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Case Report

Polycythaemia treated with nephrectomy

Anne Daugaard Thuesen and Gudrun Kjær Steffensen

Department of Medicine, Hospital Lillebælt, Fredericia, Denmark

Correspondence and offprint requests to: Anne Daugaard Thuesen; E-mail:annedt@dadlnet.dk

Abstract
Unilateral renal cystic disease (URCD) is a rare, non-familial non-progressive renal disorder not associated with cysts in other organs in contrast to autosomal dominant polycystic kidney disease. Only 55 cases have been published in the world literature. Renal diseases are a well-recognized etiology of secondary erythrocytosis but not in URCD. We can present at case of URCD and secondary polycythaemia. Only one case with similar history has been reported, but in our case, the polycythaemia was confirmed by measuring the erythropoietin (EPO) level in the cyst fluid.

Keywords: polycythaemia; renal cyst; secondary unilateral renal cystic disease

Background

Polycythaemia vera is a chronic myeloproliferative disease characterized with a great amount of red blood cells, leading to hyperviscosity and greater risk of thrombosis. Secondary polycythaemia is associated with other diseases such as heart diseases, lung diseases with reduced capacity, renal cell tumours, autosomal dominant polycystic kidney disease (ADPKD) and solitary renal cysts. In the two former, the reason may be an appropriate compensatory response, in the three latter, an inappropriate release of erythropoietin (EPO). Unilateral renal cystic disease (URCD) is a rare condition. Only 55 cases have been published in the world literature [1, 2] and only one of them with polycythaemia [1]. An additional case can be presented.

Case report

In 1997, a 45-year-old man Per Larsen (PL) was referred to a local hospital with polycythaemia. His haematocrit was 60% and hemoglobin 19.6 g/dL. On assessment, he was asymptomatic, with a history of smoking but no complaints from the lungs or heart. Blood samples showed normal serum creatinine 95 μmol/L, white cells 11.2×10^9/L and platelet count 251×10^9/L. A normal bone marrow examination was found, but an ultrasound (US) of the kidneys revealed a right polycystic kidney with a large number of cysts different sizes and a linear measurement of 22 cm. The left kidney and liver were normal. The diagnose polycythaemia vera could not be confirmed and the patient was discharged without further. In 2000, PL had a new US and an intravenous urography, showing the same result. In 2004, type 2 diabetes, hypercholesterolaemia and hypertension were diagnosed. An electrocardiogram with normal results. In June 2009, PL was referred to a haematology department because of the polycythaemia. There was no presence of thrombotic complications. Haematocrit was then 64%, haemoglobin was 21.7 g/dL and serum creatinine 92 μmol/L. A new bone marrow examination showed a slight hyperplasia with normal cytogenetics. A JAK-2 mutation could not be shown. An echocardiography with salt-water contrast was performed to find a possible arteriovenous shunt as well as a lung capacity examination to reveal reduced capacity but both examinations were normal. Venesections were performed three times but without effect, so PL was transferred to a nephrology department. A new US of the kidneys showed no change from 2000: a right polycystic kidney and a normal left kidney. ADPKD was excluded due to lack of family history, all other organs were spared inclusive the left kidney and there was no decline of the kidney function. A captopril renography showed that the left kidney function was 90% and the right 10%. A basic renography confirmed the apportionment 90/10%. S-EPO had been measured to 31 IU/L which is the upper level of normal range (5.0–30 IU/L). A low value would have been expected with the diagnose polycythaemia vera. An abnormal EPO production from the right polycystic kidney was suspected as the cause of the polycythaemia. A US cyst puncture with aspiration was done in order to measure the EPO level in the cyst liquid. The result was a high value of 767 IU/L, in contrast to the blood level of 31 IU/L. The high value supported our suspicion and a right nephrectomy was decided and performed without complication in November 2009 in the urology department. Histopathological examination of the removed kidney showed multiple cysts in different sizes lined by a flattened cuboidal epithelium separated by bands of normal renal parenchyma. The measurements of the kidney were $24 \times 15 \times 11$ cm with no signs of malignity. PL was seen 2 months after the nephrectomy and he felt well. His haemoglobin was, for the first time since

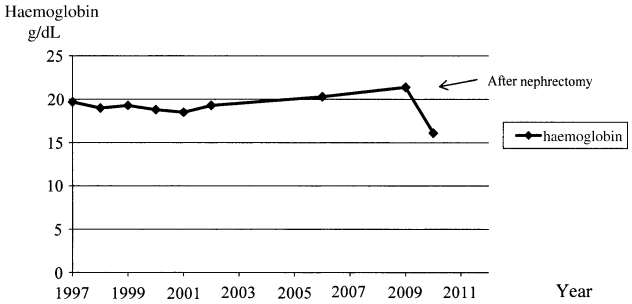

Fig. 1. Follow-up of the patient's haemoglobin levels before and after nephrectomy of the right kidney. Haemoglobin is converted from mmol/L to g/dL by the formula: × mmol/L × 1.611 = × g/dL. Normal ranges for men are 8.0–11.0 mmol/L = 12.8–17.7 g/dL.

1997, in the normal range, 16.6 g/dL, and the haematocrit 49%. Six month later, it was still normal 16.1 g/dL (Figure 1) and the EPO level was reduced to normal range 17 IU/L.

Discussion

URCD has been known as a distinct disease entity since 1987 [3] and different from ADPKD, renal neoplasm and simple cysts. The clinical importance of URCD is to make a differential diagnosis [4–6]. The histological finding of URCD is not different from those of ADPKD. Unlike ADPKD, there is no family history, normal kidney function and it only involves one kidney from just a part to most or even the whole kidney and other intra-abdominal organs are spared [2]. Our patient fills these criteria. Long-term follow-up is recommended with or without screening of family members to exclude asynchronous ADPKD and cystic neoplasm [5, 6]. Our patient was followed for 12 years and he had no family history. The diagnosis of secondary polycythaemia was confirmed by measuring the high EPO level in the cyst fluid. The concentrations of bioactive EPO in the cyst fluid are found to be from undetectable values up to 3200 IU/L (7). Measuring EPO levels in the blood and cyst fluid can be helpful to make the right diagnosis. Blake-Jones *et al.* [1] did not measure the EPO concentrations in the first and only reported case of URCD with polycythaemia. The treatment in this case was nephrectomy. In most of the cases of polycythaemia described in the literature in patients with simple cysts, the polycythaemia disappeared after surgical removal, which suggests that there is a causal relationship between the renal lesion and the haematologic disorder [8, 9]. The primary source of EPO synthesis is the interstitial fibroblasts in the kidney regulated by tissue oxygen tension, and renal diseases is a well-recognized etiology of secondary erythrocytosis [10]. It is not clear how renal cysts cause EPO production. There are experiments that suggest that the high EPO production is correlated with localized ischaemia through compressive or vasoconstrictive mechanisms [1]. Other studies point to the stroma cells of the cyst walls, which produce EPO independent of oxygen pressure inside the cysts [7]. However, measurements of levels of EPO in the blood and cyst fluid seem to be a helpful tool in diagnostic and treatment of URCD with polycythaemia.

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Conflict of interest statement. On behalf of all authors (A.D.T. and G.K.S.) there is no conflict of interest to declare.

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