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Case Report

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Association between Sclerosing Cholangitis and Paget Disease: Diagnostic Difficulties

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Abstract

A rare case of association between primary sclerosing cholangitis and Paget's disease emphasizing the diagnostic difficulties in front of increased alkaline phosphatase is reported. The association between sclerosing cholangitis and Paget's disease wasn't yet described and could thus be coincidental. However, our observation underlines the benefit of dosing ALP isoenzyme to characterize the bone or hepatic origin of ALP and therefore, help to guide the diagnosis.

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Introduction

Paget's disease (PD) is characterized by an acceleration of bone remodeling responsible for an isolated increased alkaline phosphatase (ALP) [1]. It is a frequent component of multisystem proteinopathy and may therefore lead to other medical conditions. Thus, arthritis may be caused by bowing of long bones in the leg, distorting alignment and increasing pressure on nearby joints. Moreover, cardiovascular disease can result from severe PD such as calcification of the aortic valve, aortic stenosis, left ventricular hypertrophy and eventually high-output congestive failure. Kidney stones are also more common in patients with PD. Finally, the teeth may become loose, nervous system problems may occur and angioid streaks may develop, possibly as a result of calcification of collagen or other pathological deposition [2]. However, no association with slerosing cholangitis (SC) (primary or secondary), which is due to inflammation and fibrosis of biliary tract that causes biological cholestasis [3,4], has already been described in the literature. We report a rare case of association between sclerosing cholangitis and Paget's disease emphasizing the diagnostic difficulties in front of increased ALP.

Case Report

We report the case of an asymptomatic 49 years old male patient, in which a routine check objectified a biological cholestasis (gammagmutamytransferase = 2-3N and ALP = 5-6 N without hyperbilirubinemia or cytolysis). Nos past medical facts were noted. Abdominal ultrasound, viral markers and antibodies measurement (Ac Anti-nuclear, anti-Mitochondrial, anti-LKM1, Anti-cytoplasmic) were normal. Magnetic resonance choalngiopancreatography objectified multiple biliary strictures and parietal irregularities evocative of SC (Figure 1). Colonoscopy showed no associated inflammatory bowel disease. Patient received high doses of ursodeoxycholic acid (20mg/kg) for the SC with partial improvement of liver function but persistence of a marked rise in ALP level. In order to better characterize the nature of ALP, a dosage of ALP isoenzymes was performed and objectified a predominant bone fraction (83%), while liver fractions H1 and H2 were respectively of 12% and 4%. X rays objectified bone condensations with a fibrillar appearance and bone hypertrophy suggestive of PD (Figure 2). A bone scan made for lesions mapping showed a multifocal PD (Figure 3). The patient was

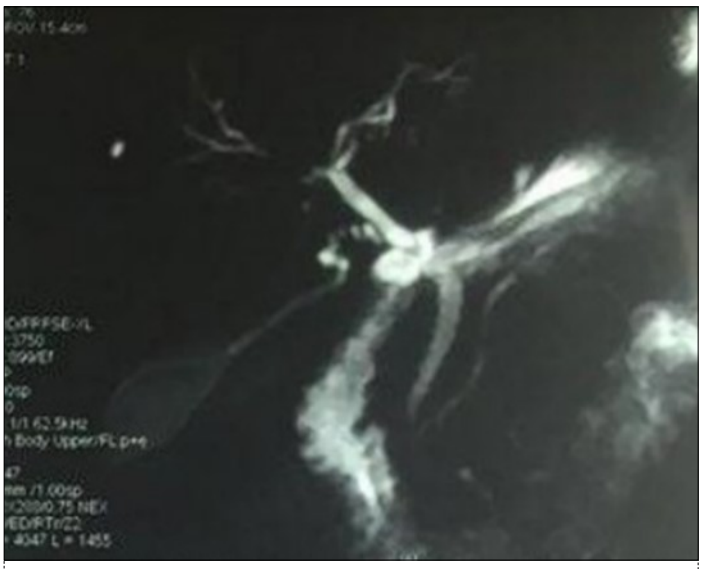

Figure 1. Magnetic resonance cholangiopancreatography showing multiple biliary strictures and parietal irregularities

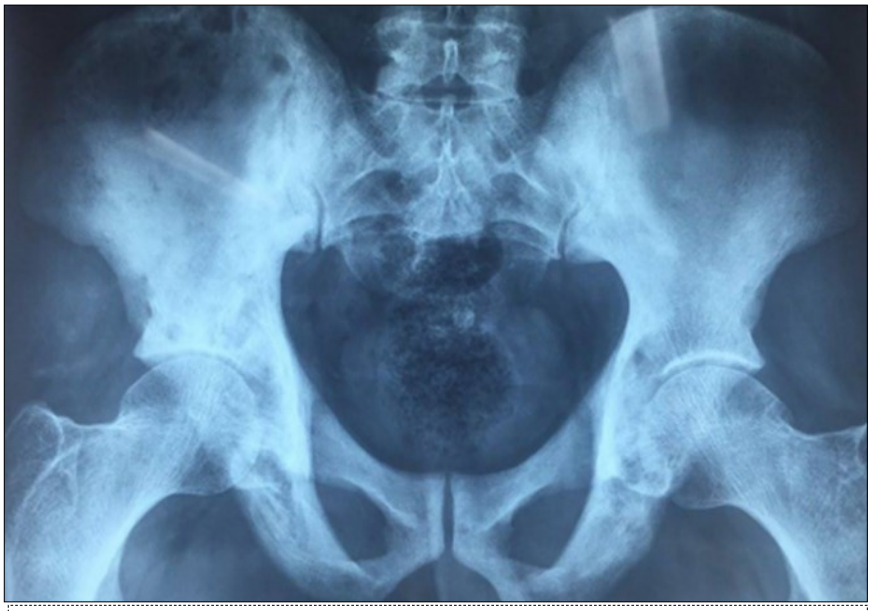

Figure 2. X ray of the pelvis showing condensations with a fibrillar appearance and hypertrophy of the bone

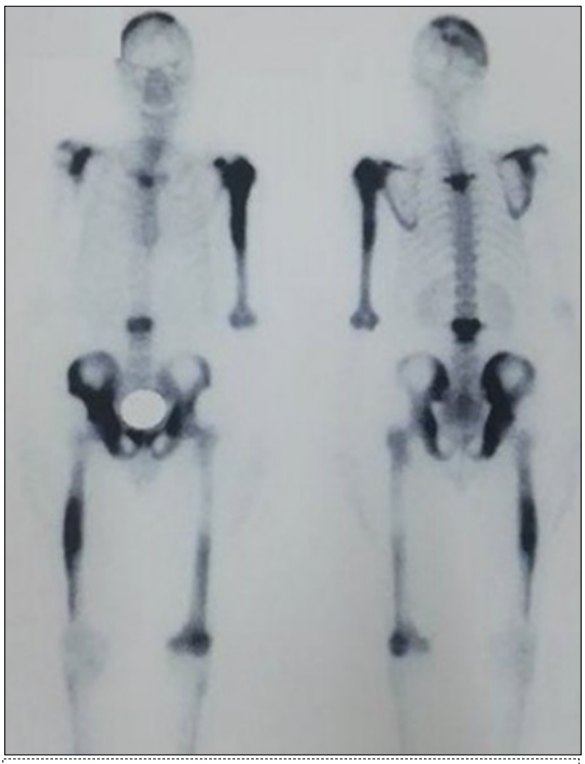

Figure 3. Bone scan mapping lesions showing a multifocal achievement of the bones

treated by bisphosphonates (injections of zoledronic acid), which was associated with a decreasing in ALP level after 6 months.

Commentary and Conclusion

Based on data from the literature, the association between SC and PD wasn't yet described, despite of the high number of secondary causes of SC [3,4]. This association could thus be coincidental, or may also be explained by immunological or genetic common disorders in both diseases [2]. No complications (nervous or cardiovascular as well as sarcoma) were noted in our case. However, in our case, a persistence of increased ALP level leaded to the diagnosis of PD in a patient having SC, and this despite a well-received treatment based on high doses of ursodesoxycholic acid [5]. One more proof of the association was the favorable outcome of biological markers after bisphosphonates treatment [6]. Alkaline phosphatase is divided into four isozymes depending upon the site of tissue expression and different biochemical and immunological methods have been used to discriminate between and selectively assay the different ALPS at the enzyme and protein level [7]. Our observation underlines the benefit of dosing ALP isoenzyme to characterize the bone or hepatic origin of ALP and therefore, help to guide the diagnosis.

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