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PHOTO QUIZ

A patient with hepatitis B, liver and kidney dysfunction and polyneuropathy

L. Klieverik[1], M. Mallant[2], M. Sekkat[1], J. Branger[1*]

Department of [1]Internal Medicine and [2]Radiology, Flevoziekenhuis, Almere, the Netherlands, *corresponding author: tel.: +31 (0)36-868 87 17, fax: +31 (0)36-539 88 98, e-mail: jbranger@flevoziekenhuis.nl

CASE REPORT

A 65-year-old male was admitted to our hospital because of postprandial nausea and vomiting, weight loss (approximately 19 kg) and night sweats in the last two months. He also reported loss of sensibility in his feet and difficulty walking for approximately two weeks. Recently, he was diagnosed with an HBe-antigen positive hepatitis B (HBV) infection.

Physical examination revealed hypertension. Furthermore, there was bilateral sensibility loss of the lower extremities and peroneal paresis, compatible with acute polyneuropathy. Laboratory investigations showed mild normocytic anaemia with an elevated erythrocyte sedimentation rate, leucocytosis (19.8 x10^9/l; 86% granulocytes), impaired kidney function (eGFR 63 ml/min) and elevated liver enzymes, predominantly cholestatic. Based on symptomatology and biochemical alterations suggestive of multi-system involvement, computed tomography was performed early on in the diagnostic process (*figures 1* and *2*).

Figure 1. *CT angiography showing maximum intensity projection (Mip) in transversal plane of liver and coeliac trunk. Note the multiple aneurysm of the medium-sized hepatic artery branches*

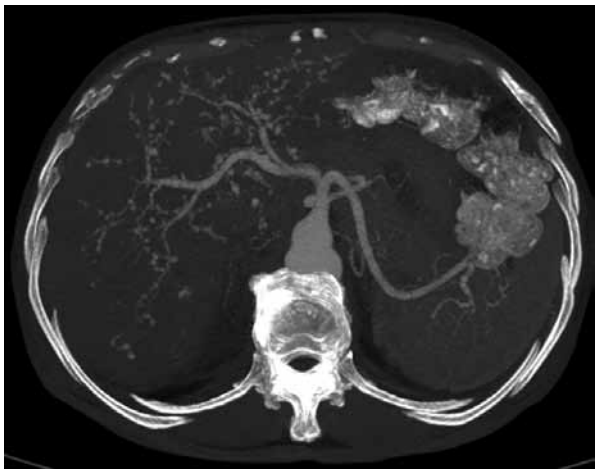
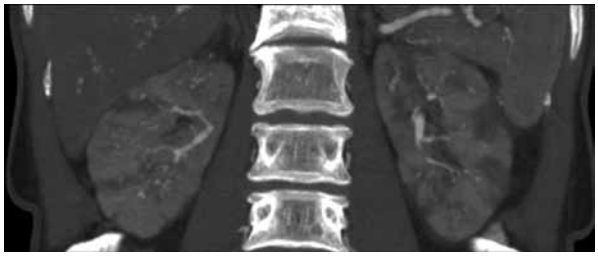


Figure 2. *Arterial phased CTA showing Mip in the coronal plane of kidneys and renal arteries. Note the multiple small aneurysms of the medium-sized branches of the renal artery and several small cortical perfusion defects*



WHAT IS YOUR DIAGNOSIS?

See page 184 for the answer to this photo quiz.

ANSWER TO PHOTO QUIZ (PAGE 181)

A PATIENT WITH HEPATITIS B, LIVER AND KIDNEY DYSFUNCTION AND POLYNEUROPATHY

DIAGNOSIS

The microaneurysms found on CT scan in combination with a recently diagnosed HBV infection raised the suspicion of polyarteritis nodosa (PAN). This diagnosis was further supported by autoimmune serology, showing negative ANCA, anti-dsDNA and low to normal complement titres.

PAN is a systemic necrotising vasculitis with typical involvement of medium-sized arteries in various organ systems. The incidence varies between $2.4/10^6$ per year in West-European countries, and $77/10^6$ per year in HBV hyperendemic areas.[1] PAN is associated with several infectious diseases, of which HBV is the most common. Among patients with PAN, approximately 7% have an active HBV infection. Of patients with HBV infection, roughly 1% develop PAN.[2] PAN typically occurs as a subacute complication of HBV infection within six months after infection. The pathogenesis of HBV-PAN is not fully understood. It is hypothesised that deposition of (HBe-anti HBe) immune complexes in vessel walls induces focal inflammation resulting in stenosis, thrombosis, formation of microaneurysms and/or rupture.[2] Symptoms are often constitutional. In addition, symptoms are related to the organ system involved. Hypertension, renal insufficiency, orchitis, livedo reticulairis and peripheral neuropathy are most frequently reported.[1] When PAN is suspected, the diagnosis is confirmed by arterial biopsy or angiography showing aneurysms or occlusions of visceral arteries. In our patient, the CT images clearly showed microaneurysms in the liver and kidneys. We therefore refrained from any further invasive diagnostic tests. Differential diagnosis includes microscopic polyangiitis, Wegener's granulomatosis, Churge-Strauss vasculitis and SLE. Therapy consists of immunosuppression (corticosteroids, cyclophosphamide) in combination with antiviral therapy.[3] Without therapy, outcome is poor. With therapy, the five-year survival is approximately 75%. HBe-antigen seroconversion is highly associated with the absence of relapses and a good prognosis.[4] In our patient, the described therapy resulted in a clear clinical and biochemical improvement within several weeks.

REFERENCES

1. Ebert EC, Hagspiel KD, Nagar M, Schlesinger N. Gastrointestinal involvement in polyarteritis nodosa. Clin Gastroenterol Hepatol. 2008;6(9):960-6.
2. Trepo C, Guillevin L. Polyarteritis nodosa and extrahepatic manifestations of HBV infection: the case against autoimmune intervention in pathogenesis. J Autoimmun. 2001;16(3):269-74.
3. Segelmark M, Selga D. The challenge of managing patients with polyarteritis nodosa. Curr Opin Rheumatol. 2007;19(1):33-8.
4. Guillevin L, Mahr A, Callard P, Godmer P, Pagnoux C, Leray E, et al. Hepatitis B virus-associated polyarteritis nodosa: clinical characteristics, outcome, and impact of treatment in 115 patients. Medicine. (Baltimore) 2005;84(5):313-22.