Exceptional Case

AA amyloidosis due to chronic oxalate arthritis and vasculitis in a patient with secondary oxalosis after jejunoileal bypass surgery

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Abstract

We report a case of a woman with secondary oxalosis after jejunoileal bypass surgery for obesity, who presented with oxalate stone disease and renal insufficiency requiring dialysis. Thirty years after surgery, longstanding osteoarticular symptoms were recognized as oxalate arthritis. Eventually, she also developed oxalate vasculitis, which improved with corticoid treatment and intensification of dialysis. Work-up for kidney transplantation revealed AA amyloidosis on gastric and colonic biopsies. Since no other cause of chronic inflammation could be identified, it was concluded that the amyloidosis was secondary to oxalate arthritis and vasculitis. To our knowledge, this is the first report on this association.

Keywords: AA amyloidosis; bariatric surgery; hyperoxaluria; oxalate arthritis

Background

Oxalosis is caused by either primary or secondary hyperoxaluria. Primary hyperoxaluria is a rare, autosomal recessively inherited metabolic disorder arising from a deficiency in either alanine:glyoxylate aminotransferase (type I) or glyoxylate reductase/D-glycerate dehydrogenase (type II) [1]. Secondary hyperoxaluria can be due to overconsumption or production of oxalate from exogenous sources [2]. More commonly, however, hyperoxaluria occurs in the setting of small intestinal malabsorption.

Well-known renal manifestations of hyperoxaluria are oxalate stone disease with or without obstructive renal failure, oxalate-induced interstitial nephritis and chronic renal insufficiency. Systemic oxalosis results in generalized calcium oxalate deposition, including the heart, blood vessels, joints, bone and retina. Among clinical manifestations that can be seen are cardiac conduction defects, distal gangrene, osteoarticular manifestations (with synovitis and subperiosteal cortical defects) and oxalate crystal deposits in retinal epithelium and macula [3].

In this report, we describe a patient with enteric hyperoxaluria after bariatric jejunoileal bypass surgery, who developed secondary amyloidosis associated with longstanding inflammation due to oxalate arthritis and vasculitis.

Case report

A female patient, born in 1949, underwent jejunoileal bypass surgery for obesity in 1975. Follow-up was uneventful and her condition remained well until the late 1990s, when bilateral nephrocalcinosis was documented on ultrasonography. Enteric hyperoxaluria was suspected. In spite of efforts to avoid progressive renal insufficiency (low salt and low oxalate diet, blood pressure control), chronic haemodialysis had to be initiated in 2004. Because of recurrent urinary tract infections and obstruction due to lithiasis, right nephrectomy was performed. Light microscopy confirmed the diagnosis of oxalosis by the finding of multiple birefringent crystalline structures (oxalate crystals) in the interstitium (Figure 1).

In April 2006, at 57 years, the patient was admitted because of persistent pain in the back, shoulders, knees, hands and feet, accompanied by acrocyanosis and livedo reticularis. She reported suffering from diffuse arthralgias for many years. Treatment with non-steroidal anti-inflammatory drugs, colchicine, and short courses of steroids and antibiotics (for tentative diagnosis of bacterial overgrowth-associated arthritis), however, had only limited effects. Retrospective review of the medical files revealed that every episode of arthralgia had been accompanied by systemic inflammation as indicated by high C-reactive protein levels (CRP) (up to 190 mg/L). Based on clinical appearance, longstanding history of...
secondary oxalosis and high oxalate serum levels (77 µmol/L, normal range 11–27 µmol/l), the diagnosis of oxalate arthritis and vasculitis was made. Initially, a favourable clinical response to maintenance treatment with steroids was seen. In September 2006, however, she presented with cutaneous vasculitic lesions and necrotic toes for which amputation of left and right forefoot was performed. After intensifying the dialysis regimen to daily high-flux sessions, control was regained over the vasculitis problems and her general condition improved even though the oxalate serum levels remained high (45–77 µmol/L).

In the autumn of 2006, the patient underwent a colonoscopy for investigation of diarrhoea. Pathological examination showed AA amyloidosis. Later on, gastrointestinal work-up for kidney transplantation confirmed the presence of AA amyloidosis on gastric biopsies (Figure 2). Hepatosplenomegaly was present to a limited extent, but no other organ involvement (adrenal glands, heart, eyes, nervous system) could be detected.
Thorough investigation, including thoraco-abdominal computerized tomography, serial blood cultures, dental examination and immunological screening for underlying rheumatoid arthritis and other autoimmune inflammatory disease, could not identify any other cause of longstanding inflammation than oxalate arthritis and vasculitis. Moreover, left nephrectomy, which was performed in order to prepare the patient for transplantation, showed only oxalate and amyloid deposition and no signs of chronic pyelonephritis.

Discussion

To the best of our knowledge, this is the first report on AA amyloidosis due to chronic oxalate arthritis and vasculitis in a patient with secondary oxalosis after jejunoileal bypass surgery.

Chronic diseases that are associated with a sustained acute-phase response (e.g. rheumatoid arthritis, spondylarthropathy, chronic infections, periodic fever syndromes) can be complicated by reactive systemic AA amyloidosis [4]. In the present case, we hypothesize that chronic inflammation due to oxalate arthritis and vasculitis caused AA amyloidosis. Indeed, the longstanding arthralgias, arthritis and eventually also vasculitis due to oxalate deposition caused a chronic inflammatory state as evidenced by the high CRP values we detected. No other causes of chronic inflammation or infection were found.

Enteric hyperoxaluria may occur in patients with intestinal malabsorption from a variety of causes. Under physiological conditions, ingested calcium binds intraluminally with oxalate to form an insoluble complex. In the case of extensive small intestinal bypass or short bowel syndrome, intraluminal calcium binds preferentially with bile salts due to malabsorption and steatorrhea. This results in excessive amounts of oxalate entering the colon, to be absorbed there. Colonic oxalate absorption occurs in this setting due to mucosal alterations brought about by the entry of malabsorbed fatty acids and bile salts in the colon [5].

The patient described here developed chronic renal insufficiency for which renal replacement therapy was required. Although far less common, the oxalosis was also complicated by arthritis and eventually vasculitis, clinically presenting as necrotic ulcers and livedo reticularis—a rare complication previously described by Nakazawa et al. [6]. Amputations of both forefeet were necessary. Under high-dose corticoids and especially after intensifying haemodialysis to daily sessions with a high-flux membrane, the clinical problem of ulcers and vasculitis stabilized. Indeed, during haemodialysis, an efficient elimination rate of plasma oxalate overcomes the production rate of endogenous oxalate. However, due to the poor solubility and the very slow mobilization rate of calcium oxalate from tissue deposits, the blood pool cannot be refilled rapidly enough to keep up with the elimination rate. Therefore, it is assumed that additional sessions rather than a prolongation of a single session of haemodialysis would improve the removal of oxalate [7].

In conclusion, this is the first report on AA amyloidosis due to chronic oxalate arthritis and vasculitis in a patient with secondary oxalosis after jejunoileal bypass surgery. The lack of earlier reports on this association may be explained by the fact that long-term survival after extensive bariatric surgery only became possible the past few decades. The medical community should be aware of this potential complication and a high index of suspicion is warranted.

Conflict of interest statement. None declared.

References

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