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Simultaneous unilateral laparoscopic adrenalectomy for pheochromocytoma and thyroidectomy in MEN 2A and MEN 2B syndrome

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A 46-year-old man with a history of left adrenalectomy performed a year earlier due to pheochromocytoma was admitted to our clinic. Subsequently, the patient was diagnosed with MEN 2A based on the results of genetic tests (genetic counselling, Medical University of Warsaw). Based on tests performed in our clinic — thyroid fine-needle aspiration (FNA) biopsy and abdominal computed tomography (CT) — he was diagnosed with malignant thyroid cancer and a tumour of the right adrenal gland. The size of the tumour of the thyroid gland was assessed by the USG of the thyroid and lymph nodes; the right lobe was 27.6 × 17.3 mm (with confluent nodules) and the left lobe was 20 × 13.8 mm. Three lymph nodes were 23 × 8.7 mm, 20 × 4.6 mm, and 10.3 × 4.6 mm in size. Histopathological examination disclosed the largest tumour up to 2.5 cm. CT of the neck revealed focal changes in the thyroid gland, visible metastatic lymph node on the right side — group III 18 mm and group IV 7 mm in size and the retropharyngeal metastatic lymph node at the level of the soft palate (10 × 7 mm in size). Eighteen lymph nodes were detected, of which 10 were metastatic lymph nodes. Preoperative USG examination revealed enlarged lymph nodes group III and IV on the right side and II and III on the left side. Abdominal CT revealed a 15 × 12 mm nodule visible in the right adrenal gland. The genetic test for *RET* gene mutation (exon 16 and codon 918) was positive. The preoperative calcitonin level was 1427.00 pg/mL Using general endotracheal anaesthesia with extended monitoring, adrenalectomy was performed transperitoneally with the patient on his side. Thyroidectomy was done afterward in the prone position. The whole procedure lasted 6 hours and 10 minutes (Fig. 1).

The patient received postoperative thyroid hormone replacement therapy. The patient had elevated serum calcitonin (42.10 pg/mL), carcinoembryonic antigen (CEA) (9.39 ng/mL), and CRP (23.47 mg/L). After 5 days the patient left the clinic in a good

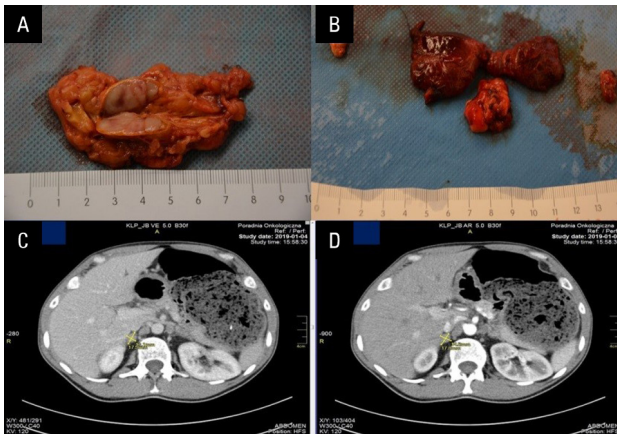

Figure 1. *Operative specimens.* **A.** *Pheochromocytoma of the right adrenal gland;* **B.** *Medullary thyroid cancer and lymph nodes;* **C, D.** *Abdominal CT scans: right adrenal pheochromocytoma*

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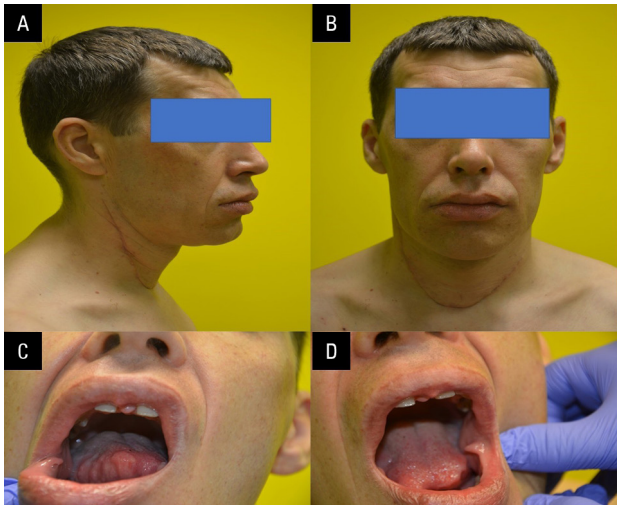

Figure 2AB. *Patient with the characteristic phenotype of MEN 2B syndrome (marfanoid habitus, bumpy lips). The patient after simultaneous right laparoscopic adrenalectomy and thyroidectomy.* **CD.** *Patient with the characteristic phenotype of MEN 2B syndrome (mucosal neurinomas and bumpy lips)*

clinical condition. The histopathological examination confirmed the MEN 2A syndrome with a diagnosis of MTC and pheochromocytoma of the right adrenal gland. The patient was followed up every 3 months postoperatively. Information about a slight metastasis of approximately 4 mm in diameter detected in the liver about one year after the surgical procedure was obtained from the patient's family. The patient died from COVID-19 complications.

A 37-year-old man was admitted to our clinic with the following symptoms: marfanoid habitus, mucosal neurinomas, and bumpy lips (Fig. 2).

Based on the FNA biopsy and abdominal CT (Fig. 3), the patient was diagnosed with malignant thyroid cancer and a tumour of the right adrenal gland.

The genetic test for *RET* gene mutation (exon 11 and codon 634) was positive. The preoperative calcitonin level was 166.00 pg/mL. USG of the thyroid revealed a tumour 10 × 6 mm and 8 × 5 mm in size in the right and left lobe, respectively. A preoperative USG revealed an enlarged lymph node in group III on the left side (12 × 8 mm in size). Group II lymph nodes — reactive changes were annotated. Group IV lymph nodes — the presence of metastasis was annotated; however, the pathologist did not provide the exact number of metastatic lymph nodes. No metastases were reported. Abdominal CT revealed a nodule in the right adrenal gland (30 × 16 mm in size). During one general endotracheal anaesthesia with extended

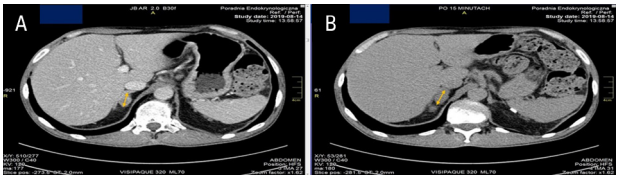

Figure 3AB. *Abdominal CT scans — right adrenal pheochromocytoma*

monitoring, adrenalectomy was performed transperitoneally with the patient arranged on the side at first. Thyroidectomy was done in the prone position afterward. The whole procedure lasted 4 hours and 55 minutes. To identify and preserve the parathyroid tissue, indocyanine green (ICG) fluorescence imaging was carried out. After visualization, parathyroid glands were exposed to near-infrared (NIR) light with a wavelength ranging from 700 to 800 nm. The camera (Quest Spectrum® NIR/ICG system) detected NIR light as a green signal. Following autofluorescence imaging, a bolus of 5 mg ICG was applied intravenously [1]. Figure 2 shows the patient after right laparoscopic adrenalectomy and thyroidectomy.

The histopathological examination of the removed tissues showed MTC — neoplastic foci were located in both lobes (maximum size of the largest foci was 2.5 cm); lymph node metastases were also found. Pheochromocytoma was diagnosed as well due to the course of MEN 2B syndrome. He received thyroid hormone replacement therapy postoperatively. The patient had elevated serum calcitonin (195.20 pg/mL) and CEA (25.74 ng/mL) levels. The patient was discharged from the clinic after 4 days and referred for further oncological treatment. The patient is still alive.

Disclosure statement
The authors have no conflicts of interest to disclose

Statements of ethics
The research was conducted in accordance with the World Medical Association Declaration of Helsinki. Data were collected retrospectively.

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Conflict of interests
None declared.

References
1. Spartalis E, Ntokos G, Georgiou K, et al. Intraoperative Indocyanine Green (ICG) Angiography for the Identification of the Parathyroid Glands: Current Evidence and Future Perspectives. In Vivo. 2020; 34(1): 23–32, doi: 10.21873/invivo.11741, indexed in Pubmed: 31882459.