A Rare Benign Renal Tumour Presenting as Polycythaemia in a Teenage Girl

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S Geoghegan, BJ McGuire, P Geurts, C Shaw, A Fabre, JM Fitzpatrick, KJ O'Malley
Department of Urology, Mater Misericordiae University Hospital, Eccles St, Dublin 7

Abstract
We present the case of a 15 year old girl who presented with polycythemia. CT abdomen revealed an enhancing mass in the upper pole of her left kidney with features suggestive of renal cell carcinoma. She underwent a laparoscopic radical nephrectomy. Histology demonstrated a well circumscribed, focally encapsulated, round blue cell tumour showing areas of microcystications and numerous psammoma bodies. Immunostaining showed diffuse positive staining for CD 57. This was consistent with a diagnosis of metanephric adenoma a rare benign epithelial renal tumour.

Case Report
A fifteen year old girl was urgently referred to a haematologist by her General practitioner with polycythaemia. She was taking regular Isoretinoin (Roacutane) for acne vulgaris and under went routine blood testing. Her Haemoglobin (Hb) was 18.6 g/dl and serum Erythropoetin (EPO) was 24.5 mIU/ml (normal 4-27 mIU/ml). A 2 mm mutation for Polycythemia Rubra Vinea was negative. Computed Tomography (CT) of her abdomen revealed a 2.7 x 3 cm enhancing mass in the upper pole of her left kidney which was suspicious for a renal cell carcinoma (Figure 1). A follow up triphasic renal CT study was performed and there was no evidence of renal vein or inferior vena cava invasion. No extrarenal extension, no lymphadenopathy and no remote metastatic disease identified. A laparoscopic radical nephrectomy was performed.

Sectioning through the nephrectomy specimen revealed a tan homogenous solid tumour, 2.9 cm in max dimension within the upper pole (Figure 2a). It was well circumscribed, focally encapsulated, round blue cell tumour showing areas of microcystications and numerous psammoma bodies (Figure 2b). The cells showed little cytology, oval round nuclei and inconspicuous nucleolus. There was very little mitotic activity and no necrosis. Immunostaining showed diffuse positive staining for Vimentin, CD 57 (Figure 2c) and focal staining for CD56, CD99 and CK 7. These findings were consistent with metanephric adenoma. She recovered well and was discharged with a Hb of 14 g/dl.

Discussion
On histology, diagnostic differential of this rare benign entity include papillary renal cell carcinoma and Wilms tumor. Metanephric adenoma (MA) is a rare benign epithelial renal tumour and the mean age of presentation is 40yrs, range (5-83yrs). There is a female predominance of 2:1. MA is generally an incidental finding however presentations include flank pain, haematuria, a palpable abdominal mass and occasionally polycythaemia. The incidence of polycythaemia in the setting of MA is 12%, and it has a tendency to present in this way in those under thirty-five years. Radiologically, MA can be difficult to distinguish from renal cell carcinoma. Ultrasonographic appearances are that of a well circumscribed mass lesion which can be hyperechoic, isoechoic or hypoechoic and tends to be hypo or avascular on doppler. The tumour has higher glomulization than the renal parenchyma on non-contrast CT, and has lower attenuation than surrounding parenchyma after contrast injection.

On histology, differential diagnoses of this rare benign entity include papillary renal cell carcinoma and Wilms tumor. Both these tumours are malignant, and may show similar histological features to MA. Pre operative or intra-operative diagnosis is difficult. There have been reports that there is sufficient cytological and cytogenetic characteristics to diagnose metanephric adenoma on fine needle aspiration cytology, this appears difficult and may require ancillary techniques including immunohistochemistry. The mechanism of EPO secretion is not yet understood. It is hypothesised that normal renal cells secrete EPO in response to hypoxia induced by the tumour. However EPO may also be produced by the tumour cells directly. The in vitro production of EPO in a MA cell line has been demonstrated. This is the first described case of MA presenting as polycythaemia in a girl of this age. It is a benign tumour, which shares histological and radiological characteristics with both renal cell carcinoma and Wilms tumour, both of these tumours may present with polycythaemia. High pre-operative diagnosis would avoid radical nephrectomy, the rarity of this tumour and its similar histological characteristics to two more common malignant tumours make definitive pre-operative or intra-operative diagnosis difficult.

Correspondence: K O'Malley Department of Urology, Mater Misericordiae University Hospital, Eccles St, Dublin 7 Tel: +353 1 803 4444 Fax: +353 1 803 2404 Email: k.o'malley@luh.com

References