MULTIFOCAL EXTRA-ADRENAL PARAGANGLIOMA – CASE REPORT

ŁUKASZ DOBOSZ, MAŁGORZATA DOBRZYCKA, PAULA FRANCZAK, JAGODA WIECZOREK, GRAŻYNA KOBERSKA-GULIDA, MAREK DOBOSZ

Department of General Surgery, Copernicus Medical Centre in Gdańsk
Kierownik: prof. dr hab. M. Dobosz

Paraganglioma is a rare neoplasm originating from extra-adrenal pheochromocytes of the sympathetic and parasympathetic nervous system. It is usually benign and the treatment method of choice is a complete resection of the tumour. The authors present a case of 66-year-old female patient with a multifocal benign retroperitoneal paraganglioma, which was completely removed during surgery.

**Key words:** paraganglioma, retroperitoneal, multifocal

Paraganglioma is a rare neoplasm originating from extra-adrenal pheochromocytes of the sympathetic and parasympathetic nervous system (1). These tumours, if originating from the cells of the sympathetic nervous system, are usually located in the area of the inferior mesenteric artery and the bifurcation of the aorta (cells of the organ of Zuckerkandl), while tumours which originate from the cells of the parasympathetic nervous system are more frequently found in the head and neck area (2). In the case of catecholamines-secreting tumours a characteristic triad of symptoms occurs including headaches, hyperhidrosis and heart palpitations, which may be accompanied by paroxysmal hypertension, pallor, nausea and vomiting (2). Lesions which do not secrete catecholamines may be asymptomatic and are often diagnosed only during autopsy and any symptoms, if present, are usually due to a mass-effect caused by the size of the tumour (1, 3). The majority of the lesions are benign and the treatment method of choice is a complete resection of the tumour (4). The authors present a rare case of a multifocal benign retroperitoneal paraganglioma, which was completely removed during surgery.

**CASE REPORT**

A 66-year-old female patient with a history of hypertension and hypothyroidism reported to the hospital for a surgical treatment of a retroperitoneal tumour. The lesion had been detected in 2012 during an ultrasound examination performed due to non-specific pain in the upper abdomen. The patient did not report any symptoms indicative of a secreting tumour such as headache, hyperhidrosis or heart palpitations. 26 years ago, the patient underwent an operation due to a retroperitoneal tumour, which was described in the ultrasound examination report as a round focus with increased echogenicity with the dimensions of 75 x 60 x 48 mm, clearly separated from the surrounding area, adjacent to the abdominal aorta. A paraganglioma was diagnosed in a histopathological examination at the time.

The patient had two CT scans. In the second scan performed in January 2014, three focal lesions were described, which were located in the retroperitoneal area. One focus was located at the antero-left-side contour of the abdominal aorta with the dimensions of 37 x 34 mm in the transverse plane and 31 mm in height, adjacent to the aorta in approximately 1/2 of the circumference. The second focus was located in the direct vicinity and to the left of the abovementioned lesion. The second focus had grown from 8 mm to 12 mm in diameter in comparison with an examination of September 2012. Another focus was located towards the medial area from the left kidney pelvis and pressed on the initial section of the ureter,
which had no signs of urinary stasis. This focus had grown from 15 to 23 mm (fig. 1, 2).

In February 2014 a fine-needle biopsy of the lesion was performed under ultrasound guidance. A paraganglioma was diagnosed in a cytology examination at the time. Serum and urine metanephrine levels were not measured in the patient due to the lack of possibility of performing such tests in the hospital.

The patient underwent open surgery in March 2014. Intraoperatively, a single soft tumour of 3-4 cm in diameter with rich blood supply, adjacent to the aortic wall was found in the area described in the CT report. It was located towards the anterior and slightly to the left of the aorta between renal vessels and the bifurcation of iliac arteries. The tumour was dissected and removed. Subsequently, after the descending colon was mobilise, the left kidney was reached and a second tumour of the size of a hazelnut was found in the area of the hilum between the renal pelvis and left renal vein and it was removed. No complications were observed during the surgery or in the postoperative period. A histopathological examination of the specimen revealed yellow and brown tumours of polycyclic contours, covered with a capsule. Microscopic analysis confirmed the presence of a paraganglioma in the samples from both specimens (fig. 3).

DISCUSSION

According to the Pick classification of 1912, neoplasms originating from pheochromocytes can be divided into those originating from the adrenal medulla, called pheochromocytomas and those originating from the cells of the sympathetic and parasympathetic ganglia, forming beyond the adrenal medulla, called paragangliomas (5). The prevalence of these neoplasms is 2-8 cases per million, of which paragangliomas account for 10-18% (6, 7). The paraganglioma originating from the cells of the sympathetic nervous system is usually located in the abdominal cavity, in the organ of Zuckerkandl, which is a collection of pheochromocytes around the bifurcation of the aorta or at the origin of the inferior mesenteric artery. The paraganglioma originating from the cells of the parasympathetic nervous system is usually located in the head and neck area; however, cases of such neoplasms being located in the urinary bladder, pelvic or chest cavity have also been reported (2, 3, 8). The paraganglioma usually occurs as a single tumour; however, in 15-24% of cases multiple foci are found (3).

It is estimated that the chance of a relapse following a complete resection of the lesion five years after the surgery is approximately 15% (9). However, in the present case new, multiple foci formed after 26 years. A local relapse after such a long time of observation is highly unlikely and these foci probably formed de novo. However, the lesions which appeared in less than a year after the surgery seem to be relapse lesions.

Catecholamines-secreting tumours account for the majority of cases and occur more frequently in the abdominal or pelvic cavity (3). Typical symptoms for these tumours (associ-
ated with excessive secretion of mainly noradrenaline, less frequently dopamine) include headaches, hyperhidrosis, paroxysmal heart palpitations, surges in blood pressure, nausea and vomiting (2, 3, 8). In the case of non-secreting paragangliomas, the most commonly reported complaints include increasingly stronger abdominal pain or symptoms associated with metastases, predominantly to the lungs, liver, bones and lymph nodes (1). Symptoms of gastrointestinal tract or ureter obstruction or deep vein thrombosis caused by the compression of these structures by a paraganglioma have also been reported (10, 11, 12). In the present case, the only symptom was non-specific pain in the upper abdominal area. No symptoms associated with excessive secretion of catecholamines were observed, which suggested a non-secreting nature of the tumour.

Secreting tumours produce, accumulate, synthesise and metabolise catecholamines therefore, their diagnosis is based on biochemical testing of the secretion and metabolites of catecholamines (2, 3, 4). It is recommended that the tests be based on the measurement of fractionated serum and/or urine metanephrine (2). Biochemical tests should be carried out mainly in symptomatic patients, patients in whom an adrenal tumour was found by chance and in patients with a hereditary risk of a pheochromocytoma or paraganglioma. The basic imaging examinations of paragangliomas allowing to detect a tumour and determine its location and size include computed tomography and magnetic resonance imaging. Both methods demonstrate similar sensitivity, while MRI is preferred for patients allergic to the contrast medium, children and pregnant women (1).

A method of choice for the treatment of paragangliomas is a complete surgical resection of the lesion. If the tumours secrete catecholamines and symptoms of their excess occur, preoperative treatment is introduced (7). In order to decrease blood pressure and prevent a hypertensive crisis during the operation, alpha-adrenergic receptor blockers, and, for patients with persistent tachycardia and/or arrhythmia, betablockers are administered for two weeks before the procedure (2, 7). At present, the laparoscopic procedure is preferred. However, for multiple tumours (as in the present case), recurring tumours or metastatic ones open resection is a more desirable method to ensure that the lesions are completely removed (13).

REFERENCES


Received: 10.05.2015 r.
Adress correspondence: 80-803 Gdańsk, ul. Nowe Ogrody 1-6
e-mail: lukaszdobosz@gumed.edu.pl