

plementemia require elucidation but may relate to immune complexes in the blood and those detected in some organs, particularly the kidney and pancreas. Two major questions are the participation of other IgG subclasses in immune-complex formation and the direct role — if any — of IgG4 in this process.

Finally, Vaglio and Zwerina note elevated serum IgG4 concentrations in the Churg–Strauss syndrome. Th2 cytokine pathways are typical of both diseases, and eosinophilia occurs in the blood and tissue of both. Important differences exist, too, such as the absence of granulomatous inflammation and necrotizing vasculitis in IgG4-related disease. But the clinical features of these two disorders do indeed overlap, and each must be considered in the differential diagnosis of the other.

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Since publication of their article, the authors report no further potential conflict of interest.

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Case 2-2012: Dyspnea and Rapidly Progressive Respiratory Failure

TO THE EDITOR: In the Case Record, Kotton et al. (Jan. 19 issue)¹ describe a case of angiosarcoma of the chest wall associated with the Kasabach–Merritt syndrome (KMS). This case was reminiscent of one at our hospital involving a 77-year-old man who presented after 1 month of progressive neck swelling and purpura with respiratory failure. Initial laboratory findings revealed anemia, thrombocytopenia, and coagulopathy with elevations in prothrombin time, partial-thromboplastin time, and D-dimer. A parotid biopsy with immunohistochemical staining confirmed the diagnosis of angiosarcoma. Eventually, the patient died from worsening coagulopathy.

KMS was first described as a consumptive coagulopathy in a 2-month-old boy with a giant capillary hemangioma and purpura.² The underlying pathophysiology is the sequestration of platelets and clotting factors within the vascular lesion, leading to disseminated intravascular coagulation, severe thrombocytopenia, microangiopathic anemia, hypofibrinogenemia, and elevated fibrin split products, which can result in a fatal event.³ However, the case reported by Kotton et al. involved only thrombocytopenia without coagulopathy, which is a vital component of KMS.

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No potential conflict of interest relevant to this letter was reported.

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THE DISCUSSANTS AND A COLLEAGUE REPLY: The laboratory findings mentioned by Lee et al. are characteristic of patients with KMS. However, case series of pediatric patients have shown that laboratory-proven coagulation defects (apart from thrombocytopenia) do not appear to be invariable features of KMS.^{1,2} In our patient, there was histologic evidence of intravascular and extravascular coagulation, with fibrin deposition in the lungs. Testing for D-dimers was not done by the clinicians but would have been helpful in confirming the presence of intravascular coagulation.

The clinicians found that the patient's international normalized ratio was essentially normal with a range of 1.1 to 1.2, but the prothrombin time was slightly prolonged (range, 13.1 to 14.2 seconds [reference range, 11.0 to 13.3]). Her severe thrombocytopenia, slightly prolonged prothrombin time, and normal partial-thromboplastin time were similar to those reported in the aforementioned case series.^{1,2} The hyperfibrinogenemia can be attributed to an acute-phase reaction, given the concomitant elevation in ferritin and C-reactive protein. Although this case may differ slightly from the initial pediatric case reported by Kasabach and Merritt, we believe the two cases share a common pathophysiological mechanism by which aberrant activation and trapping of platelets within a vascular neoplasm leads to thrombocytopenia.³

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Dr. Van Cott reports no potential conflict of interest relevant to this letter. Since publication of their article, Drs. Nishino and Kotton report no further potential conflict of interest.

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Resolution of Recurrent Focal Segmental Glomerulosclerosis after Retransplantation

TO THE EDITOR: Recurrent primary focal segmental glomerulosclerosis (FSGS) develops in over 40% of renal-transplant recipients and presents a major therapeutic challenge.¹ The first marker of disease recurrence is often proteinuria in the nephrotic range that can appear 2 to 3 days after transplantation.² Experimental findings supporting the theory of a circulating factor as the cause of primary FSGS include the rapid induction of proteinuria and foot-process effacement after the injection of FSGS serum into rats.^{3,4} Although recent research has identified soluble urokinase plasminogen activator receptor (suPAR) in the serum as the responsible circulating factor, a complete understanding of the involved cellular cascade has been elusive.⁵ Plasmapheresis often serves as adjunctive therapy to immunosuppression in patients with recurrent primary FSGS after transplantation, but the clinical benefits vary widely and have not been tested in large, randomized trials. In this report, we describe the successful retransplantation of an allograft that was failing in the first recipient owing to recurrent primary FSGS.

A 27-year-old man with end-stage renal disease caused by primary FSGS (Patient 1) received a kidney transplant from his healthy 24-year-old

sister. Given the known risk of recurrent FSGS after transplantation, plasmapheresis was performed before surgery (two sessions) and after surgery (five sessions), and standard immunosuppressive therapy was administered. Although the renal allograft functioned immediately (Fig. 1), marked proteinuria (>10 g of protein per 24 hours) developed on the second post-transplantation day. A biopsy specimen from the allograft obtained on day 6 revealed a normocellular glomerulus with prominent podocytes and marked podocyte foot-process effacement and loss of the interdigitating arrangement, consistent with disease recurrence (see Panels A and B of the figure in the Supplementary Appendix, available with the full text of this letter at NEJM.org).

The renal allograft was removed on post-transplantation day 14 because of persistent proteinuria, worsening hypoalbuminemia (1.5 g per deciliter), rising creatinine (8.3 mg per deciliter [733.7 μ mol per liter]), and the development of an intraabdominal hematoma. At this time, after consulting with the hospital ethics committee and internal review board and obtaining informed consent, we approached the first recipient about donating his kidney transplant to another patient on the transplant waiting list.