Lupus cofactor phenomenon

I read with interest the recent paper by Mathey et al about a case of familial antiphospholipid syndrome. The authors stated that the lupus anticoagulant could not be confirmed in the father, although the APTT did not correct with normal plasma. The results showed that the addition of normal plasma further prolonged the APTT by a few seconds, making it seven seconds prolonged. This is an example of the lupus cofactor phenomenon.

Although the exact nature of this cofactor is unknown, it cannot exert its effects unless the lupus anticoagulant is present. This is indirect confirmation that the lupus anticoagulant is present in this patient. The fact that the dilute Russell’s viper venom time (DRVVT) was equivocal does not change the conclusion as a recent study has shown that the DRVVT will not detect all lupus anticoagulants. Perhaps a further confirmatory test would have been useful for this patient—a thromboplastin inhibition test or platelet neutralisation procedure.

Dr Mackie et al comment:

We stated that the APTT was performed as a screening test, using control plasma, patient plasma, and a 50/50 mixture, and that the presence of a lupus anticoagulant was confirmed by a more specific technique. The APTT alone is generally not suitable for determining the presence or absence of lupus anticoagulant because even if a sensitive reagent is used, it is not specific, and may be influenced by factor deficiency, increased concentrations of coagulation factors, as well as by various inhibitors, including: antiphospholipids, antibodies against coagulation factors, and heparin.

We used the DRVVT as a confirmatory test with a platelet neutralisation procedure, using freeze-thaw lysed washed normal platelets. Tissue thromboplastin inhibition tests are less sensitive and give false negative results in many patients, especially those with IgM lupus anticoagulant. Most recent comparisons of lupus anticoagulant tests have found that the DRVVT and kaolin clotting times are the most sensitive and reliable, although no single test has a 100% detection rate. Unfortunately, it is not always possible to perform more than one of these tests.

In the family described the APTT did not correct in the father, but APTT tests are notoriously erroneous, and this result did not fulfill our criteria for the presence of lupus anticoagulant. As the father was asymptomatic, there was no justification for further studies at this stage, and the question of whether he had a lupus anticoagulant remains academic. On the basis of an abnormal, though equivocal DRVVT result, and positive antiphospholipid antibodies, with his family history it is very likely that future samples would give unequivocally positive lupus anticoagulant tests, and development of suitable clinical criteria would classify him as a true antiphospholipid syndrome patient.
Lupus cofactor phenomenon.

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