Clinics in Surgery

Clinical Image
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Anterior Paravertebral Paraganglioma Revealed by Nutcracker Syndrome

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Clinical Image

A 49-year-old man with a history of paroxysmal postprandial epigastric pain for 2 years presented with intermittent hematuria. The abdominal pain was occasionally accompanied by lightning headache and sweating. His blood pressure was not detected before. The patient was initially diagnosed as gastroduodenal ulcer by gastroscopy, his symptoms didn't relieve after a standard anti-ulcer therapy. He was admitted for further assessment and examinations. Physical examinations revealed grade 3 hypertension, with the highest blood pressure 180/110 mmHg. His heart rate was within normal range. No abdominal mass was palpitated. Blood investigations of tumor biomarkers including AFP, CEA, and CA19-9 were within normal range too. Estimation of renal function in subjects with normal serum creatinine levels and cystatin C. Routine urinalysis presented RBC 500 cells/μl. With a normal range of urinary calcium secretion, stool guaiac test was negative. Endocrinal test identified elevated levels of urine catecholamine, Norepinephrine 296.07 μg/24 hr (normal range 16.69 μg/24 hr to 40.65 μg/24 hr), epinephrine 3.18 μg/24 hr (1.74 μg/24 hr to 6.42 μg/24 hr). Further imaging studies were evaluated. Abdominopelvic CT scan showed an irregular retroperitoneal soft tissue lesion, ranging from the inferior margin of the left adrenal gland towards the bottom edge of vertebrae L3. The lesion was supplied by left branch of abdominal aorta and left jejunoileal artery. It was fully enhanced in arterial phase. The body of pancreas and left renal vein were notably compressed. Somatostatin receptor and I-123-MIBG scintigraphy were positive. 18F-fluorodeoxyglucose PET-CT revealed highly metabolic mass at the same location with SUV 6.9, no other distant metastatic diseases were observed. A diagnosis of paraganglioma was confirmed. Further abdominal vascular ultrasound scan suggested a 4.5 cm × 2.7 cm solid hypoechoic mass between inferior margin of the superior mesenteric artery and abdominal aorta. Left renal vein was apparently compressed, with the narrowest site around 0.13 cm, peak velocity 164 cm/s. The radius of distal left renal vein was 0.75 cm. The diagnosis of nutcracker syndrome secondary to paravertebral paraganglioma was confirmed.

Treatment

After pre-operative treatment of alpha-adrenergic blocker (phenoxybenzamine) for 4 weeks, the blood pressure fluctuated around 100-115/70-80 mmHg, heart rate 80 to 87 per-minute, distal limb circulation was massively improved. A resection of the primary tumor, including peripheral

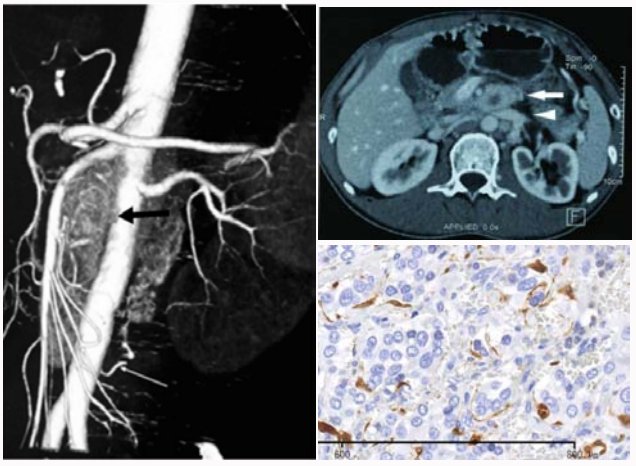

Figure:

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adherent adipose tissue and retroperitoneal lymph nodes, was done. During the operation, we observed a drastic elevation of blood pressure up to 210/140 mmHg and a rigorous decrease to 70/40 mmHg after tumor resection. Pathological analysis confirmed paraganglioma, immunohistochemical test of chromogranin A was positive and Ki-67 was 3%.

Outcome and Follow-up

The patient underwent an uncomplicated postoperative phase in intense care unit, his blood pressure gradually dropped to around 120/80 mmHg, serum NE was within normal range. Neither red blood cell in urinalysis nor postprandial epigastric pain happened after surgery. At a 3-month follow up, no blood pressure variation or symptoms recurrence were noted. Annual imaging screening as CT scan was suggested, considering the unpredictable nature of the tumor. In view of other possible hereditary syndrome involving paraganglioma, we applied a genetic test screening for multiple endocrine neoplasia type 2, von Hippel-Lindau syndrome, and Neurofibromatosis type 1, no relating gene mutation was observed. But a heterozygotic point mutation was discovered on the splicing site of SDHB gene was noted.