BILATERAL ADRENALECTOMY AND SEGMENT VI RESECTION OF LIVER IN A YOUNG MAN WITH VON HIPPEL-LINDAU DISEASE
Nitin Agarwal*, Rana Anil Kumar Singh, Pooeyush Kumar. Department of Surgery and Renal Transplant Postgraduate Institute of Medical Education and Research (PGIMER) and Dr Ram Manohalohia Hospital, Baba Kharak Singh Marg, New Delhi, India

Background Bilateral pheochromocytomas can be seen in von Hippel-Lindau disease (VHL) and Multiple Endocrine Neoplasia (MEN) syndrome. Metastases occur in 10%–15% of all pheochromocytomas. Resection of the primary, metastasectomy and/or debulking are acceptable surgical options; despite liver being the second most common site of metastases, only a few reports of concomitant adrenalectomy and liver resection for pheochromocytoma are available.

Methods A 33-year-old man with a history of excision of hemangioblastoma of the cauda equina and retinal detachment presented with secondary hypertension and palpitations. Urinary catecholamine levels were raised, and CT scan revealed ~4.5 cm-sized, well-defined heterogeneously-enhancing lesions in bilateral adrenals with a 6.5 × 5 cm lesion in segment VI of the liver. MRI showed heterogeneous signal intensity in bilateral adrenals (predominantly hypointense on T1W and hyperintense on T2W) and also in segment VI of the liver with loss of fat planes. 131-I-MIBG scan with SPECT/CT showed uptake of tracer in the area of the bilateral adrenal masses and liver. With a diagnosis of VHL disease with bilateral pheochromocytomas, recurrent/residual spinal hemangioblastoma, right simple renal cyst, retinal detachment and a metastatic primary liver lesion, the patient was explored, and bilateral adrenalectomy with segmentectomy VI was performed.

Results The resection was performed with curative intent; histopathological examination revealed malignant pheochromocytoma with infiltrates in the liver. Resection margins were negative and the patient is normotensive six months postoperatively. Though bilateral pheochromocytomas are well-known in many familial syndromes, this is probably the first case of VHL to have undergone bilateral adrenalectomy and curative liver resection for metastatic pheochromocytoma in the available English-language literature. Hepatectomy for neuroendocrine tumours especially pheochromocytoma can be technically easier due to the well-circumscribed nature of the lesion, and also rewarding as complete removal leads to sudden relief from symptoms of hormone excess. Due to favourable histopathology in our patient, we are hopeful for a good long-term survival.

Conclusions Familial bilateral pheochromocytomas have a high propensity for malignancy. Surgical resection should be attempted for metastatic pheochromocytoma whenever possible, i.e., with limited metastatic disease, good performance status, and in an equipped centre.