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CASE REPORT

Intractable bleeding from the renal pelvis in a patient with hereditary haemorrhagic telangiectasia (Rendu-Osler-Weber disease)

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Case report

The patient was a 68-year-old female with a confirmed genetic diagnosis of HHT (mutation in the ACVRL1 gene) with a strong family history of the disease. She had typical manifestations of telangiectasias in the skin, nasal septum (with recurrent epistaxis) and the gastrointestinal tract. There were known arteriovenous malformations in the liver and in the left renal pelvis. In addition, she had chronic anaemia and pulmonary hypertension, disorders often associated with HHT. The patient also had metastatic breast cancer treated with hormone therapy and her nutritional status was poor. Long term anticoagulant therapy was given due to a deep venous thrombosis of the upper extremity.

She was admitted to the emergency department due to left-sided flank pain and gross haematuria with clot retention. There were no physiological signs of circulatory instability and initial treatment was manual evacuation of blood clots from the urinary bladder and continuous bladder irrigation.

CT-scan demonstrated the left renal pelvis obstructed by blood clots with secondary parenchymal hypoperfusion. There were signs of multiple arteriovenous malformations with an enlarged, draining vein in relation to the renal pelvis (Figure 1). The patient developed clinical signs of systemic infection and was therefore treated with intravenous antibiotics and a percutaneous nephrostomy tube.

The bleeding did not subside and required blood transfusions and frequent manual irrigations of the nephrostomy tube and bladder catheter. The former eventually occluded and the patient was taken to the operating theatre for placement of an internal ureteric stent instead of the percutaneous drainage. Gross haematuria continued over the next days with continuous bladder irrigation and repeated blood transfusions. An attempt to obtain endovascular haemostasis was performed and multiple pathologic vascular malformations were embolized through right femoral access (Figure 2). Angiography demonstrated reduced feeding of the venous sinuses and no obvious extravasation to the urinary collecting system (Figure 3).

The results of the endovascular treatment were observed over the following days, but the bleeding persisted and required repeated blood transfusions. After nine days of attempts at conservative and minimal invasive approaches to the condition, it was decided to perform laparoscopic nephrectomy with the patient's consent. The operating time was 119 min with an estimated blood loss of 100 ml. The postoperative course was without complications and there were no

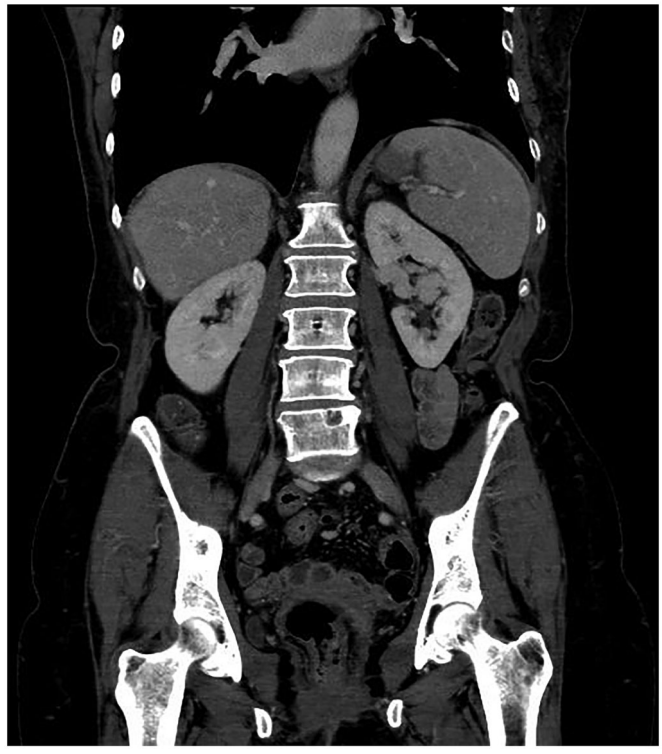

Figure 1. Coronal reconstruction of contrast enhanced CT scan in nephrographic phase showing on the left side an enlarged, draining vein in the central part of kidney hilum.

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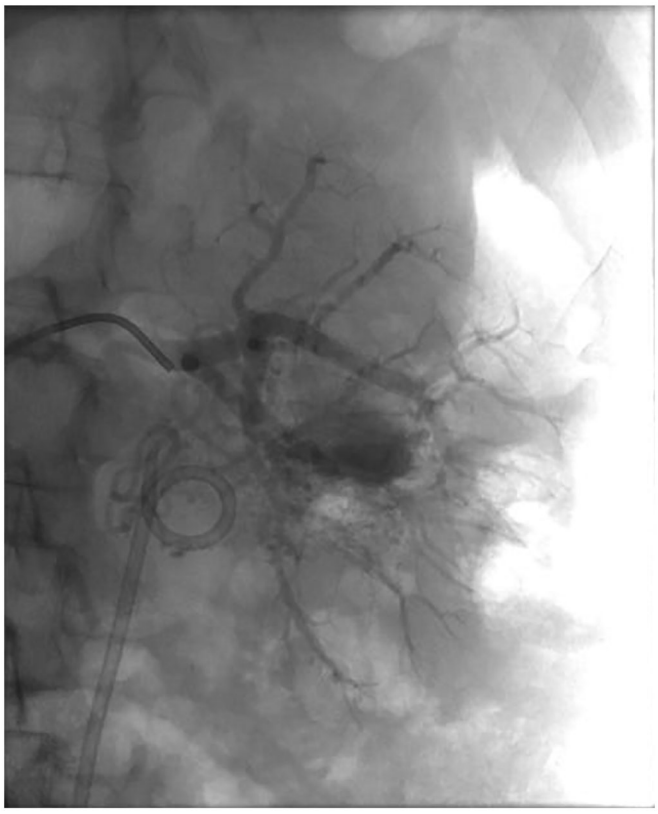

Figure 2. Renal angiogram showing arteriovenous malformation with pathologic vessels and early filling of large draining veins.

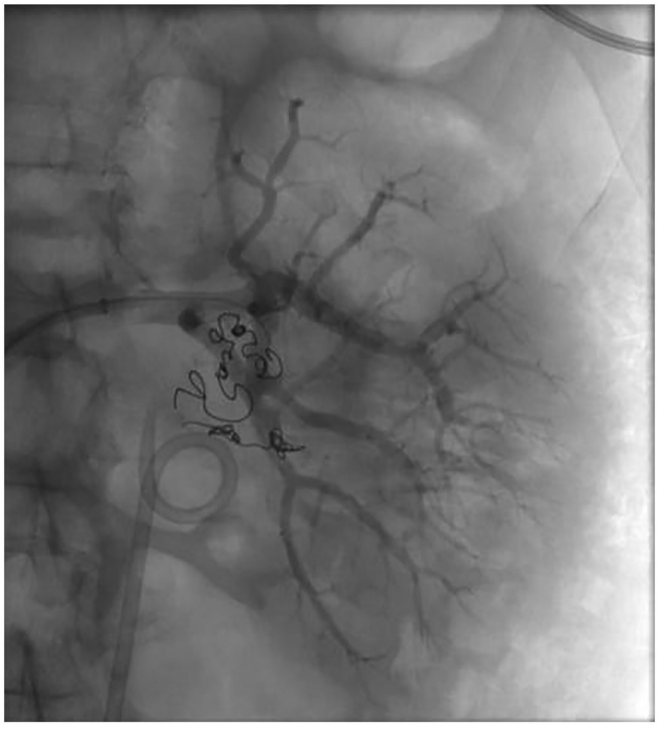

Figure 3. Following coilembolization of multiple arterial feeders reduced early filling of draining veins. No contrast filling of the urinary collecting system seen.

further need for blood transfusions. The serum creatinine value increased from 45 μmol/l (with an estimated GFR of 99 ml/min/1.73 m^2) to 75 μmol/l (with an estimated GFR of 71 ml/min/1.73 m^2). The patient was discharged from the hospital five days postoperatively.

Gross examination of the kidney specimen showed hemorrhage with clot formation in the renal pelvis (Figure 4(A)). Histologic evaluation revealed hemorrhage and an increased number of malformed vessels in the pelvic area characteristic of arteriovenous malformations (Figure 4(B)). The vascular malformation consisted of an admixture of arteries, veins/venules and capillaries. An area with rupture of a dilated vein into the renal pelvis was identified (Figure 4(C)). The vessels showed abnormal dilatation and abrupt changes in thickness of vessel wall (Figure 4(D–G)). There were also areas with increased numbers of capillary-like vessels (Figure 4(H–I)).

Discussion

Hereditary haemorrhagic telangiectasia is an autosomal dominant disease that affects 1 in 5–8000 and leads to the development of abnormal vascular structures such as telangiectasias and arteriovenous malformations, in which the latter most commonly affects pulmonary, hepatic and cerebral circulations [1]. Affection of the renal pelvis is an exceptionally rare manifestation and has only been described in two previous case reports requiring embolization [2] and surgery [3], respectively. One cohort study including 11 patients with HHT, where renal structure and function were assessed, found no evidence supporting renal involvement in this disease [4]. Renal arteriovenous malformations are rarely seen unless it is after trauma or iatrogenic injury, in which case it would be defined as an arteriovenous fistula. Further research would be needed to define the role of renal arteriovenous malformations in the context of HHT. This case reports a female patient with an advanced stage of HHT, including systemic comorbidities, where the intractable bleeding from the renal pelvis demanded a sequential approach, gradually increasing the invasiveness, before ultimately being successfully treated with laparoscopic surgery.

Due to severe comorbidities and a poor nutritional status, the patient was initially deemed less eligible for major surgery and the minimally invasive approaches were perceived as more suitable treatment options. However, as the condition did not respond adequately, the decision to advance to major surgery was unavoidable. The patient tolerated the treatment well. This highlights the importance of a multidisciplinary approach to such a patient, involving close cooperation with interventional radiology services.

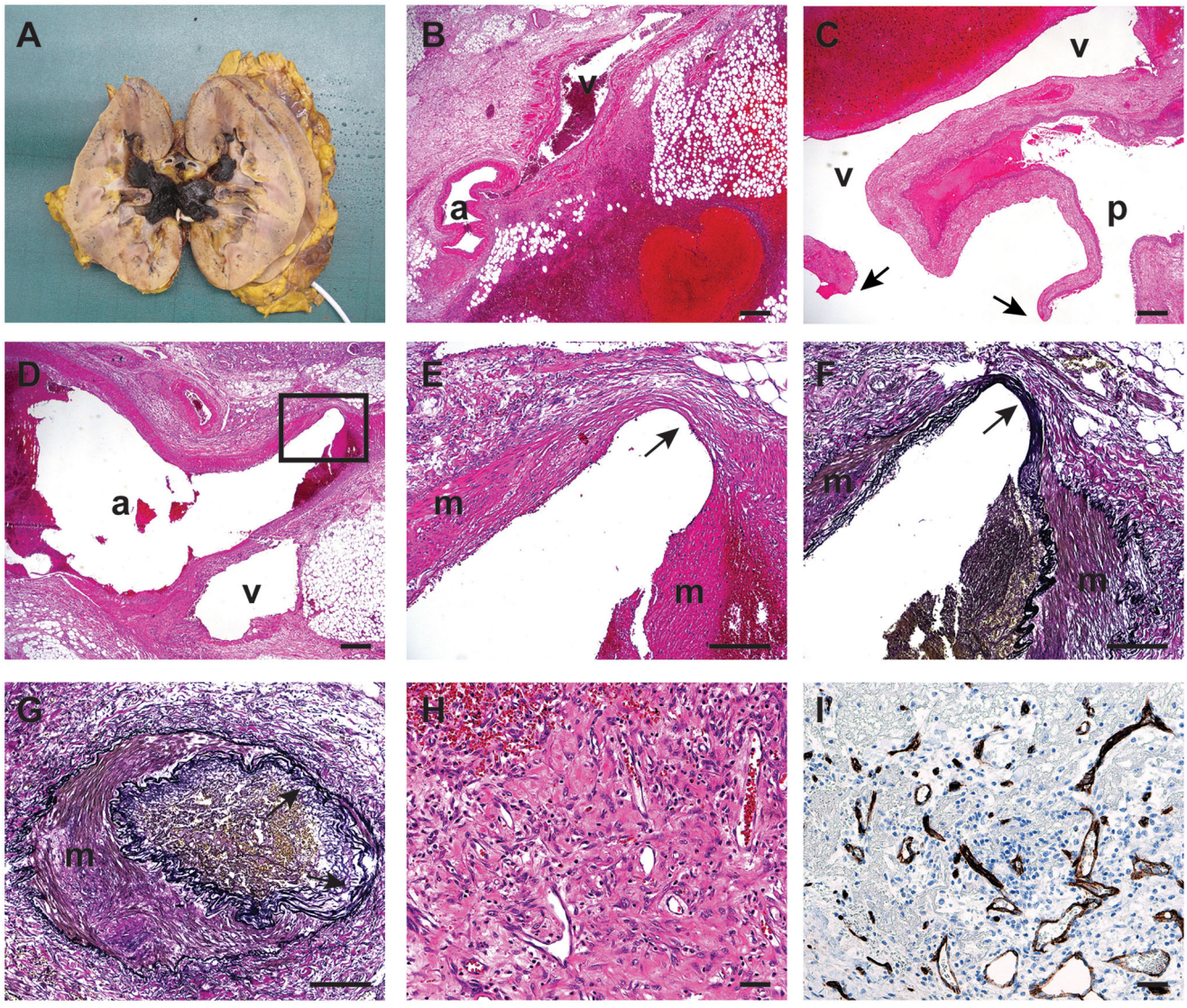

Figure 4. (A) Gross examination of the resected kidney shows blood clots in the renal pelvis. (B–E) H&E (Hematoxylin and Eosin)-stained sections from the renal pelvis with arteriovenous malformation. B: Section from the renal pelvis with hemorrhage, dilated artery (a) and vein (v). (C) A dilated vein (v) with rupture into the pelvis (p). Arrows demarcate point of rupture. (D) Dilated artery (art) and vein (v). (E) Higher magnification of box in D shows abrupt change in vessel wall thickness. Arrow points at section with thin wall lacking tunica media (m). (F) Elastic stain highlights the inner and outer elastic lamina. (G) Elastic stain of another artery showing variation in the thickness of tunica media (m). Arrows at segment of artery with thin wall. (H) Area with increased numbers of capillary-like vessels. (I) Immunostaining for the endothelial marker CD34 from the same area as in H, highlights the capillary-like vessels. (B–D) Original magnification ×20, scale bar 400 μm. (E–G) Original magnification ×100, scale bar 200 μm. (H–I) Original magnification ×200, scale bar 200 μm.

Disclosure statement

No potential conflict of interest was reported by the author(s).

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