A 12-year-old girl with severe idiopathic panuveitis and macular cystoid edema, requiring adalimumab (40 mg every 2 weeks) and cyclosporine (3 mg/kg/d) for the past 20 months, presented with a 4-month history of multiple mucocutaneous lesions. The first began as an area of superficial erythema over the third toe of her right foot, which subsequently evolved into a vegetative lesion. A dermatologist diagnosed this as a wart, but it was refractory to topical treatment with imiquimod 5%. One month later, an erythematous vegetative mass appeared in the left nasal septum, with posterior extension to the hard palate and left tonsil. The lesion was unresponsive to topical bacitracin and tobramycin and to oral flucloxacillin. The patient denied fever, weight loss or cough. She had contact with horses and had an aquarium but denied any preceding trauma or previous travels.

On physical examination, she was well appearing and had a temperature of 36.2°C, heart rate of 88 beats/min, blood pressure of 116/62 mm Hg and respiratory rate of 25 breaths/min. She had a nodular, 2 × 2 cm erythematous vegetative lesion on the left side of her nasal septum and hard palate with smaller ones on her left tonsil. On the dorsum of the third toe of her right foot, she also had an erythematous verrucous plaque that was hardened and rough with some crusts (Fig. 1). There was no cervical or submandibular adenopathy and no hepatosplenomegaly. The rest of her physical and neurologic examination was normal.

Laboratory studies showed a hemoglobin level of 12.8 g/dL, white blood cell count of 14.9 × 10^9/L, with differential of 56.4% neutrophils, 36.8% lymphocytes, 4.2% monocytes, 2.1% eosinophils and 0.5% basophils, platelet count of 322 × 10^9/L, erythrocyte sedimentation rate of 53 mm/h and C-reactive protein level of 0.9 mg/L. Transaminases, serum electrolytes and creatinine were all normal. Interferon-gamma release assay and HIV and Epstein-Barr virus antibody tests were negative. A computed tomography scan (Fig. 2) demonstrated a 3-cm cavitary mass in the left nose extending to the basal vestibule, with no evidence of invasion or infiltration of adjacent structures.

An additional test confirmed the diagnosis.

For Denouement see P. 1160.