Clinics in Surgery

Case Report
Published: 27 Apr, 2017

Pheomochromoblastoma: Case of 19-Year Follow-Up Study

Tronko MD, Kvachenyuk AM*, Kovalenko AY, Bolgov MY, Tarashchenko YM, Zinych PP, Omelchuk AV and Guda BB

Department of Endocrine Surgery, State Institution "V.P. Komisarenko Institute of Endocrinology and Metabolism, Natl. Acad. Med. Sci. Ukraine

Abstract

Catecholamine-secreting tumors are one of the most complex and, at the same time, interesting problems of endocrinology. So far, the morphological criteria of malignancy of catecholamine-secreting tumors remain complex, and evidence of malignancy may manifest several years after a successful operation. It presents an interesting clinical case of a long 19-year follow-up of patients with primary inoperable pheochromoblastoma. After the radiotherapy, polychemotherapy, endovascular occlusion of tumor vessels, which subsequently reduced the signs of tumoral invasion and made possible a complete surgical removal of the tumor. The only radical method for treatment of catecholamine-secreting tumors is surgery. However, in the case of inoperable primary tumor is necessary radiotherapy and polychemotherapy.

Introduction

Catecholamine-secreting tumors are one of the most complex and, at the same time, interesting problems of endocrinology. In a population, pheochromocytomas are relatively rare, with a maximum frequency of 1:200,000 per year and a maximum incidence of one person per two million people. So far, we cannot consider a 100% *in vivo* diagnosis of adrenal chromaffin tumors. According to literature data as a whole, in 30%-70% of followed cases the diagnosis of "pheochromocytoma" is made post mortem. The five-year survival rate is 34%-60% [1,2].

Approximately 10%-15% of pheochromocytomas are malignant [3,4]. The probability of malignancy is elevated, with a significant increase in urinary dopamine excretion, a tumor size exceeding 6 cm, and signs of extra adrenal growth. So far, the morphological criteria of malignancy of catecholamine-secreting tumors remain complex and evidence of malignancy may manifest several years after a successful operation. The only absolute proof of malignancy is metastatic spreading in those anatomical areas where paraganglia are not present in normal condition [5].

Case Presentation

Patient D, born in 1968, has been admitted for the first time to the Surgery Department of the State Institution "V.P. Komisarenko Institute of Endocrinology and Metabolism of the Natl Acad Med Sci Ukraine" on April 06, 1998, complaining of arterial pressure rises, nausea, vomiting at the level of crisis, headache, dizziness, weight loss (of 10 kg), blurred vision.

History of the disease. The patient considered he ill since ≈ one month, when hypertensive crises appeared, with an increased arterial pressure up to 200/130 mmHg. The patient considered his disease to be not associated with anything. He received antihypertensive drugs.

On admission general state of the patient of moderate severity. Height: 183 cm; weight: 92 kg; BMI: 28. Uniform distribution of subcutaneous fat. Normal humidity of skin. No pathological pigmentations and trophic skin changes. No thyroid enlargement. Heart tones are muffled. Heart rate: 120 bpm; arterial pressure: 190/130 mm Hg. (s:d). Lungs: vesicular breathing. Soft painless abdomen. No liver enlargement. Kidney region painless at palpation. Pasternatsky's symptom "-" on both sides.

Results of instrumental research methods

Adrenal glands not visualized in ultrasound. Additional neoplasias in their projection not identified. Examination is extremely difficult, due to increased intestine pneumatization. In the region of liver gate, an additional neoplasia of about 50 mm cannot be excluded. Adrenal CT is recommended. ECG: sinus tachycardia, heart rate: 120 bpm. Hypertrophy and systolic overload of left ventricle. Pronounced myocardial metabolic disorders.

OPEN ACCESS

Correspondence:
Andrey Kvachenyuk, Department of Endocrine Surgery, State Institution "V.P. Komisarenko Institute of Endocrinology and Metabolism, Natl. Acad. Med. Sci. Ukraine, E-mail: kvachenyuk@yandex.ru

Received Date: *10 Feb 2017*
Accepted Date: *20 Apr 2017*
Published Date: *27 Apr 2017*

Citation:
Tronko MD, Kvachenyuk AM, Kovalenko AY, Bolgov MY, Tarashchenko YM, Zinych PP, et al. Pheomochromoblastoma: Case of 19-Year Follow-Up Study. Clin Surg. 2017; 2: 1442.

Results of laboratory research methods

17-CS: 64.0 mcmol/24 h; 17-ACS: 17.1 mcmol/24 h; epinephrine: 85.41 nmol/24 h; norepinephrine: 1060.8 nmol/24 h; dopamine: 4777.5 nmol/24 h; vanillylmandelic acid (???): 102.1 nmol/24 h. Plasma electrolytes: K: 4.44 mmol/L; Na: 133.0 mmol/L; Ca: 2.4 mmol/L. Blood glucose: 5.0 mmol/L.

Taking into account the patient's complaints, medical history data, clinical examination, and results of laboratory and instrumental (ultrasound) methods of study, the patient's diagnosis was "pheochromocytoma of right adrenal".

In preparing the operation of April 9th, 1998, on a background of a relatively favorable condition the patient developed an uncontrolled hemodynamic syndrome. Arterial pressure rose to 280/150 mmHg, being accompanied by sharp headache, nausea, vomiting, tachycardia, blurred vision. After i/m injection of 10 ml of Redzhitin, arterial pressure momentarily decreased to 110/70 mmHg, and then rose again to 260/140 mmHg. After repeated administration of Redzhitin, blood pressure decreased to 140/100 mmHg, and then rose again to 260/150 mmHg.

Given his condition, the patient was urgently taken to surgery. A right-side lumbotomy and exploration of retroperitoneal space were performed.

Protocol of operation

Right-side lumbotomy. Exploration of the upper pole of the right kidney showed a dense conglomerate that consisted of a retroperitoneal tumor, inferior vena cava, upper pole of the kidney, invading the parietal peritoneal sheet. At this stage, the size of the above tumor, the extent of tumor growth spreading cannot be evaluated. Resection of a right rib.

With great technical difficulties, the right kidney was mobilized and removed. Further exploration revealed a dense, hilly, sometimes decaying tumor, originating from the right adrenal, without clear-cut contours; it extends into the abdominal cavity, invades the lower surface of the liver, inferior vena cava on a considerable length. General tumor size: 15.0 × 10.0 × 7.0 cm. In addition, in the region of liver gate tumor's metastases are identified, measuring 2.0-3.0 cm. The tumor was recognized to be inoperable.

Pathohistological conclusion

The kidney capsule is smooth; epithelium of proximal tubules of nephrons is swollen and dystrophically changed by the type of a cloudy swelling. Their lumens sometimes contain erythrocytes.

The postoperative course was uneventful. The patient was prescribed a course of external beam radiotherapy to the area of right adrenal.

On July 1st, 1998, the patient was re-admitted to the surgery department for a planned follow-up examination. At the same time, he complained of weakness, drowsiness, lack of appetite, rare dizziness, and periodic rise in temperature up to 37.5°C-38°C. Hypertensive crises persisted with a frequency of about one per month and ceased spontaneously.

Patients underwent CT of the adrenals

CT-picture improved. The tumor in the right adrenal projection decreased in size, and infiltrate decreased as well. Liver structure was heterogeneous, possibly a secondary liver lesion. Inferior vena cava was differentiated, with no increase in size. Hormonal tests showed in 24-h urine increased levels of epinephrine, norepinephrine, dopamine, vanillylmandelic acid.

The patient had the same complaints as before Year 2000. MRI of retroperitoneal space (October 16th, 2000) in the right adrenal and kidney beds a neoplasia was determined, of irregular shape, heterogeneous structure, dimensions 28.8 × 42.7 × 49.5 mm, adjacent to the inferior vena cava. Epinephrine, norepinephrine, dopamine, vanillylmandelic acid levels remained elevated. Given the presence of neoplasia, inefficiency of previous treatment (hypertensive crises, increased catecholamine levels in 24-h urine), the patient was prescribed a course of intratumoral chemotherapy with Adriablastin. Also, a control angiography showed branches to the right adrenal to be occluded. After treatment, the patient noted an improvement in his condition, a sharp decrease in the number of hypertensive crises. At follow-up examination, CT showed no signs of progression of the underlying disease. The 24-h urine revealed several times increased levels of epinephrine, norepinephrine, vanillylmandelic acid.

In October 2001, given the tumor size, relatively non-invasive process according to MRI data, the inefficiency of previous courses of chemotherapy, the patient was operated - the tumor with the adrenal was removed for life-threatening reasons. A right-side adrenalectomy with tumor was performed.

Pathohistological conclusion

Pheochromoblastoma, trabecular variant, with areas of tumor tissue disintegration. From 2003 to 2007 control routine examinations were carried out. Arterial hypertension persisted. At control CT, MRI, there were no signs of tumor recurrence. At ultrasound, the adrenals were not visualized, no additional neoplasias were identified. In the 24-h urine, no increase in epinephrine, norepinephrine, dopamine, vanillylmandelic acid was noted. There were no symptoms of chronic adrenal insufficiency.

Since the autumn of 2007, the patient's condition has deteriorated, hypertensive crises have reappeared (arterial pressure up to 200/120). During the spiral CT with i/v contrast, no areas of pathological accumulation of contrast material were found, no evidence of continuing growth was noted. In the 24-h urine, an increase in epinephrine, norepinephrine, and dopamine was noted. The patient was prescribed Bisoprolol at 2.5 mg/24 h, Kardur at 2 mg/24 h.

From 2009 to 2016, the patient had periodical hypertensive crises (increased blood pressure from 160/100 to 200/140 mmHg). He was permanently receiving Metoprolol at 50 mg-100 mg/24 h, Zokson 2 mg/24 h. In the 24-h urine, periodically increased epinephrine, norepinephrine, dopamine, vanillylmandelic acid levels were noted.

During a spiral CT in 2016, a secondary focus in the liver and in the region of inferior vena cava was found. At the moment, the issue of re-operation is considered, namely, liver resection and prosthetics of a portion of inferior vena cava.

Clinical detailed diagnosis

Pheochromoblastoma of right adrenal $pT_4N_xM_x$. Status after right side adrenalectomy, radiotherapy, polychemotherapy, right-side nephrectomy. Secondary arterial hypertension. First degree hypertensive disease. Left kidney remains in place.

Conclusion

The only radical method for treatment of catecholamine-secreting tumors is surgery. However, in case of inoperable primary

tumor the patient was referred to a course of telegammatherapy and polychemotherapy, which subsequently reduced the signs of tumoral invasion and made possible a complete surgical removal of the tumor. A repeat tentative of surgical removal (after polychemotherapy and radiotherapy) appeared to be successful. More than 10 years after the operation, the patient has been kept under observation in the absence of a significant local recurrence of the disease, with biochemical persistence of the disease which is docked by adrenergic blockers.

The present clinical observation is interesting owing to the opportunity of a long-term combined treatment of pheochromoblastoma, including surgery, radiotherapy, polychemotherapy, endovascular occlusion of tumor vessels, with a patient's lifespan of 19 years after diagnosis.

References

1. Pacak K, Linehan WM, Elsenhofer G. Recent advances in genetics, diagnosis, localization, end treatment of phaeochromocytoma. Ann Intern Med. 2001;134:315-29.
2. Manger WM, Gifford RW. Pheochromocytoma. Springer-Verlag. New York. 1977;398 c.
3. Ajallé R, Plouin PF, Pacak K, Lehnert H. Treatment of Malignant Pheochromocytoma. Horm Metab Res. 2009;41(9):687-96.
4. Brancíková D, Mechl Z, Adam Z, Jandáková E, Pavlovský Z, Válek V, et al. Patient with inoperable pheochromocytoma. Curr Oncol. 2015;22(3):216-9.
5. Mohammed AA, El-Shentenawy AM, Sherisher MA, El-Khatib HM. Target Therapy in Metastatic Pheochromocytoma: Current Perspectives and Controversies. Oncol Rev. 2014;8(2):249.