A patient with polycythaemia vera associated with membranoproliferative glomerulonephritis

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Introduction

Myeloproliferative disorders (MPDs) are clonal disorders of the haematopoietic stem cells. These are classified as polycythaemia vera (PV), essential thrombocythaemia (ET), chronic myeloid leukaemia (CML) and myelofibrosis (MF) depending on the main haematopoietic lineage involved. Glomerulonephritis (GN) refers to a pattern of glomerular injury based on characteristic histopathology findings. GN associated with MPDs are rare in clinical practice. Although cases of polycythaemia with concomitant GN have occasionally been reported, there are few reports regarding PV. Amongst the few reported cases, focal segmental glomerulosclerosis (FSGS) is one of the most frequent forms that has been described. As far as we know PV associated with membranoproliferative glomerulonephritis (MPGN) has not been described before.

Case report

A 43-year old woman was admitted to hospital with a history of red eyes, painful red swollen hands and palpable erythematous rash over the limbs of three weeks duration. She also complained of headache and impaired vision. She was found to have high blood pressure one week prior to admission. On examination she was plethoric with conjunctival suffusion. Her hands were red and puffy. There was a palpable purpuric rash distributed predominantly in the extremities. Blood pressure was 110/90 mmHg. There was a 2 cm firm splenomegaly. Examination of the fundi revealed hyperaemic discs and tortuous vessels.

Investigations revealed haemoglobin of 22.5 g/dL, haematocrit of 70.7% red blood cell count of 8.06x10¹²/L, total leukocyte count of 12.4x10⁹/L and platelets of 430x10⁹/L. Blood picture showed elevation of all three cell lines. Urine analysis showed proteinuria of 3g/L without any dysmorphic red cells. There was a 2 cm firm spleenomegaly. Examination of the fundi revealed hyperaemic discs and tortuous vessels.

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Serum LDH was elevated. Serum erythropoietin was 4.6 IU/L (normal 3.7-29.5 IU/L). Vasculitic screening and ANA were negative. Serum complement levels were normal. Hepatitis B, C and HIV screening were negative. JAK2V617F and JAK2Exon12 mutations were absent. Bone marrow aspiration and trephine biopsy showed a markedly hypercellular marrow with erythroid predominance.

The diagnosis of (PV) was made as our patient fulfilled all three major criteria of polycythaemia vera study group (PVSG) diagnostic criteria. Due to constraints in assessing absolute red cell mass, indirect indices that reflect the red cell mass such as haemoglobin and haematocrit were considered. Percutaneous renal biopsy was performed to investigate proteinuria further and its findings confirmed MPGN.

Patient was subjected to several venesections once the diagnosis of PV was made and a target haematocrit of less than 45% was achieved during the hospital stay. She was started on aspirin and folic acid. Once the MPGN was diagnosed oral prednisolone was started 1mg/kg body weight/day. She was given nifedipine and captopril to keep the target blood pressure less than 120/70mmHg. Osteoporosis prophylaxis was started with alendronate. Currently she is on tapering doses of prednisolone and other medication. 24 hour urine protein excretion is checked every third month and the current value after six months of therapy is 0.357 g.

Discussion

MPGN may be primary or secondary. Majority of MPGNs are primary. The commonly identified secondary causes are autoimmune diseases, chronic viral, bacterial, protozoal infections, carcinoma of lung and breast, paraprotinaemias and chronic and recovered microangiopathies which includes polycythaemia [1]. Detection of MPGN is important as fifty percent of patients with MPGN develop end stage disease ten years after diagnosis, and ninety percent have renal insufficiency after twenty years. Early initiation of immunosuppressive treatment is worthwhile though there is only some evidence regarding steroids in MPGN [2].
Polycythaemia may be primary which is known as PV or secondary. PV results from clonal expansion of a transformed haematopoietic stem cell. This is associated with a prominent overproduction of erythrocytes and to a lesser extent, expansion of the granulocytic and megakaryocytic elements. PV is usually associated with genetic mutations such as JAK2V617F and JAK2Exon12. There are case reports of PV associated with glomerulonephritis such as FSGS, membranous GN and IgA nephropathy [3-7]. But there are no reported cases of PV associated with MPGN to our knowledge.

Some case reports on PV and GNs show improvement of cell counts and proteinuria after repeated venesections alone. The improvement of cell counts in our patient with repeated venesections was not sustainable and there was no improvement of proteinuria. However once she was commenced on oral prednisolone, she showed a sustained improvement in cell counts with dramatic improvement in her proteinuria. She did not require any more venesections thereafter. Long-term follow-up of this patient will be required to show whether the remission of proteinuria indicates resolution of the renal changes and a good prognosis.

References

An unusual cause for shock in dengue fever

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Introduction

Dengue shock syndrome carries poor prognosis. Shock in dengue may not always be due to shock syndrome. Spontaneous splenic rupture can mimic shock syndrome, which is rare but potentially fatal. We report a case of spontaneous splenic rupture in a dengue patient treated by emergency splenectomy, which was curative.

Case report

An otherwise healthy 30-year old man was admitted with six days fever and malaise. He was haemodynamically stable on admission and capillary refilling was less than 2 seconds. His platelet count was 99 × 10⁶/ ml and packed cell volume was 43%. On the following day he experienced abdominal discomfort and distension. Few hours later patient collapsed and systolic blood pressure was 80 mmHg.

His haemoglobin dropped from 15.6 g/dl to 6 g/dl and he went into refractory shock despite resuscitation with blood and volume expanders in the intensive care setting. Further, he developed features of intra-abdominal hypertension with anuria and low cardiac output. Ultrasoundography showed gross ascites and hepatomegaly. Needle paracentesis showed frank blood in the peritoneal cavity. Resuscitative laparotomy was decided upon.

There was about 3 liters of blood in the peritoneal cavity on laparotomy. Splenic capsule was found to have torn exposing the raw oozing surface. Splenectomy was performed and the patient recovered and went home after 14 days of hospital stay. Pathological examination of the

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