Normal black kidney

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Abstract

A black kidney has 3 major differential diagnoses: hemosiderosis, lipofuscin pigment and melanotic renal cell carcinoma. Excluding lipofuscin, the other 2 are accompanied by an abnormal renal function. We report on a 25-year-old man who intended to donate a kidney to his cousin. On the operating room table when we incised the left flank region and exposed the kidney, we found a firm and black kidney so the operation was cancelled due to potential vascular injuries. Days after the incomplete procedure, we reviewed the donor's biochemistry and imaging to reassess his renal function, but the results showed quite normal renal function again. The result of Ham test was also negative. Two weeks later, we began the operation, removed the same left kidney and found that it was in the same conditions as it was before. We took the opportunity to send needle biopsies of the kidney for histopathologic analysis. The analysis showed a melanotic kidney without pathological changes in glomeruli and interstitium and vessels. A black kidney may result in hemosiderin, lipofuscin or melanin deposits in the kidney, which can confirm the diagnosis; however, special tests for underlying disease and renal function should be considered. Some causes of black kidney lead to abnormal function, but our patients's kidney returned to normal.

Introduction

Kidney transplantation is preferred for patients with end-stage renal disease (ESRD) or chronic kidney disease (CKD).[1,2] Organ sources are limited to live and cadaveric human donors. The first kidney transplant in Iran was in 1967; in 2002, Iran was ranked fifth in kidney transplants.[2] Mashhad University of Medical Sciences, in north east part of Iran, started performing kidney transplants in 1988. So far, as one of the pioneering universities in the region, it has recorded about 3000 kidney transplants and has become the transplant centre for our neighbouring states.

Black kidney has 3 major differential diagnosis: hemosiderin deposits,[3] lipofuscin pigment[4] and melanotic renal cell carcinoma (RCC); all cases are very rare.[5] The differential diagnosis for melanin in cancer cell usually leads to the diagnosis of melanoma, melanotic neurofibroma, melanotic dermatofibrosarcoma protuberans, pheochromocytoma, basal cell carcinoma, seborrhetic keratosis or breast cancer.[6]

Hemosiderosis is a form of iron overload disorder resulting in the accumulation of hemosiderin in different organs. This disorder is a disease, like sickle cell anemia and thalassemia, in which chronic blood loss requires frequent blood transfusions (though beta minor thalassemia has been associated with hemosiderin deposits in the liver in patients with non-alcoholic fatty liver disease independent of any transfusions).[7,8] Also, renal hemosiderosis is a complication of chronic intravascular hemolytic states, such as hemolytic anemia, paroxysmal nocturnal hemoglobinuria (PNH) and mechanical hemolysis after inserting a prosthetic cardiac valve[9,10] or black-water fever as well.[3] Renal hemosiderosis (blue kidney) is the anatomic indicator of severe intravascular hemolysis.[3]

The exact pathogenesis of renal failure in blue kidney is unknown, but the iron chemical activity in hemosiderin may cause tubular damage and, eventually, cell death.[11] The role of hemosiderin in acute renal toxicity remains controversial.[12] Calazans and colleagues reported on a 37-year-old woman with renal hemosiderosis and sickle cell anemia, which caused renal failure.[13] In another case, a 68-year-old non-diabetic male, with a history of metastatic colon cancer, was evaluated for a rising serum creatinine level after clinical evaluations and imaging; in this case, the renal hemosiderosis was considered the cause of the renal failure.[9]

Lipofuscinoses occur due to abnormal accumulation of this pigment.[14,15] Pathologic lipofuscin can lead to blue kidney,[4] macular degeneration and other diseases.[16-19] In melanotic RCC, dark-brown endogenous pigments can be found; the kidney may appear black one, and the differential diagnosis may be hemosiderin, homogentisic acid and lipofuscin.[5] Black or blue kidney is difficult to diagnose without advanced paraclinical studies.

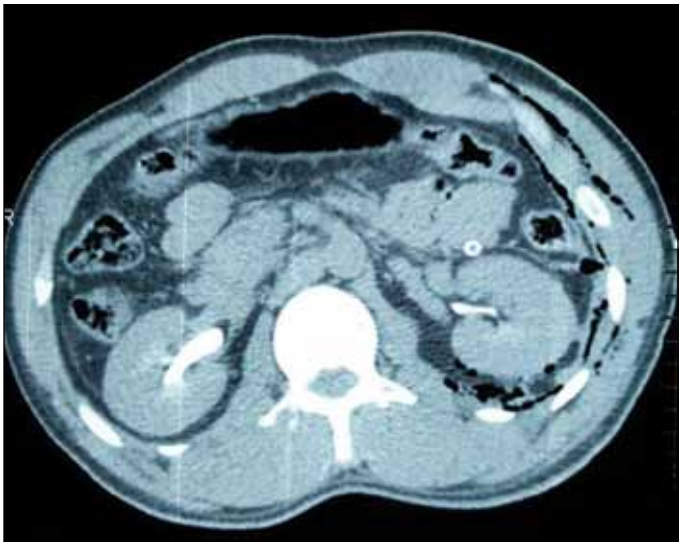

Fig. 1. Normal renal computed tomography angiography was done before operation.

We report the case of a 25-year-old male kidney donor who was referred to our centre for an unusually coloured kidney with normal function, but with a diagnosis of melanotic kidney. Written informed consent was obtained from the patients (both donor and recipient) for publication.

Case presentation

A 25 year old man, who wanted to donate a kidney to his cousin, was referred to our organ transplantation centre (Montaserieh Educational Hospital). On October 2012, we arranged a full biochemistry, imaging (Fig. 1), psychological and histocompatibility tests, followed by physical examinations. On the operating room table when we incised the left flank region and exposed the kidney, we found a firm and black kidney (Fig. 2). After consulting with the transplant team, we stopped the operation for further assessment. Days after the incomplete procedure, we reviewed the donor's biochemistry (blood urea nitrogen, creatinine, 24-hour urine collection) and imaging to reassess his renal function, but the results were normal again (Fig. 3, parts A and B). The result of Ham test (to rule out PNH) was also negative. Two weeks later we repeated the operation and removed the same kidney. We found the left kidney in the same condition it was before (black, firm with good dieresis) and we sent needle biopsies of the kidney for histopathologic analysis. The analysis showed a melanotic kidney without pathological changes in glomeruli and interstitium and vessels. Based on pathology report, glomeruli are normal and there were coarse, tubular intracytoplasmic granules (Fig, 4, part A). These brown granules were positive for Fontana (black) (Fig. 4, part B) and negative for modified Ziehl-Neelsen[20,21] for lipofuscin (Fig. 4, part C). They were also was negative for other specific staining (repeat periodic acid Schiff [PAS], Pearl and trichrome). These findings were associated with heavy renal melanin pigmentation. One week after the second operation, the donor and recipient were checked by blood urea nitrogen and creatinine; both values were normal postoperation and the patients were discharged.

Discussion

The normal size of a kidney is 10 to 12 cm in length, 5 to 7 cm in width and 2 to 3 cm in thickness.[22,23] Its normal colour is reddish-brown due to the number of small capillaries.[24] Our patient had a black kidney. After the renal vascular and renal function tests, we ruled out acute vascular disorders. After the transplant, the pathology reports showed

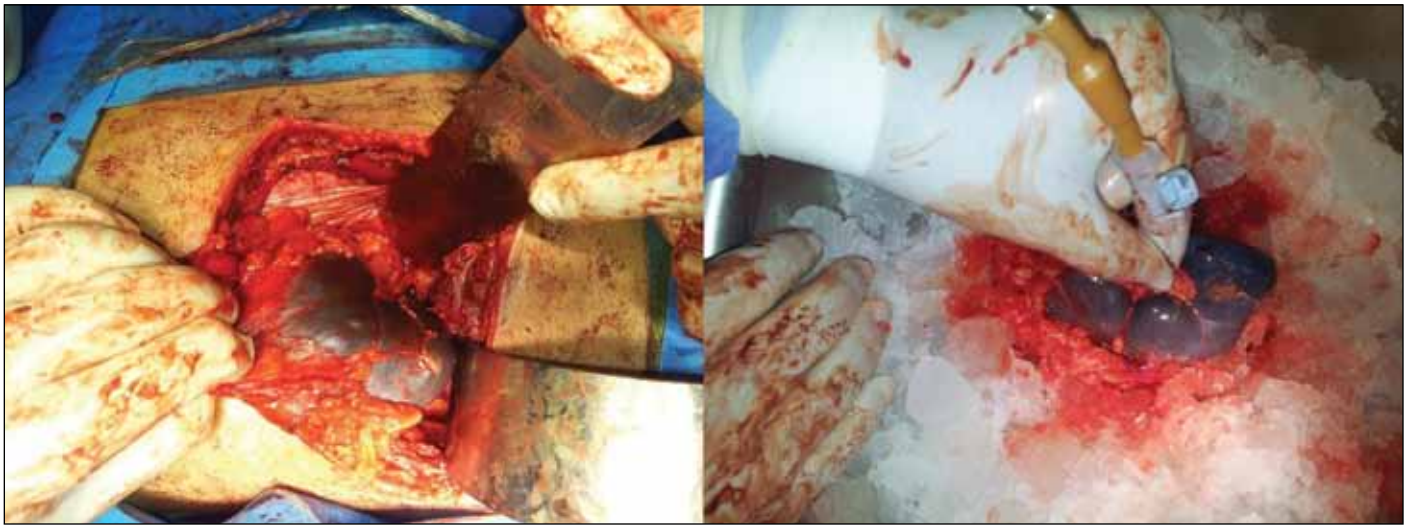

Fig. 2. A black renal with normal arterial pulse and palpable appeared.

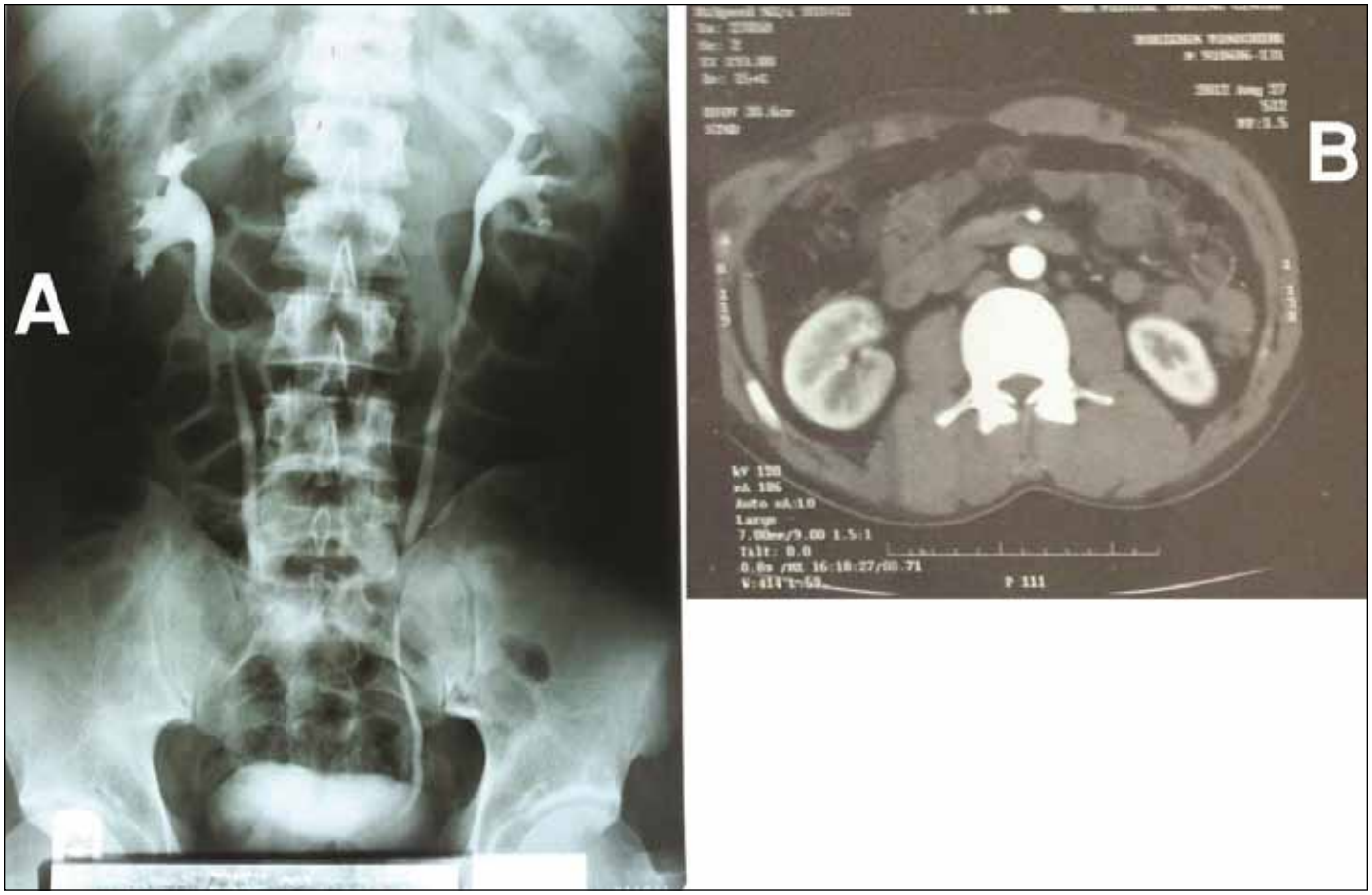

Fig. 3. A: Normal in vitro produced secretion and function, B: A computed tomography scan with intravenous contrast with normal results.

black-brown pigment deposits in the patient's renal cells.

One of the differential diagnoses of black pigment in renal cells is renal hemosiderosis. Renal hemosiderosis is a rare cause of renal failure that can occur in diseases characterized by chronic intravascular hemolysis and lead to a black kidney. Free hemoglobin released in sickle cell anemia is filtered by the glomeruli and reabsorbed by proximal convoluted tubules, leading to renal hemosiderosis.[13]

Siddappa and colleagues presented a rare case of renal hemosiderosis appearing with renal failure due to repeated blood transfusion (about 2 per month) over past 2 years for chronic refractory anemia.[25] Three cases of paroxysmal nocturnal hemoglobinuria with renal hemosiderosis and renal failure were reported by Nair and colleagues.[10] Most patients with renal hemosiderosis also have renal failure.

Although the pathogenesis of renal failure in a blue kidney is unknown, iron depositing in glomeruli and interstitium is a trigger of renal malfunction. Additional tests, like sucrose hemolysis test and Ham's test for acid hemolysis, were recommended by Siddappa and colleagues.[25] These tests were negative in our patient.

In 1987, Mocelin and colleagues reported on a 29-year-old living donor with a blue kidney. At the 6-month follow-up, renal function of the donor and recipient were normal, but pigmentation in this case was lipofuscin;[4] lipofuscin pigmentation has been called the aging pigment or phenacetin analgesic nephropathy.[26,27] In 1964, Biava and colleagues reported on 114 patients with lipofuscin-like granules in renal vascular smooth muscle cells. Patient age ranged from 2 to 70 years. The authors found that the number of granules, such as lipofuscin, in different patients correlated with age and hypertension.[28]

In 2001, Lei and colleagues reported an unusual case of melanin-pigmented clear cell RCC with melanocytic differentiation.[29] In their case, upon gross examination, they found a solid yellow and tan tumour measuring 8 × 7 × 6 cm occupying the upper pole of the kidney. Upon light microscopic evaluation, they found abundant black pigment.[29] Moreover, many cases were melanocytic differentiation, which led to RCC.[5]

Conclusion

Black or blue kidney may be caused by vascular or non-vascular factors. Non-vascular black pigments in light microscopic examination may result in hemosiderin, lipofuscin or

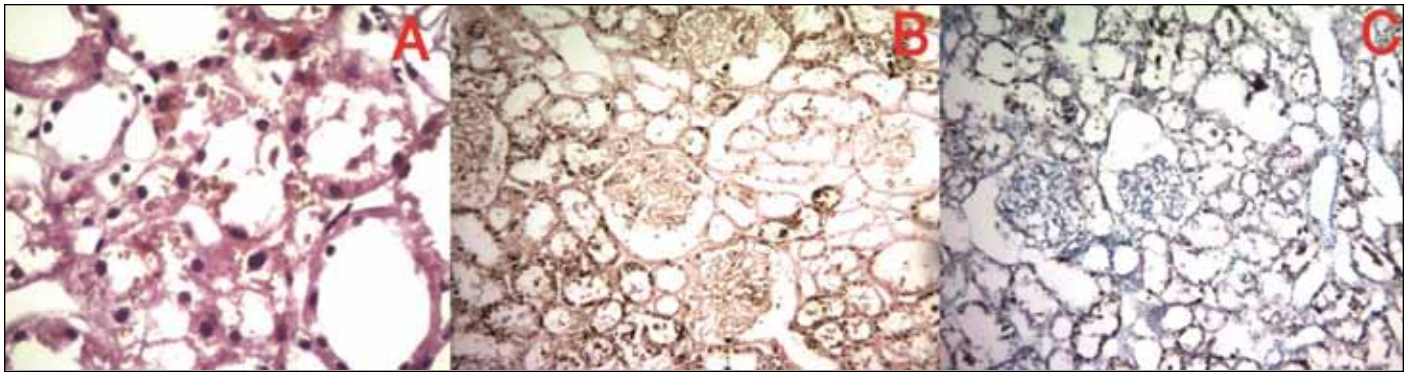

Fig. 4. A: Coarse tubular intracytoplasmic brown granules, glomeruli are completely spared (hematoxylin and eosin stain 400×); B: Coarse Fontana positive cytoplasmic granules (100×); C: Intratubular granules negative in modified Ziehl Neelsen for lipofuscin (100×).

melanin deposits in the kidney. A histopathological study can confirm the diagnosis; moreover, special tests for underlying disease and renal function should be considered.

Competing interests: Dr. Yarmohamadi, Dr. Rezayat, Dr. Memar and Dr. Rahimi all declare no competing financial or personal interests.

This paper has been peer-reviewed.

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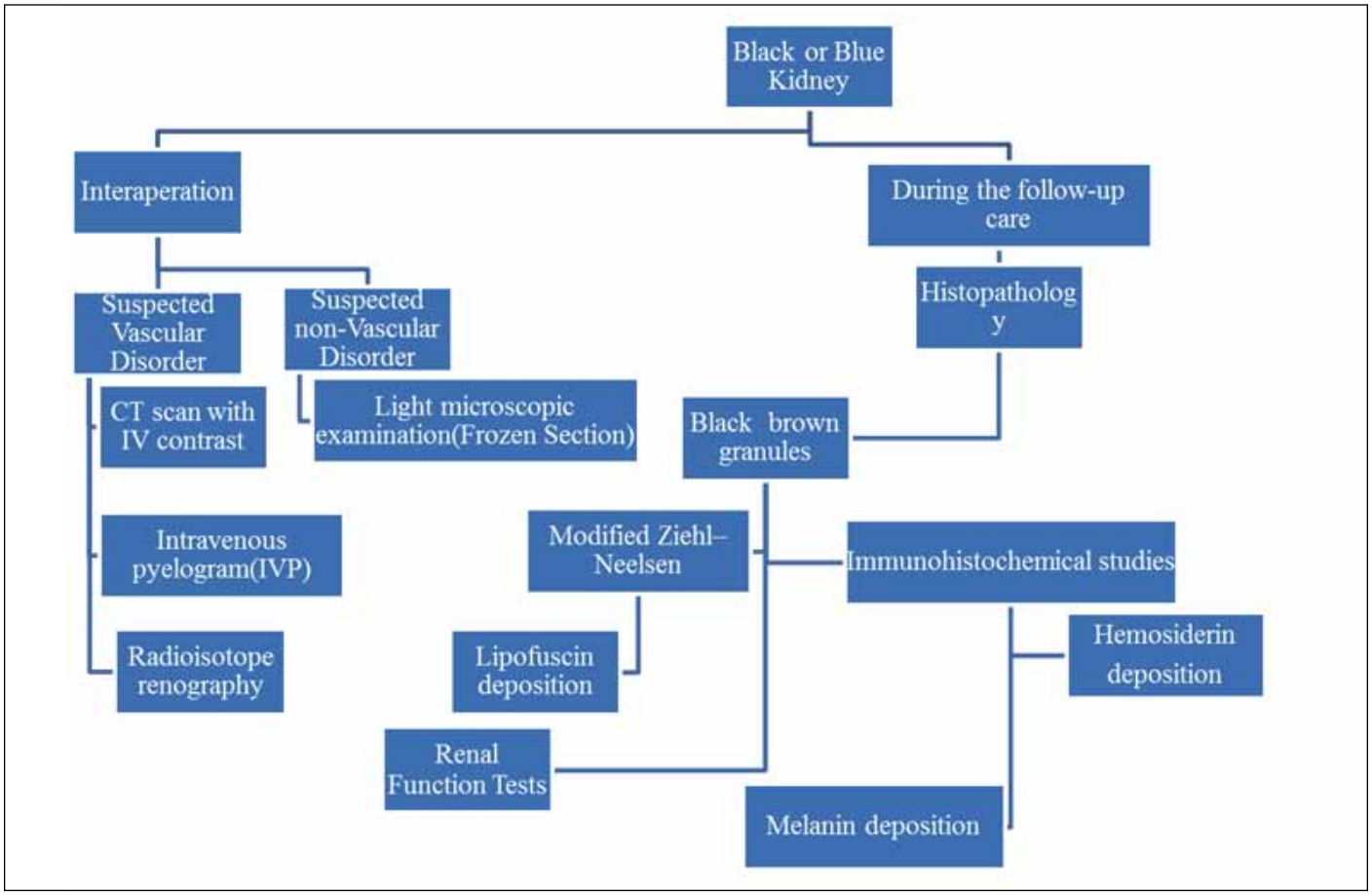

Fig. 5. Black or blue kidney diagnosis fellow-chart.

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