The patient was diagnosed with systemic vasculitis, probably eosinophilic granulomatosis with polyangiitis (EGPA) based on the pathologic features. Prednisone 60 mg/d and cyclophosphamide (CYC) 100 mg/d were administered orally. Then, he gradually improved. His steroid dose was tapered to 10 mg/d, and CYC was stopped 3 months later.

Six months later, the skin and sclera turned yellowish again. Meanwhile, the patient experienced progressive fatigue and itching, which caused him deep concern. His biochemical test showed elevated total and direct bilirubin, 15.3 mg/dL and 13.8 mg/dL, respectively, with high alkaline phosphatase (ALP) (1289 U/L) and gamma-glutamyltransferase ($\gamma$-GT) (1837 U/L). The white blood cell and eosinophil counts were also elevated, amounting to 25.93 × 10^9/L and 18.84 × 10^9/L (72.7%), respectively. A repeat CT scan showed a solitary mass embracing the head of the pancreas, ductus choledochus, and duodenum (Figure 1). Percutaneous transhepatic biliary drainage was performed to relieve the jaundice, and a repeat biopsy by endoscopic retrograde cholangiopancreatography (ERCP) suggested non-Hodgkin’s lymphoma (NHL).

In this case, a middle-aged male had an abdominal mass and obstructive jaundice. The common causes of obstructive jaundice and mass include biliary tract or pancreatic cancer and gallbladder carcinoma. IgG4-related diseases may be rare causes. This patient was suspected of having cancer or carcinoma when he underwent the first operation, but the...
biopsy did not support these working diagnoses, instead suggesting an inflammatory mass. He also had an increased acute-phase reactant suggestive of systemic inflammatory diseases. Considering his elevated serum acute-phase reactants, high peripheral eosinophil count, being negative for autoantibodies, and normal serum IgG4 level, he was diagnosed with systemic vasculitis, most probably EGPA. He responded well to glucocorticoids and immunosuppressants. However, the patient had jaundice within 6 months, and a repeat biopsy proved that he had NHL.

Hypereosinophilia is caused by various factors, including parasitic infections, allergic disorders, drug hypersensitivity, adrenal insufficiency, and connective tissue diseases such as EGPA, sarcoidosis, IgG4-related disease, and lymphomas. Idiopathic hypereosinophilic syndrome (HES) neoplastic diseases, such as primary (or neoplastic) HES, acute eosinophilic leukemia, systemic mastocytosis, and lymphoma, although rare, may also be implicated and should be differentiated.

EGPA is a major vasculitis syndrome associated with eosinophilia. Patients typically present with asthma or allergic rhinitis as prodromal symptoms, which most commonly involve the lung, skin, cardiovascular, gastrointestinal, renal, and peripheral nervous systems (PNS).[1] Vasculitic symptoms typically develop over the years. Positive ANCA is found in 20–50% of patients with EGPA, and the main pathological features include eosinophilic infiltration, eosinophilia, giant cell vasculitis, and sometimes interstitial and perivascular necrotizing granulomas.[2] The patient had no history of asthma or allergic rhinitis and did not have typical manifestations of vasculitis-related skin or PNS involvement. He was diagnosed with EGPA because of eosinophilic infiltration and vasculitis on biopsy. Furthermore, mesenteric masses are not common in EGPA.

Further pathological examination of the dissected mass showed lymphocytes cells at the peripheral area of the mass, vasculitis, eosinophilic infiltration, and necrosis. These features can mimic vasculitis. In a retrospective study, 16 out of 2642 patients evaluated for eosinophilia were found to have lymphoma.[3] Therefore, lymphoma should be considered an important differential diagnosis for patients with atypical clinical manifestations of EGPA.

Figure 1: The mesenteric mass wrapped around the head of the pancreas, ductus choledochus, and duodenum. (Blue arrow).
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