Intestinal Perforation in Temporal Arteritis, Associated with Paroxysmal Nocturnal Hemoglobinuria


Temporal arteritis (TA) is an adult-onset, focal granulomatous inflammatory disorder of the small and medium sized arteries. Intestinal perforation is a rare complication of TA. Regarding its etiology, steroid-induced or arteritis-induced ulceration have been proposed. We describe a patient who developed TA in addition to preceding paroxysmal nocturnal hemoglobinuria. During steroid therapy for TA, intestinal perforation manifested, and it was proven to be arteritis induced perforation on histological examinations. The patient may be the 5th reported case of TA complicated with arteritis-induced intestinal perforation. The possibility of polyangitis overlap syndrome of TA and polyarteritis nodosa is discussed.

Key words: temporal arteritis, intestinal perforation, arteritis, polyangitis overlap syndrome, polyarteritis nodosa

Introduction

Temporal arteritis (TA), an adult-onset, focal granulomatous inflammatory disorder of the small and medium sized arteries, is rarely complicated by intestinal perforation. Regarding its etiology, steroid (1)- or arteritis-induced ulceration has been proposed; with respect to the latter etiology, arteritis close to the perforated ulcer has been histologically demonstrated in only 4 patients (2–5). We describe here a patient who developed TA in addition to preceding paroxysmal nocturnal hemoglobinuria (PNH). During steroid therapy for TA, intestinal perforation manifested, and was proved to be arteritis-induced on histological examinations.

Case Report

A 63-year-old man was hospitalized because of fever, general malaise, stiffness of the neck and jaw claudication, muscle pain of the chest and back, and loss of more than 4 kg body weight. When he was 17 years old, he developed pulmonary tuberculosis and later underwent left thoracoplasty twice at the ages of 24 and 37. At the time of these surgeries, he received blood transfusions and subsequently developed chronic hepatitis which was later identified as type C. At the age of 38, he was diagnosed as having aplastic anemia-PNH syndrome based on pancytopenia, reticulocytosis, hemoglobinuria, low neutrophil alkaline phosphatase activity, and positive results for acidified serum lysis (Ham) and sucrose hemolysis tests. The pancytopenia and hemoglobinuria were well controlled by persistent androgen therapy and by short-term administration of corticosteroids (for the hemoglobinuria).

On admission, his body temperature was 38.3°C, and tremor in the hands and jaw was noted. The patient did not have muscle pain or stiffness in the extremities. Focal sensory or motor disturbance (mononeuropathy) was observed in the distal regions of the extremities. Bilateral superficial temporal arteries were markedly hypertrophic. Blood pressure was normal. Hematological examination revealed a white cell count of \(4.5 \times 10^9 \text{L}^{-1}\), platelet count of \(90 \times 10^9 \text{L}^{-1}\), hemoglobin concentration of 125 g L\(^{-1}\) and reticulocytes 6.3%. Bone marrow aspirate showed normocellular marrow with erythroid hyperplasia. Serum haptoglobin level was low, while C-reactive protein (CRP) was elevated to 132 mg L\(^{-1}\). Serum creatinine phosphokinase level was normal. Renal function was intact. Tests for rheumatoid factor, antinuclear factor, anti-DNA antibody, and complement (C3 and C4) as well as Coombs’ test all...
Tsuyuoka et al yielded nonspecific results. A biopsy specimen of the left superficial temporal artery showed proliferative granulomatous arteritis with giant cells (Fig. 1). From these findings, a diagnosis of TA was made in addition to aplastic anemia-PNH syndrome.

The patient was given prednisolone (orally), and striking clinical improvement including a decreased CRP level was observed. The initial dose of prednisolone was 20 mg/day, then 7 days later, the dosage was elevated to 30 mg/day. The low dosage of prednisolone was determined based on the persistent chronic bronchitis and active chronic hepatitis. However, 20 days after the original initiation at 20 mg/day, the patient developed severe abdominal pain and melena. Plain X-ray film of the abdomen demonstrated free air under the diaphragm. At emergency laparotomy, a perforated jejunal ulcer and generalized peritonitis was observed. After partial resection of the jejunum, jejunostomy was performed. Additional surgery was required because panperitonitis was caused by a ruptured suture in the artificial anus. Disseminated intravascular coagulopathy developed and he died of respiratory failure. Histological examination of the resected jejunum showed arteritis of the small arteries in the submucosa close to the perforated ulcer (Fig. 2). Generalized arteritis of the small arteries, involving the gastrointestinal tract, liver, lung and kidneys, was demonstrated at autopsy. Giant cell arteritis, however, was not observed in these small arteries.

Discussion

In TA, intestinal manifestations are uncommon, and especially, intestinal perforation is a rare complication of this disease. On the other hand, intestinal perforation caused by arteritis is rather common in polyarteritis nodosa (PN). Although physical and laboratory findings showed typical features of TA in the present patient, 3 of 10 criteria for PN (6) were present, namely, weight loss of more than 4 kg, myalgias, and mononeuropathy. In 1986, Leavitt and Fauci proposed a new syndrome of polyangitis overlap (PO) syndrome (7) based on the fact that overlap can exist among many of the well-defined vasculitic diseases. As one component of PO syndrome, they reported one patient with features of TA and PN. From this point of view, the present patient might have been diagnosed as having PO syndrome of TA and PN. Intestinal perforation was not described in their patient, while in our patient, histological examination revealed arteritis of the small arteries in the portion of perforated jejunal ulcer, indicating that the perforation was due to the arteritis. It would be interesting to determine whether TA patients who developed arteritis-induced intestinal perforation had features of PN because of the rarity of intestinal perforation in TA and the relatively common incidence of intestinal perforation in PN. Therefore, we reviewed the 4 TA patients cited above (2–5); however, we could not determine whether their disease was PO syndrome of TA and PN due to insufficient information. Nevertheless, PO syndrome of TA and PN should be taken into consideration when treating a TA patient complicated with arteritis-induced intestinal perforation.

The association of TA and PNH appears extremely rare; to our knowledge, this association has never been reported. The exact relationship of these two diseases is unclear. Environmental factors such as infections or contact with birds have been proposed as possible factors in the pathogenesis of TA (8); therefore, immunological stimulation may be important in the initiation of this disease. In the present patient, long sustained immune responses caused by pulmonary tuberculosis, chronic hepatitis or PNH might have contributed to the development of TA.
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References