Answers

1. (d) (HIT)

HIT or heparin induced thrombocytopenia, is divided into type I and II. Type I consists of a mild non-immune thrombocytopenia, often seen within a few days of starting heparin. This is typically transient, requires no intervention and improves despite continued heparin administration. Type II, in contrast, is an immune-mediated, idiosyncratic reaction with potentially severe consequences. Severe thrombocytopenia with haemorrhage, purpura and a platelet count of less than 20, is uncommon, with the complications coming mainly from the associated thrombotic tendency. Both arterial and venous thromboses occur. The thromboses may be seen even with a ‘normal’, albeit falling platelet count. Onset is usually 5–14 days after starting heparin, but can occur earlier if there has been previous heparin exposure. HIT may also occur with low molecular weight heparin, albeit more rarely. Treatment is to stop the heparin and give an alternative anticoagulant.

PNH: paroxysmal nocturnal haemoglobinuria. Chronic haemolysis punctuated with acute episodes, major cause of morbidity arises from venous thrombosis and the progressive pancytopenia.

TTP: thrombotic thrombocytopenic purpura. Thrombotic tendency is most marked in brain and kidney. It may present with headache, confusion, focal signs and fits.

HUS: haemolytic uraemic syndrome. Thrombocytopenia, microangiopathic haemolytic anaemia and renal failure; 90% secondary to E. Coli 0157 infection; patients may have headache and confusion.

FURTHER READING

2. Intracranial venous thrombosis secondary to Behçets disease.

FURTHER READING

3. Combined immunodeficiency syndrome. His neurology is due to subacute combined degeneration of the spinal cord secondary to a gut enteropathy, in turn secondary to low plasma IgA. The recurrent chest infections are due to the low IgG, with associated bronchiectasis.

FURTHER READING

4. Myotonic dystrophy. Percussion of the muscle being recorded helps elicit this repetitive firing of individual muscle fibres. These discharges wax and wane in frequency (20–150Hz) giving a characteristic Doppler-like sound over the loudspeaker.

FURTHER READING

5. (a) and (b). PMP22 point mutations and deletions cause hereditary neuropathy with liability to pressure palsies (HNPP). The characteristic neurophysiology findings reveal conduction delay which involves atypical locations, for example delay of ulnar conduction across the wrist or median nerve conduction along the forearm. Susceptible areas such as the carpal tunnel or around the fibular head may also reveal demyelinating features but this does not necessarily indicate HNPP.

FURTHER READING