A Young Female Case of Polyarteritis Nodosa Strongly Suspected by Typical Angiographic Findings Which Improved Rapidly after Prednisolone and Cyclophosphamide Therapy

Hideo Yamada, Masashi Oka, Seiko Mori, Takeshi Takahashi, Shoji Kuwata, Chikara Aizawa, Gotaro Toda and Kiyoshi Kurokawa

A 16-year-old girl was admitted with the complaints of headache, chest pain, low abdominal pain and left hemi-numbness. Her blood pressure was high and plasma renin activity and aldosterone levels were elevated. Renal angiography revealed vascular stenoses and micro-aneurysms although the renal artery and its main branches were not involved. Polyarteritis nodosa (PN) was strongly suspected and oral prednisolone and intravenous pulse therapy of cyclophosphamide were started. The second renal angiography which was performed 11 days after the therapy was started, showed marked improvement of vascular lesions. This is a case which suggests that the angiographic findings of PN can improve very rapidly with therapy.

Key words: hypertension, micro-aneurysm

Introduction

Polyarteritis nodosa (PN) is a disease of unknown origin which usually occurs in middle-aged men. The diagnosis of PN is made based on the morphological evidence of vasculitis, which is often difficult to obtain. On the other hand, there are several reports (1–3) that emphasize the importance of angiography for the diagnosis of PN. We report here a young female who had the typical angiographic findings of PN, which rapidly improved in response to steroid and cyclophosphamide therapy.

Case Report

A 16-year-old girl was admitted with the complaints of headache, chest pain, low abdominal pain and left hemi-numbness. Her blood pressure was 170/120mmHg and high plasma renin activity and aldosterone levels were observed (17.3 ng/ml, 49 pg/dl, respectively). Her blood pressure was normal six months earlier and she denied any drug abuse. Then, chest pain, abdominal pain and left hemi-numbness also appeared in the following months and she was admitted to our hospital on August 1989.

Her blood pressure on admission was 162/110 mmHg bilaterally. On physical examination, there was no edema and no skin eruption. Lymph nodes were not palpable. There were no abnormal findings in the chest and abdomen, and no pathological sign was seen neurologically. Laboratory data showed red blood cell and white blood cell counts of 463 X 10^4/μl, and 5,000/μl, respectively, and eosinophilia was absent. CRP was negative but ESR was slightly elevated (15 mm/h). Speckled type antinuclear factor (ANF) was weakly positive (40×). Hepatitis B antigen was negative. Routine blood chemistry was normal except for a low potassium level (3.2 mEq/l). Urinalysis revealed proteinuria of 0.6 g daily and urine sediments showed only a few mixed casts. Plasma renin activity and aldosterone levels at rest in the morning were extremely elevated (14 ng/ml, and 27 pg/dl, respectively). Catecholamine series and ACTH-cortisol series were normal. On the studies of abdominal ultrasonography and computed tomography (CT), the size of both kidneys was the same but their surfaces were irregular. Neither enlargement of adrenal
glands nor an abnormal mass was seen. Renal angiography was performed, which showed multiple vascular stenoses and micro-aneurysms in peripheral branches of the renal arterial tree, as depicted in Fig. 1. At the same time, renin activity and aldosterone levels in both renal veins were checked, which were proven to be high bilaterally (right 23.8 ng/ml and 26.3 pg/dl; left 28.6 ng/ml and 22.8 pg/dl, respectively). Although the muscle biopsy of biceps did not reveal any sign of vasculitis, we strongly suspected this case to be PN due to the multiple symptoms and typical renal angiographic findings.

As the left hemi-paresis and convulsion appeared and the abdominal pain rapidly worsened after the renal angiography, oral prednisolone therapy (60 mg per day) was started followed by intravenous pulse therapy (1 g cyclophosphamide). Her symptoms disappeared gradually and ESR was also decreased. Hypertension was controlled by the angiotensin-converting enzyme inhibitor and nifedipine (Fig. 2). To evaluate whether the abdominal pain, left hemi-paresis and convulsion...
were also related to the vascular abnormalities, visceral, superior and inferior mesenteric and the right carotid arteriographies were performed 11 days after the prednisolone therapy was started (3 days after cyclophosphamide injection), but no specific lesions were detected. Therefore, in order to confirm the abnormal findings detected in the first renal angiography, a second renal angiography was performed. It showed remarkable improvement of the vascular stenoses and a decrease in the number and size of the micro-aneurysms (Fig. 3). The third angiography, performed 52 days after the therapy was started, confirmed the improvement seen at the second angiography.

Discussion

Here we report a young female with PN in whom renal micro-aneurysms rapidly improved after prednisolone and cyclophosphamide therapy. There are many reports (3–7) which indicate a high incidence of male PN patients. Adue et al (7) reported that two-thirds of 43 PN patients were males, but the three patients under the age of 30 were all females.

The final diagnosis of PN is generally made by biopsy of muscle, skin, or kidney. Positivity of muscle biopsy is, however, low (20–25%) (8), and in some cases renal biopsy is not recommended because it may be accompanied by massive bleeding due to ruptured aneurysms (1). Thus the importance of angiography in the diagnosis of PN has been emphasized and it is thought that microaneurysms are specific for PN when rheumatoid disease, systemic lupus erythematosus, gram-negative septicemia and malignant hypertension are ruled out (1–3). Furthermore, this case satisfied at least two items of "1990 criteria for the classification of polyarteritis nodosa" of the American College of Rheumatology (9). Therefore, we strongly suspected PN although the clinical symptoms and signs were not typical and the muscle biopsy was negative.

At the second angiography, arteries in addition to the renal arteries were examined because the patient had obvious abdominal pain, left hemi-paresis and convulsion, but there was no evidence of vasculitis. We considered the possibility that some psychological factors might be involved in these symptoms because she was reluctant to go to high school and they appeared just after she entered, and because her complaints were sometimes inconsistent with the physical findings and laboratory data. But the relation with PN could not be completely ruled out, as these symptoms improved dramatically after therapy was started, and the abnormal angiogram can not always be obtained even if symptoms and/or signs exist (6).

This case showed the improvement on renal angiogram findings only 11 days after the therapy was started. McClude and Westcott (10) reported one case in which renal micro-aneurysms decreased 20 days after steroid therapy was started. This case, however, had a poor prognosis. Since it has been suggested that angiographic improvement does not always indicate a good prognosis (11), a long-term follow-up will be necessary for this patient.

References