Overtight nappy precipitating thrombosis in antithrombin III deficiency

R Soutar, V Marzinotto, M Andrew

Abstract

An antithrombin III deficient infant presented with iliac vein thrombosis, apparently precipitated by an overtight nappy. Venous thrombosis is unusual, both in normal and in antithrombin III deficient children, but children with venous thrombosis should have their natural anticoagulants assayed and obvious risk factors avoided.

(*Arch Dis Child* 1993; 69: 599)

Heterozygote antithrombin III deficiency is an autosomal dominant abnormality predisposing to venous thromboembolism.¹ Altogether 50% of affected individuals will develop a venous thrombosis during their lifetime,² but this is very uncommon in the first decade of life.

We report an unusual case of antithrombin III deficiency presenting in a neonate who developed femoral and iliac vein thrombosis, probably secondary to an overtight nappy.

Case report

A healthy girl was born at term by vaginal delivery. There were no obstetric or neonatal difficulties. She presented two weeks after birth with a cold, swollen, blue left leg. There was an obvious circumferential line of bruising around the mid thigh. Non-accidental injury was initially suspected. Venography, however, demonstrated occlusion of the left external iliac and femoral veins (figure). Ultrasonography of the pelvis was normal. Platelet count, prothrombin time, and activated partial thromboplastin time were normal. Antithrombin III was measured and found to be normal (75%, in-house clotting method).

On review the line of bruising was found to correspond to the occlusive leg elastic of the nappy.

Treatment with heparin and warfarin was complicated by gastrointestinal haemorrhage, requiring blood transfusion, and warfarin resistance. Warfarin was stopped eight weeks after diagnosis and the patient discharged.

Nine years later the patient represented to the same paediatric hospital. In the intervening period she had been well with no further thrombotic problems. Her father and paternal

uncle had recently been diagnosed as having type I antithrombin III deficiency. On this occasion both the patient and her asymptomatic, elder sister were found to be antithrombin III deficient. Their functional antithrombin III was 66% and 58% respectively (normal range 74–126%) and immunological antithrombin III antigen in both 0.44 U/ml (normal range 0.62–1.05 U/ml).

Discussion

Thromboembolic complications are rare in childhood. There are no reports in the literature of healthy infants developing spontaneous deep venous thrombosis. When apparently well infants present with thrombotic events either an underlying prethrombotic state, or recently acquired disorder, are present.³ Thromboembolic events in healthy antithrombin III deficient infants are also rare.⁴

In the infant described, compression of the femoral vein by overtight nappy elastic appears to have impaired venous return and precipitated thrombosis in an antithrombin III deficient individual. Impaired blood flow is a recognised thrombotic risk factor in the neonate.³

The erroneously normal antithrombin III value initially reported is explained by the clotting method employed, which was technically difficult and prone to error. Nine years later, when the analysis was repeated, the assay had been superseded by more accurate immunological and chromogenic methods.

This report illustrates that seemingly minor secondary events superimposed upon an inherited prethrombotic disorder may result in the early presentation of the affected individual. Mothers of antithrombin III deficient children should be warned about the risks of venous stasis and overtight nappies.

- 1 Hirsh J, Piovella F, Pini M. Antithrombin III deficiency. *Am J Med* 1989; 87 (suppl 3B): 34s–8.
- 2 Demers C, Ginsberg J, Hirsh J, Henderson P, Blajchman M. Thrombosis in antithrombin III deficient persons. Report of a large kindred and literature review. *Ann Intern Med* 1992; 116: 754–61.
- 3 Gibson B. Normal and disordered coagulation. In: Hann I, Gibson B, Letsky E, eds. *Fetal and neonatal haematology*. London: Ballière Tindall, 1991: 123–88.
- 4 Mitchell L, Piovella F, Ofusu F, Andrew M. Alpha-2-macroglobulin may provide protection from thromboembolic events in antithrombin III deficient children. *Blood* 1991; 78: 2288–304.

Department of Paediatrics, McMaster University, Hamilton, Ontario, Canada

R Soutar
V Marzinotto
M Andrew

Correspondence to:
Dr R Soutar, Ward 47,
Aberdeen Royal Infirmary,
Aberdeen AB9 2ZB.

Accepted 28 July 1993

Left leg venogram showing occlusion of the external iliac and femoral veins, with collateral filling through the long saphenous system.