A pelvic kidney occurs in between 1 in 2200 and 1 in 3000 people, due to failure of ascent during development. It is commonly asymptomatic and usually functions normally. Pelvic ureteral junction obstruction can either be congenital or acquired, and is characterized by intrinsic stenosis or extrinsic compression of the ureter at the junction with the pelves–lyceal renal system. This can cause symptomatic or asymptomatic hydronephrosis. We describe the complex case and management of a patient with a massive pelvic ureteral junction obstruction in a pelvic kidney.

CASE PRESENTATION

A 45-year-old female presented with abdominal pain and recurrent urinary tract infections, associated with acute exacerbations of her neurological and respiratory sarcoidosis.

A CT scan showed a right pelvic kidney with a massively enlarged pelvis secondary to pelvic ureteral junction obstruction (PUJO) (Fig. 1). A percutaneous ultrasound-guided nephrostomy was placed for initial management, following which a dimercapto succinic acid scan demonstrated 32% function on the right kidney. Creatinine was 70 µmol/L with an estimated glomerular filtration rate (eGFR) of 84 mL/min/1.73 m².

The presence of large uterine fibroids measuring up to 16 × 13 cm indicated a possible extrinsic cause of PUJO. The patient subsequently had an open myomectomy, and trial of clamping of the nephrostomy tube. However, a
postoperative CT scan revealed persistence of PUJO. After a further symptomatic episode of pyelonephritis and an eGFR drop to 59 mL/min/1.73m², she proceeded with a reconstructive pyeloplasty.

A prepyeloplasty CT angiogram revealed the complex vascular anatomy of the pelvic kidney with 3 renal arteries arising respectively from the aorta, left common iliac, and right internal iliac artery (Fig. 2). An open dismembered pyeloplasty of the pelvic kidney was performed (Fig. 3). Recovery was uneventful. At last follow up, eGFR was 60 mL/min/1.73m² and MAG3 renogram showed well perfused kidneys with no evidence of obstruction.