Obstructive uropathy due to extramedullary haematopoiesis in beta thalassaemia/haemoglobin E

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Summary: An 18 year old woman with beta thalassaemia/haemoglobin E developed a large pelvic tumour resulting in bilateral obstructive uropathy. Technetium-99m sulphur colloid marrow image, computed tomographic scan of the abdomen and needle biopsy of the mass confirmed the diagnosis of extramedullary haematopoiesis. Although radiation is the treatment of choice for decompression, the mass in this patient did not respond satisfactorily due to its multiple area of tumour autoinfarction. Obstructive uropathy due to extramedullary erythropoiesis has not to our knowledge been previously described.

Introduction

Extramedullary haematopoiesis, a common manifestation of severe thalassaemia, occurs as a consequence of uninhibited erythropoiesis. Common sites of involvement include the posterior mediastinum, liver and spleen. Generally these extramedullary haematopoietic masses are asymptomatic, although spinal cord compression has been frequently cited. A patient with beta thalassaemia/haemoglobin E in whom an unusually large pelvic extramedullary haematopoietic mass resulted in severe bilateral obstructive uropathy prompts this report.

Case report

An 18 year old woman with the diagnosis of beta thalassaemia/haemoglobin E since the age of 10

References

years presented with a slowly progressive enlarged pelvic mass of 2 months duration. She had received 6–10 units of packed red cells annually and the haemoglobin level ranged between 5 and 7 g/dl.

On physical examination the patient was pale and icteric; secondary sexual characteristics were scarcely developed. A large firm smooth mass, size 17 × 18 cm, extending from the suprapubic bones to the umbilicus was noted. The liver was 4 cm below the right costal margin and the spleen was 10 cm below the left rib cage. An initial complete blood count revealed a haemoglobin of 5.8 g/dl, a white blood count of 6.6 × 10⁹/l and there were 12 nucleated RBC/100 WBC in the differential count. Red blood cells were markedly hypochromic with prominent anisopoikilocytosis. Blood chemistries revealed a blood area nitrogen of 17 mg/dl and a serum creatinine level of 70 μmol/l.

Computerized tomography (CT) of the abdomen demonstrated two large retroperitoneal soft tissue masses, each 10 cm in diameter, extending from just below the renal hila down to the pelvis (Figure 1a). Multiple hypodense areas were observed inside the masses. Both pelvicalyceal systems and proximal ureters were markedly dilated and the intestine was displaced forward and laterally (Figure 1B). A direct contiguity between the masses and the lumbosacral spine was also observed. Technetium-99m (Tc-99m) sulphur colloid marrow image revealed a large lobulated area of abnormal accumulation of radiotracer at the pelvic region with a large infarction area at the right lateral portion of the mass (Figure 2).

Stenting catheters were placed into both ureters and radiation was given to the masses at a dosage of 200 rads per day for 10 days. Repeated bone marrow images showed a slight decrease in tumour size with a significant decrease in uptake of radiotracer. Six weeks after radiation, the mass recurred at its original size and the patient was rehospitalized for surgical intervention. However, a generalized convulsion developed after 4 units of packed red cell transfusions and the patient died from aspiration pneumonia. An autopsy was not performed. Needle biopsy of the pelvic mass demonstrated active marrow tissue consistent with a diagnosis of extramedullary erythropoiesis.

Discussion

Pelvic extramedullary erythropoiesis in patients with thalassaemia has been rarely described; a review disclosed only three case reports, all with thalassaemia intermedia.4–6 In two of these cases,4,5 the masses exerted a pressure effect mainly on the rectum while it was asymptomatic in the other case.6 We have not been aware of any previous description of extramedullary erythropoiesis caus-
ing obstructive uropathy including reports of pelvic extramedullary erythropoiesis among the non-thalassaemic disorders.7-9

The cortical irregularity and coarsening of the trabecular pattern of the lumbosacral spines as shown in the CT scan suggested that the extramedullary erythropoiesis in our patient represented extrusion of proliferating marrow through the cortex rather than embolization of circulating totipotential cells as was proposed by Ask-Upmark.3 In the previous study of the CT features of presacral extramedullary erythropoiesis masses in thalassaemias, irregularity and erosion of the anterior cortex of the sacrum were also observed.4,6

In order to decompress the erythropoietic mass, radiation is often the treatment of choice10 since the mass is highly radiosensitive and surgical interven-
tion may result in massive haemorrhage due to the high vasculature of the tumour.11 Issaragrisil et al. reported a prompt response of the mass with spinal cord compression to deep X-ray therapy at a dose of 2,000–3,000 rad.12 The erythropoietic mass in our patient however did not respond satisfactorily to radiation. The relatively huge mass and hypoxic autoinfarction, as demonstrated by Tc-99m sulphur colloid marrow image, could be responsible for this radiotolerant effect,13 and early surgical decompression should be considered in such cases.

Acknowledgement

The authors wish to thank Professor Henry Wilde for his review of the manuscript. Dr Ti is a recipient of China Medical Board Scholar Development Fund No. 85-441.

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Malignant phaeochromocytoma and hypercalcaemia

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Summary: We describe a case of hypercalcaemia secondary to recurrent malignant phaeochromocytoma. Parathyroid-related protein (PTHrp 1–86) immunoreactivity was identified in plasma and PTHrp was identified by immunocytochemistry in tumour tissue.

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Accepted: 23 July 1992
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Postgrad Med J 1993 69: 75-77
doi: 10.1136/pgmj.69.807.75

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