Hyperviscosity Syndrome

Hyperviscosity refers to any state in which there is increased viscosity of the blood. Increased serum viscosity usually results from increased circulating serum immunoglobulins (eg, macroglobulinaemia, multiple myeloma) and can also result from increased cellular blood components (eg, red or white blood cells) in hyperproliferative states - eg, leukaemias, polycythaemia and thrombocythaemia.

Epidemiology

- The precise incidence of hyperviscosity syndrome is not known, as it may occur in a large number of conditions.
- Hyperviscosity may occur at any age but the aetiology of that seen in infants is different from that seen in adults.

Conditions in which hyperviscosity may occur

Hyperviscosity occurs as a result of a raised haematocrit or due to increased levels of circulating plasma components. Many conditions may produce this state, including:

- Waldenström's macroglobulinaemia (most common cause).
- Multiple myeloma.
- Polycythaemia rubra vera.
- Leukaemia (both acute and chronic).
- Connective tissue disorders - eg, rheumatoid arthritis.
- Retinoic acid therapy.
- Cryoglobulinaemia.
- Chronic hypoxia.
- Paraneoplastic syndromes (hyperviscosity results from the large amounts of circulating immunoglobulins, cryoglobulins, paraproteins or antibodies, or due to an excessive increase in blood cells).

In infants, hyperviscosity may occur as a result of the polycythaemia which develops in response to intrauterine hypoxia or hypoxia during delivery.

Presentation

Increased viscosity and reduced blood flow may result in a variety of clinical manifestations, including:

- Central nervous system: lethargy, headache, nystagmus, deafness, convulsions.
- Visual: papilloedema, fundal haemorrhages, dilation of the retinal vessels, loss of vision.
- Cardiovascular system: hypertension, heart failure.
- Haematological: dilutional anaemia, abnormal bleeding (eg, bruising, mucosal bleeds, rectal bleeding, menorrhagia), thrombosis, leukocyte dysfunction (sepsis), crossmatch difficulties.
- Renal: renal failure, proximal renal tubular acidosis.

Investigations

- Plasma viscosity - increased.
- FBC and differential cell count.
- Blood film may show rouleaux formation.
- Platelet count.
- Clotting screen.
Other investigations to determine the underlying cause include bone marrow aspiration, urine electrophoresis, auto-antibody levels.

Management

Non-drug

- Patients with a hyperviscosity syndrome should be advised that this may recur; they should be advised to look for signs of bleeding or infection.
- Some conditions producing hyperviscosity may be helped by regular venesection - eg, polycythaemia rubra vera.
- Unfortunately, repeated procedures may lead to iron deficiency, resulting in microcytic erythrocytes, which induce higher viscosity than normocytic erythrocytes. This may increase the risk for veno-occlusive events. [5]
- Infants may be treated using partial exchange transfusion.
- In adult patients, plasmapheresis to remove excess numbers of cells or circulating complexes remains the treatment of choice. [6] 1-2 procedures are advised for the treatment of hyperviscosity syndrome in Waldenström’s macroglobulinaemia. In patients who are drug-resistant this may be indicated as long-term management. [1]

Drugs[1]
The underlying cause of the hyperviscosity syndrome may be treated with chemotherapy where appropriate. Rituximab is a commonly used agent.

Complications

Complications may occur as a result of bleeding, thrombosis or sepsis and may result in neurological deficit, heart failure and renal failure.

Prognosis

The overall prognosis for any patient will depend on the underlying condition and severity of any complications of hyperviscosity.

Further reading & references

- Ehrly A; Therapeutic Hemorheology, 2012.

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