last fifteen years. On physical examination, he had apthous penile ulcers. Due to the swelling of the supraclavicular fovea, the jugular veins and the right arm, obstruction of the superior vena cava and the right brachiocephalic vein was suspected. Computed tomography of the chest and magnetic phlebography of thoracic vessels showed thrombosis with partial recanalization of the superior vena cava and thrombosis in the right brachiocephalic vein. An extensive laboratory evaluation for thrombophilia was negative. Papulopustules developing at venipuncture sites were considered as an equivalent to a positive pathergy test. The therapeutic approach involved the administration of glucocorticoids, anticoagulant therapy and cyclophosphamide.

This case report confirms the rare but documented appearance of obstruction of the superior vena cava in patients with ABD. Venous thrombosis occurs in 7% to 34% of patients with ABD. Its mechanism remains largely unknown, but it is suggested to be more closely related to vasculitis than to clotting disorders. Male gender and a positive pathergy test are associated with higher risk of venous thrombosis[3]. Some authors support the use of corticosteroids in all cases of deep venous thrombosis. In cases of thrombosis of the vena cava or cerebral venous sinuses, an additional immunosuppressive agent should be administered[4]. Nevertheless, thrombosis in the dural sinuses, vena cava and Budd-Chiari syndrome carry a poor prognosis.

To our knowledge, stenosis of the right brachiocephalic vein has not yet been described in patients with ABD, and it was attributed to multiple venous thromboses with partial recanalization of the right brachiocephalic vein. A case of complete thrombosis of the supraazygos superior vena cava, brachiocephalic veins and right internal jugular vein in a patient with ABD has only recently been reported[5]. It seems likely that unusual deep venous thrombosis will be detected more often in disorders of hypercoagulation with the use of modern imaging techniques.

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Acute vulvar edema a rare consequence of preeclampsia may characterize the severity of the disease

Dear Editor

Preeclampsia is a disease unique to human pregnancy characterized by new-onset hypertension and proteinuria after the 20th week of gestation. It affects 6-8% of all pregnancies and is one of the leading causes of maternal and fetal morbidity and mortality worldwide. Voluminous vulvar edema in association with preeclampsia is a rare sign during pregnancy. In our case a primigravida women with acute appearance of massive vulvar edema in ongoing twin pregnancy with severe preeclampsia further developed an acute renal failure. Vulvar edema during pregnancy may characterize the severity of the disease. Because of poorly controlled blood pressure of our patient, deteriorated renal function the following 9 hours from her admission and the non-reassuring recordings of the fetal heart rates a caesarean section (CS) was decided. Because of prematurity, neonates were transferred in the Neonatal Intensive Care Unit (NICU), while mother was transferred to the Intensive Care Unit because of severely deteriorated renal function and of the uncontrolled blood pressure. Forty-eight hours later biochemical blood tests, physical findings improved dramatically, while the vulvar edema almost disappeared. The first infant died 10 days after birth because of intraventricular hemorrhage while the second one remained in NICU for 60 days and was discharged when her body weight was 2880 grams.

Preeclampsia ranges from mild disease to a severe form that endangers the life of both mother and the fetus. Severe edema in preeclamptic patients is very rare. The current evidence encloses only five cases related to this issue [1-4]. Preeclampsia associated with acute renal failure presents in 5-10% of severe cases of preeclampsia with a predicted mortality rate of 10% [5]. Recovery is usually complete if the acute phase passes without further complications.

Persistent hypertension despite anti-hypertensive therapy, progressive deterioration in renal function and non-

reassuring fetal testing were the evidence of disease progression and the reason of our decision to perform CS in the 32 weeks of gestation. Two cases of massive antepartum vulvar edema in patients with preeclampsia were reported who also failed to be managed with medication and delivered by a CS due to fetal distress [3]. In another case of severe preeclampsia with vulvar edema whose condition deteriorated despite medical treatment with the development of ascites and signs of impending eclampsia [4]. CS again was considered the best choice for a safe termination of the pregnancy.

Severe preeclampsia during pregnancy can be associated with acute appearance of vulvar edema complicated with acute renal failure which usually disappears after delivery. Early recognition of such cases is essential to plan a safe delivery preventing from adverse outcomes both mother and fetus.

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Bullous pemphigoid: combined diagnostic criteria are still needed

Dear Editor

There is no doubt that the pathogenetic pathway of bullous pemphigoid (BP) is undiscovered in its whole spectrum. Literature is being enriched with further experience on combined diagnostic procedures.

In our study we used combined methods to establish diagnosis of BP in 40 patients (75.1 ± 11.8 years), 17 men and 23 women. 15 healthy volunteers and 15 patients with pemphigus vulgaris were enrolled as controls. Enrollment of patients was based on the presence of typical clinical and histological picture, as well as positive direct or indirect immunofluorescence (DIF or IIF). Blood sera were examined by BP180NC16A and BP230 ELISAs, prior to initiation of treatment.

Anti-BP180ELISA had a sensitivity of 67.5%, while sensitivity of anti-BP230 ELISA was 40%. Total sensitivity (for both antigens) was 77.5%, a value that was competitive to DIF. Specificity of anti-BP180 and anti-BP230 assay was 100%, while diagnostic accuracy was 81.4% and 65.7%, respectively. When both ELISAs were used in combination, specificity was 100% and diagnostic accuracy was 87.14%. Levels of autoantibodies were higher in patients with widespread lesions.

Atypical features do not allow clinical picture of BP to become a definite criterion. The percentage of atypical BP presentations is estimated to be around 40% [1]! Joly et al, has considered significant the co-existence of four clinical criteria: age > 70 years, absence of residual scars, absence of mucosal lesions and lesions on the head and neck [2]. In that study, 75% of patients had positive at least 3 out of 4 criteria [2]. In our study, 77.5% of patients were positive for all clinical criteria, while 22.5% of patients had 3 out of 4.

Histological findings are diagnostic in 50-70% of cases[1,3]. Histology is connected to the age of lesion. Old blisters or lesions with poor inflammatory infiltrates may be misdiagnosed.

DIF is often performed in inadequate specimens, resulting in a pathology report with the conclusion “suggestive”. IIF depends on the phase of BP. Chaidemenos et al, comparing clinical, histological and immunofluorescence findings, observed a variety of results without intracorrelation. The most important conclusion was that 25% of patients would have been under a wrong diagnosis if IIF was not performed [1].

Immunoblotting is a method with high potential in identifying pathogenic antigens. However, it isn’t a practical method for everyday use. On the other hand, ELISA is an easy technique, able to detect specific circulating autoantibodies and quantitate their levels. Sensitivity increases with ELISAs that detect several BP180 extra or intracellular epitopes or proteins of hemidesmosomes [4].

In conclusion, combined diagnostic criteria are still needed for the diagnosis of BP.

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