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Von Recklinghausen's Disease Associated with Angiodysplasia of Colon and End-Stage Renal Disease

Dear Sir,

We present a case of von Recklinghausen's disease associated with hypertension, angiodysplasia of colon and end-stage renal disease. Our patient, a 62-year-old farmer, was admitted to hospital because of anuria of 5 days' duration. The signs and symptoms compatible with von Recklinghausen's disease had drawn attention already 20 years earlier. Since then, the quantity and size of these signs had increased progressively. The patient had a strong family history that we could suggest autosomal dominancy. He had been found to be hypertensive for the first time 6 months before.

Physical examination revealed multiple soft, small tumoral masses on his face and body which were pathologically diagnosed as being fibromas. He also had café-au-lait spots and diffuse freckles. We observed respiratory distress, distended jugular veins and subcrepitan rales. The laboratory values on the first day of admission were as follows: Hb 6.8 g/dl, WBC 11,500/mm^3, platelets 374,000/mm^3, ESR 60 mm/h, BUN 129 mg/dl, Cr 11.6 mg/dl, ANF (–), anti-DNA (–), HbSAg (–), anti-HCV (–), urinary Na 84 mEq/ml. Abdominal ultrasonography (USG) revealed congestive hepatomegaly and bilateral parenchymatous renal disease; the kidneys were smaller than normal. Pelvic USG was normal. Radiologic examination of lung and colon and computed tomographies of the brain, abdomen and thorax were normal. ECG revealed T negativity at DII, DIII, avF, V_5 and V_6.

Because of hyperkalemia, uremic encephalopathy and congestive heart failure, a peritoneal dialysis was performed. On hospital day 2, these signs and symptoms disappeared. He began to extract urine. He was started on maintenance hemodialysis 2 months after the initial admission to hospital. We observed hematochezia on hospital day 30. Nasogastric lavage was clear and upper gastrointestinal endoscopy was type B gastritis, monilial esophagitis and bulbitis without any bleeding focus. But by colonoscopy, multiple bleeding angiodysplastic lesions were found on the lumen of the transverse colon. Six units of blood were transfused. Approximately on day 10 of bleeding, he received an estrogen pill which consisted of 0.03 mg ethinyl estradiol/day, orally. On day 3 of estrogen therapy, hematochezia stopped and blood transfusion was not required. Rectal biopsy was found to be normal and negative for amyloidosis. Renal biopsy was performed on hospital day 18. There was complete sclerosis in 50% of glomeruli, and the other ones revealed enhancement of Bowman's capsule. Periglomerular and interstitial fibrosis, focal calcification, tubular atrophy, hyaline casts in tubulus lumens, endothelial and medial thickening of arterioles were also observed. Focal deposition of IgG, IgA and IgM were found positive with immunohistochemical methods. The biopsy diagnosis was tubulointerstitial nephritis and hyaline atherosclerosis.

Urinary complications or hypertension may occur in neurofibromatosis in three separate circumstances; involvement of the urinary appartus by the neurofibromatosis and spread to the kidney of the vascular lesions of von Recklinghausen's disease; pheochromocytoma is found in 3% of patients with neurofibromatosis. We did not find pheochromocytoma in our patient. Neurofibromatosis may itself produce vascular lesions. Aneurysm of renal artery, causing partial stenosis, may be responsible for hypertension. In other cases, there may be diffuse vascular involvement, sometimes accompanied by secondary parenchymatous lesions, which may cause hypertension and proteinuria [1].

The cause of angiodysplasia is unknown but is most probably acquired and the result of a degenerative process associated with aging [2]. Chronic low-grade submucosal venous obstruction, hypo-oxygenation of the intestinal mucosa due to atherosclerotic peripheral vascular disease, connective tissue and ground substance abnormalities of the vessels as in connective tissue disease or with prolonged topical corticosteroid therapy and aluminum hydroxide antacids are some causes of arteriovenous malformations [3]. Angiodysplasia has been purported to occur with higher frequency in patients with renal failure. However, it is not known whether formation of these lesions precedes or follows onset of renal dysfunction. In this case we draw attention to the possible association between neurofibromatosis and chronic renal disease, and between neurofibromatosis and angiodysplasia [1]. And also, for gastrointestinal bleeding due to angiodysplasia, ethinyl estradiol can be used effectively and safely in addition to other therapeutic modalities.

References

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