A rare erythropoietin secreting adrenal adenoma

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Adrenal adenomas, carcinomas and phaeochromocytomas are commonly listed as tumours that secrete erythropoietin. A literature search identified numerous cases of polycythaemia associated with adrenal adenomas secreting cortisol and testosterone. A few case reports also suggested additional erythropoietin secretion in such adenomas. We failed to identify a single case solely secreting erythropoietin.

Case History

41 year old female presented to the haematologists with the coincidental finding of polycythaemia at investigation of dyspepsia: haemoglobin 198 g/L, white blood cell count 8 x 10⁹/L, platelets 236 x 10⁹/L, haematocrit 0.57 L/L and red blood cell count 6.16 x 10¹²/L. Six years earlier haemoglobin had been 151 g/L, white blood cell count 7.7 x 10⁹/L, platelets 297 x 10⁹/L, haematocrit 0.4 L/L and red blood cell count 4.53 x 10¹²/L. She was a non-smoker taking no medications. There was no history of cardiovascular or respiratory disease.

Investigations: Exon 12 of the JAK2 gene and Exon 9 of the CALR gene analyses were normal. Erythropoietin 15 mU/ml (5-25) was inappropriately normal for the prevailing haemoglobin level.

Ultrasound of kidneys confirmed them to be normal but a 6.7 cm rounded mass with internal calcification was present between the liver and the right kidney (Figure 1).

CT imaging pre-contrast confirmed the lesion to be a 5.8 x 5.7 x 5.2 cm adrenal mass with a focus of calcification. Post-contrast there was peripheral heterogeneous enhancement with a large central area of necrosis (fig 2). Fat planes with adjacent organs were well maintained.

Post uncomplicated adrenalectomy, there was a fall in erythropoietin, haemoglobin and red blood cell count initially to sub-normal levels with rapid normalisation which has persisted (figure 3).

Microscopic examination of the adrenal lesion showed extensive central infarction focal ossification. The residual lesion subcapsularly was composed predominantly of spindly cells having limited cytoplasm and a population of small sized cells showed granular positivity for chromogranin, synaptophysin, Melan A, inhibin, pankeratin, S100 protein, GFAP, CD10 and HHV-8. Occasionally cells expressed CD56. A few small sized cells showed granular positivity for Melan A scattered amongst the cells containing hyaline globules.

Based on these findings the lesion was thought to be an unusual adrenocortical adenoma.

Conclusion

We describe a case of an adrenal adenoma causing severe polycythaemia by sole secretion of erythropoietin. We believe this to be the first such description.