Polycystic Kidney Disease Associated with Polycystic Ovarian Syndrome

M.		Segasothy
M.Y.		Norazlina
P.H.		Ong
M.		Jamil

Departments of Medicine, Radiology and Obstetrics and Gynecology, University Kebangsaan Malaysia, Kuala Lumpur, Malaysia

M. Segasothy, Department of Medicine, Faculty of Medicine, University Kebangsaan Malaysia, Jalan Raja Muda Abdul Aziz, 50300 Kuala Lumpur (Malaysia)

Dear Sir,

Autosomal dominant polycystic kidney disease (ADPKD) is commonly associated with other systemic manifestations such as cardiac valvular lesions, intracranial aneurysms, hepatic cysts and diverticula [1]. Rarely, ovarian cysts have been associated with ADPKD [1]. Polycystic Ovarian Syndrome (PCOS) has, however, not been documented in patients with ADPKD. We report a patient with PCOS and ADPKD.

A 30-year-old female presented to the Obstetrics and Gynecology Department with hirsutism of 2 years' duration and amenorrhea and weight gain of 6 months' duration. On examination, she was found to be overweight (weight 65 kg, height 160 cm) with gross hirsutism. There was male distribution of body hair. The breasts and genitalia were normal. There were no masses palpable on abdominal examination. Pelvic examination revealed normal findings. Blood pressure was 140/90 mm Hg. Cardiovascular examination revealed no murmurs or abnormal heart sounds. There was no neurologic deficit.

Investigations revealed elevated serum testosterone (5.5 nmol/L; normal range: 0.9-2.8), raised luteinizing hormone/follicle-stimulating hormone ratio 10.7:5.2 (normal range: < 2) and low serum estradiol (216 pmol/L; normal range: 588-1,397 pmol/L). Serum prolactin (190 mU/L; normal range: 117-68), dehydroepiandrosterone (3.5 µmol/L; normal range: 1.1-10.7) and dexamethasone suppression test were normal. Vaginal probe ultrasound demonstrated the classic 'necklace'-like arrangement of cysts in both ovaries (fig. 1). The adnexa were normal.

Ultrasonography and computed tomography of the abdomen revealed multiple cysts of

The patient was treated with oral cyproterone acetate and premarin for her PCOS and was referred to the nephrology unit for evaluation and management of her ADPKD. By this time, the patient developed hypertension, the blood pressure being 190/110 mm Hg, and symptoms of renal colic. Investiga-

08*02.81 M4 15.OZ.41

t F8.ü D 18.♦HH 21.2HH

JAB. OBSTETRIK & GINEKOLOGI, UKM IJEBU NEES 21761

Pot, CYSTIC OVARY, ,

m»m i *

g.iXl,f*. 3/8781.
5/81113/85% %00
me.4 o

G 78 DBS45 EE *

β-εε M-Mc»RK . B-RE VERKE

Fig. 1. Polycystic ovary. Transvaginal sonography shows multiple peripheral cysts in a normal-sized left ovary.

©1992 S.KargerAG, Basel

0028-2766/92/

0624-0482\$2.75/0

Fig. 2. Adult polycystic kidney disease. Enhanced CT scan shows multiple bilateral renal cysts of varying sizes.

tions showed hemoglobin 13.3 g/dl, urea 4.5 mmol/l, creatinine 81 μmol/l and uric acid 305 μmol/l. Urinalysis showed traces of protein, leukocytes $30 \times 10^6/l$, red cells $50 \times 10^6/l$, epithelial cells $3 \times 10^6/l$, no casts and staphylococcus 100,000 colony-forming units/ml on culture. She was treated with a course of antibiotics for her urinary tract infection and commenced on prazosin for her hypertension.

This patient had clinical features of PCOS such as hirsutism, weight gain and amenor-rhea. The diagnosis was confirmed by hormonal assays and ultrasonography. She also had features of ADPKD such as hypertension, renal colic, microscopic hematuria and urinary tract infections. Ultrasonography and computed tomography confirmed the presence of bilateral polycystic kidneys with multiple calculi in the right kidney.

Extrarenal involvement of ADPKD includes the cardiovascular, gastrointestinal, neurological, musculoskeletal and the genitourinary system. Cardiovascular abnormalities include mitral valve prolapse, aortic valve incompetence, tricuspid valve prolapse [2], bicuspid aortic valve, aortic root dilatation, aortic aneurysms, coarctation of the aorta, dissecting thoracic aortic aneurysms [3] and left ventricular hypertrophy [2]. Gastro-intestinal abnormalities include hepatic cysts [4] and colonic diverticula [5]. Neurological abnormalities include intracranial aneurysms [6]. Musculoskeletal abnormalities include inguinal and umbilical hernias [7] and other genitourinary abnormalities include ovarian cysts [1]. The association of ovarian cysts and ADPKD has not been well studied and PCOS has not so far been reported to occur with ADPKD. Our patient may well be the first case to have PCOS in association with ADPKD.

In view of the involvement of the cardiovascular, gastrointestinal and genitourinary systems, it has been suggested that ADPKD should be viewed as a systemic disease [1]. Our findings of genitourinary as well as endocrine abnormalities in a patient with ADPKD lend support to the above view.

References

Gabow PA: Autosomal dominant polycystic kidney disease – more than a renal disease. *Am J Kidney Dis* 1990; 16:403-413. Hossack KF, Leddy CL, Johnson AM, Schrier RW, Gabow PA: Echocardiographic findings in autosomal dominant polycystic kidney disease. *N Engl J Med* 1988; 319:907-912. Torres VE, Holley KE, Offord KP: General features of autosomal dominant polycystic kidney disease; in Grantham JJ, Gardner KD (eds): *Problems in Diagnosis and Management of Polycystic Disease*. Polycystic Kidney Res Found, Kansas City, 1985, pp 49-67. Dalgaard OZ: Bilateral polycystic disease of the kidneys: A follow-up of two hundred and

eightyfour patients and their families. *Acta Med Scand Suppl* 1957;323:326-329. Scheff RT, Zuckerman G, Harter H, Delmez J, Koehler R: Diverticular disease in patients with chronic renal failure due to polycystic kidney disease. *Ann Intern Med* 1980;92:202-204. Bigelow NH: The association of polycystic kidneys with intracranial aneurysms and other related disorders. *Am J Med Sci* 1953;225:485-494.

Sedman A, Bell PA, Manco-Johnson M, Schrier R, Warady BA, Heard EO, Butler-Simon N, Gabow P: Autosomal dominant polycystic kidney disease in childhood: A longitudinal study. *Kidney Int* 1987;31:1000-1005.