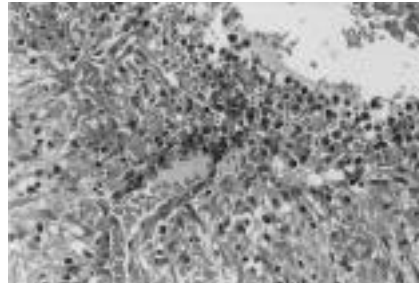


## Letters to the Editor



(a)



(b)

**Fig. 1.** (a) Significant intimal fibrous thickening of the temporal artery (hematoxylin-eosin x 40); (b) fibrin deposition in the luminal surface of the intima of the temporal artery, with numerous eosinophils present (hematoxylin-eosin x400).

healthy looking young man with a completely normal physical examination and painless, normally palpated temporal artery pulses. Chest x-ray was normal as were his biochemical and serologic profile, including CRP. His ESR was 3 mm, his Hb 13.6 g%, the WBC count 6620/mm<sup>3</sup> with 8.5% eosinophils. The biopsy was re-evaluated and the diagnosis of JTA was made (Fig. 1). The patient has remained asymptomatic, since then, performing normally.

Four cases of the disease were reported for the first time in 1975 by Lie *et al.* (1), who coined the term "juvenile temporal arteritis" for an otherwise asymptomatic disease presenting with a painless lump at the temporal artery region in older children and young adults. The histopathology had revealed intimal proliferation and a lymphohistiocytic infiltrate containing many eosinophils but no giant cells, and some degree of focal disruption of the internal elastic lamina. A fifth case was reported in 1986 (2), the sixth in 1994 (3), two cases of bilateral JTA in 1995 (4) and 1996 (5) each, a ninth case in 1999 (6), and the tenth in 2002 (7), interestingly in an elderly woman who otherwise fulfilled the criteria for the disease. Besides the absence of clinical symptoms and the histopathologic picture characteristic for the disease, there is some question regarding the presence of peripheral eosinophilia. In 6 of the 10 cases so far described, an eosinophil count was not reported (1,3,4). Out of the remaining four, two had impressive peripheral eosinophilia (2,5), one had a normal count (6), and the other had a count just above the upper normal limit (7). Our patient had also an eosinophil count just above normal.

Watanabe *et al.*, who reported the tenth case (7) in an elderly female, suggested that JTA is an accessory form of Kimura disease (8), which is a persistent and recurrent illness with peripheral eosinophilia and histologic findings very similar to JTA involving several arterial regions, albeit most commonly the head.

Two other diseases in which a non-giant

cell eosinophilic arteritis of the temporal artery has been reported include the acquired immunodeficiency syndrome (one case) (9) and Buerger's disease (3 cases) (10). Our patient had no evidence of either of the two conditions. Furthermore, the possibility of Churg-Strauss syndrome, which has been occasionally reported to involve the temporal artery, was easily ruled out in our patient in view of the absence of any typical or serologic finding of this disease.

In conclusion, our patient represents the eleventh case of JTA described in the literature, and as such it is worth reporting, in order to provide as much information as possible for a disease that quite rare, as has already been suggested by several investigators. Furthermore, this report, being the second of this rare disease described in the rheumatologic literature, will sensitize colleagues regarding this unusual temporal vasculitis affecting children and young adults.

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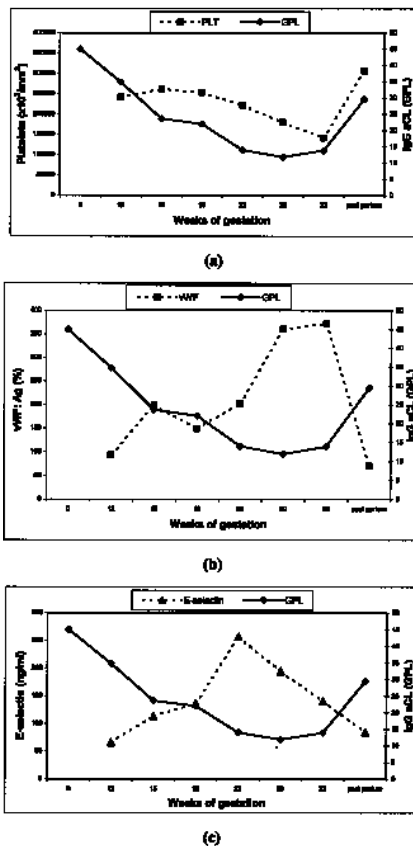
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### Could placental abruption be an antiphospholipid antibody related disorder?

Sirs,

A 35-year-old woman was referred to us at her 12th week of gestation. An unexplained placental abruption occurring at the 22nd week of pregnancy with loss of a normal male fetus had taken place one year before. Antiphospholipid antibodies (aPL) tested according to International Consensus statement (1) repeatedly revealed a medium level of immunoglobulin (Ig) G anticardiolipin antibodies (aCL), while IgM aCL, lupus anticoagulants and IgG/IgM anti- $\beta_2$ -glycoprotein I antibodies (anti- $\beta_2$ GPI) were absent.

As the patient's clinical and laboratory data were in accordance with the International Classification Criteria (1), definite APS was considered to be present and anticoagulant prophylaxis was started with nadroparin (Seleparina, Italfarmaco, Italy) at a dosage of 2,850 anti-Xa U self-administered subcutaneously twice a day. Subsequently nadroparin doses were gradually increased to 6,650 U twice daily in order to guarantee an anti-factor Xa level of between 0.1 and 0.6 U/mL over a 24-hour period. By the 12th week of gestation a steady fall in IgG aCL titer was observed, reaching its lowest values at the 22nd, 29th and 32nd weeks, when the most significant decreases in the platelet count were recorded (Fig. 1). During these same periods all coagulation



**Fig. 1.** (a) Pattern of IgG aCL levels and platelet number; (b) pattern of IgG aCL levels and von Willebrand values; (c) pattern of IgG aCL levels and E-selectin values.

parameters and antibodies to platelet factor 4-heparin complex were within the normal range. At the 32nd week of pregnancy, vaginal bleeding associated with abdominal pain manifested unexpectedly. Ultrasonography showed a large area of placental separation with a number of retroplacental clots and fetal heart activity was absent. The patient was thus delivered by uncomplicated caesarean section of a dead fetus. The placenta weighed 370 g and a retroplacental haematoma on the maternal surface without villous tissue damage was observed. Histological examination confirmed that the blood clot was situated outside the intact placental wall. The fetus was a normal male, weighing 2040 g (65th percentile). Available stored sera were retrieved and some markers of endothelial perturbation including von Willebrand factor antigen (vWF:Ag), intracellular cell adhesion molecule-1 (ICAM-1), vascular cell adhesion molecule-1 (VCAM-1), E-selectin and antibodies to endothelial cell (AECA) were tested for. ICAM-1, VCAM-1 and AECA were within the normal ranges in all serum samples, while the highest levels of vWF:Ag and E-Selectin were found at the 22nd,

29th and 32nd weeks, when the lowest values of IgG aCL were noted (Fig. 1). Spiral artery changes are generally considered to be important to the pathogenesis of retroplacental haematoma (2). This endothelial cell disorder may lead to several alterations, including vessel ruptures followed by retroplacental haemorrhage (3). Antiphospholipid antibodies (aPL) have been considered a cause of abnormal physiological changes in the spiral arteries of the placental bed (4). Using *in vitro* studies Di Simone *et al.* (4) demonstrated that either  $\alpha_2$ -GPI-dependent or  $\alpha_2$ -GPI-independent aCL bind to trophoblast cells. They thus postulated that this binding might represent a potential pathogenetic mechanism for a defective placentation in women with APS. Chamley *et al.* (5) observed that antibodies to human cardiolipin alone, to  $\alpha_2$ -GPI alone or to cardiolipin/ $\alpha_2$ -GPI complexes inhibit the proliferation of human choriocarcinoma cells. They therefore speculated that proliferation of endovascular trophoblast cells could be inhibited in women with aCL. In accordance with these studies we hypothesize that the decreases in IgG aCL titers occurring in our patient as the pregnancy progressed might be a consequence of aCL binding to trophoblastic cells during placentation. The occurrence of some signs of endothelial perturbation such as a rapid increase in vWF:Ag and E-selectin levels along with a decrease of platelet number, when the lowest values of IgG aCL were observed, might be indicative of endothelial damage causing spiral artery rupture and placental abruption.

Moreover, the absence in our patient of anti-endothelial antibodies and abnormalities in ICAM-1 and VCAM-1 levels during pregnancy would exclude the presence of endothelial activation with an inflammatory response. This later mechanism has been described by several authors (6-8) who report endothelial activation by anti- $\alpha_2$ -GPI or  $\alpha_2$ -GPI-dependent aCL through the adherent cofactor  $\alpha_2$ -GPI. Enhanced levels of ICAM-1 and/or VCAM-1 were correlated in these studies with increased adhesion of leukocytes to endothelium and with vascular thrombosis.

Our data suggest that in APS women with a history of placental abruption and positivity for IgG aCL, monitoring aCL, platelet, vWF:Ag and E-selectin levels might be a useful procedure during pregnancy. A steady fall in aCL titers associated with a decrease in the platelet count and an increase in vWF:Ag and E-selectin might in fact predict placental abruption.

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## Pulse steroid treatment of polymyalgia rheumatica

Sirs,

Corticosteroids are the standard treatment for polymyalgia rheumatica (PMR). Their clinical response is dramatic. However, the total dose of steroids used by PMR patients is high and associated with side effects in as