly prepared patient, this can be as high as 50%. The greatest frequency occurs in the fourth and fifth decades of life, with slightly higher female preponderance (60%).Incidence of pheochromocytoma in VHL syndrome has been reported between 10% and 20%. Clinically silent pheochromocytomas can become troublesome when subjected to stress [5,6].

Our patient had a bilateral secreting pheochromocytoma which resulted in persistently elevated BP and did not respond to conventional antihypertensive treatment. Epidural analgesia has been safely used to reduce the need for vasodilators and inhalational anaesthetics in these patients [4].

Controversy continues regarding the safety of performing laparoscopic surgery in patients with VP shunt.In previous studies monitoring of CSF shunt function was limited to clinical observation. The major concerns in patients with VP shunt, during laparoscopic procedures, are reduced visualisation due to the possibility of the presence of peritoneal adhesions related to previous abdominal interventions, and the risks of acute shunt dysfunction during peritoneal insufflation, shunt occlusion by soft tissue impaction, infection, and rarely retrograde carbon dioxide insufflation through the distal end of the catheter (Catheter/valve malfunction) [7-9].

Pneumoperitoneum pressures exceeding 15 mm Hg carries risk of shunt dysfunction contributing to raised ICP in patients with decreased cerebral compliance [9]. Patients with shunted hydrocephalus could be considered as being at low risk for laparoscopic procedures provided low insufflation pressure are used and if shunt function could be monitored intraoperatively.

In our case, we resorted to vigilant clinical monitoring to detect early haemodynamic signs of increase in ICP which enabled us to promptly identify and manage the episode of increased of ICP. Many techniques have been proposed to reduce the risk of retrograde shunt failure like clamping of the intraperitoneal end of the catheter, exteriorising and clamping the subcutaneous portion of the catheter and maintaining lower intra-abdominal pressures during pneumoperitoneum [10]. In our case patency of the shunt was demonstrated intraoperatively by visualization of CSF drainage.

CONCLUSION

Anaesthetic management of VHL with pheochromocytoma is by itself a challenge this case was more challenging by presence of a VP shunt.The case was successfully managed by vigilant clinical and invasive haemodynamic monitoring without worsening the co-existing neurological disorder.

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