

A case of Polyarthritits Nodosa

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Abstract : A 76-year old man was admitted with disturbance of consciousness, epigastric pain, appetite loss. Clinical evaluation revealed congestive heart failure, cardiogenic shock, pre-renal renal failure, urinary tract infection liver congestion, liver dysfunction. Treatment with intravenous infusion, inotropic agents, diuretic drugs, antibiotics improved circulation status, renal function, infection. But he died of ventricular tachycardia suddenly. Autopsy revealed fibrinoid necrotizing inflammation of small and medium-sized muscular arteries accompanied by infiltration of eosinophilic leukocyte in kidney, lung heart. This suggests the possibility of Polyarthritits Nodosa. This disease reveals various symptoms, and it is difficult to diagnose. Polyarthritits Nodosa is one of systemic vasculitis, whose classification is complicated and not perfect. Although we couldn't diagnose before death, congestive heart failure, liver dysfunction, renal dysfunction were all explained by systemic vasculitis in retrospect. So, in multi-organ dysfunction whose cause is unclear, we should keep it mind the possibility of systemic vasculitis.

Key words : Polyarteritis Nodosa, systemic vasculitis, autopsy

I . Introduction

Systemic vasculitis represents various symptoms, and there are many forms of systemic vasculitis, besides its classification is very complicated and not perfect. Therefore, it is said to be difficult to make a diagnosis of systemic vasculitis. In the present case, we report a case of polyarteritis nodosa (PN) that was made a diagnosis of at autopsy.

II . Case Report

Patient : 76 years old male

Chief Complaint : disturbance of consciousness, epigastric pain, appetite loss

Past History : cerebral infarction at the age of 72, gastric ulcer treated with internal medication at the age of 69.

Family Histroy : mother died of colon cancer at

the age of 78

Present Illness : He had lived in a nursing home for aged since he suffered from cerebral infarction at the age of 72. Despite of dementia, he could support himself. At the beginning of August, he started complaining of epigastric pain and appetite loss, and suffered from disturbance of consciousness. Sometimes, he complained of muscle pain, even though his complains were vague because of dementia. His symptoms had developed unfavorably, so he was seen in consultation to our department on August 27. Laboratory data revealed elevation of inflammatory reaction, liver dysfunction, renal dysfunction, metabolic acidosis, bacilluria, and electrocardiogram (ECG) revealed anterior old myocardial infarction. So he was hospitalized urgently.

Physical Examination on Admission : height and weight couldn't be measured, body temperature 35.

1°C, heart rate 60 bpm irregular, blood pressure 118/40 mmHg, respiratory rate 36/min, Kussmaul respiration, Consciousness Japan Coma Scale (JCS)3 restlessness, Eyes: not anemic, not icteric, no palpable superficial lymph node, dry skin, Heart: irregular beat, no murmurs, Lung: no rales, Abdomen: flat and soft, no mass, no tenderness, no peripheral edema, no jugular vein dilatation.

Laboratory data indicating elevation of inflammatory reaction (WBC 8,270/ μ l, CRP 8.61 mg/dl), liver dysfunction (GOT 189 IU/l, GPT 106 IU/l, LDH 617 IU/l, ALP 253 IU/l), renal failure with participation of dehydration (Cre 2.9 mg/dl, BUN 76.9 mg/dl, UA 9.6 mg/dl), increase of globulin dominated by IgG, bacilluria, metabolic acidosis compensated by respiratory system, increased

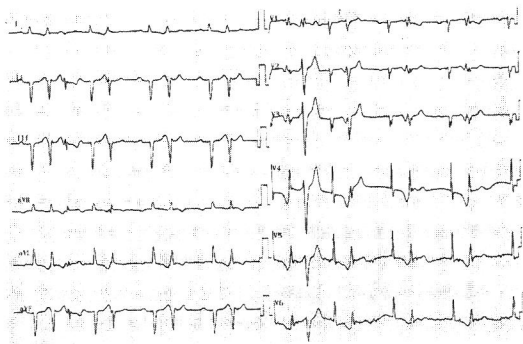


Figure 1 ECG at admission: sporadic PAC and old myocardial infarction were found

alveolar arterial difference of oxygen pressure difference (pH 7.345, PaO₂ 92.4 mmHg, PaCO₂ 12.4 mmHg, Anion Gap 25.1 mEq/l, A-aDO₂ 43 mmHg). ECG (figure 1) revealed sporadic paroxysmal supraventricular contraction (PAC) and QS pattern in leads II•III•aV_r, poor R progression in leads V₁₋₃. A chest X-ray (figure 2) showed slight lung congestion and cardiothoracic ratio was 63%. Abdominal ultrasonograph (AUS) (figure 3) revealed liver congestion and hypertrophy of gallbladder wall and no precipitate in gallbladder.

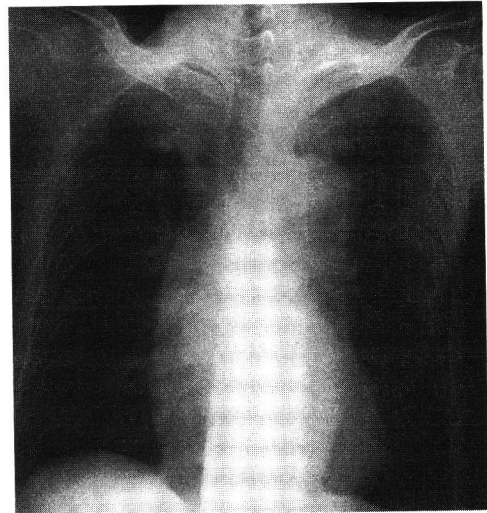


Figure 2 Chest X-ray at admission: Cardiomegaly and slight lung congestion were found

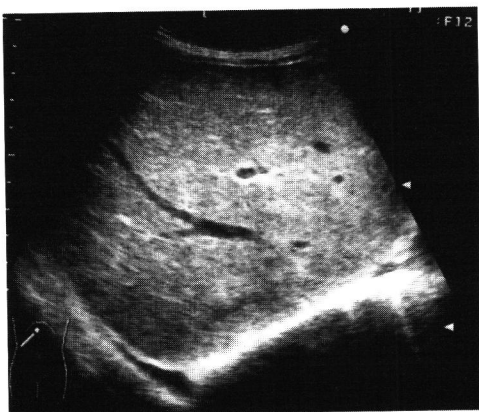


Figure 3 AUS at admission: Liver congestion were found (subcostal scan)

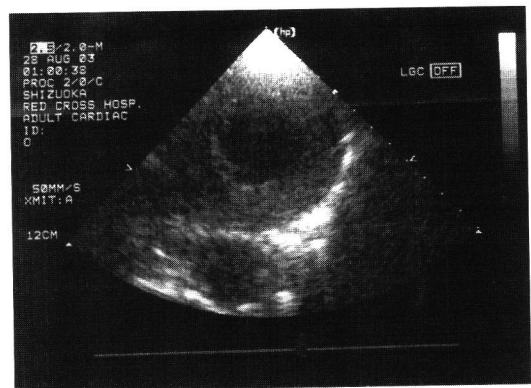
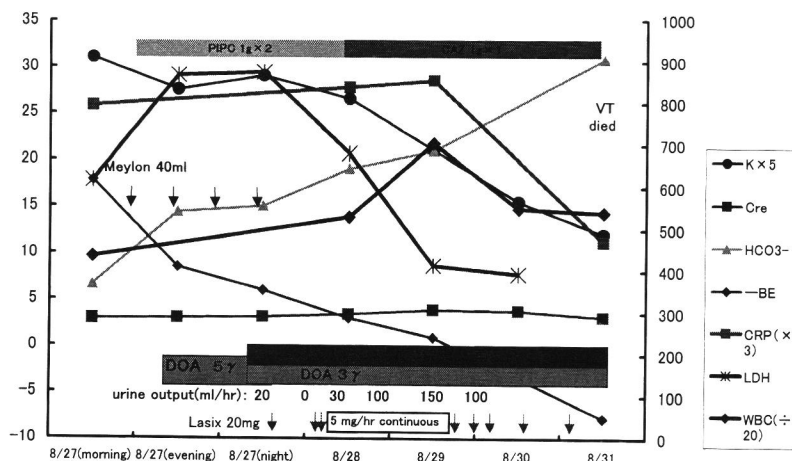


Figure 4 Echocardiogram soon after admission: Diffuse hypo cardiac wall motion and dilatation of IVC were found

Table 1 Clinical Course



Clinical course (table 1): We made a diagnosis of congestive heart failure, liver congestion, pre-renal renal failure, urinary tract infection (UTI) by cardiomegaly and lung congestion in the chest X-ray, diffuse hypo cardiac wall motion and dilatation of inferior vena cava (IVC) in the echocardiogram (figure 4), roughness of hepatic parenchyma and dilatation of hepatic vein in the AUS, elevation of liver and biliary system enzyme and renal failure and metabolic acidosis and elevation of inflammatory reaction and bacilluria in the laboratory data. A few hours after admission, the systolic blood pressure fall to 70 mmHg, and peripheral skin got cold, so we made a diagnosis of cardiogenic shock and administered intravenous infusion (2000cc/day) and dopamin at dosage of 5 μ g/kg/minute. And intravenous sodium bicarbonate and furosemide were administered as the treatment of acute renal failure. For urinary tract infection, PIPC 1g \times 2/day were administered first, but replaced with CAZ 1g \times 1/day for certain improvement. In spite of administration of furosemide, urine output was 0ml/12hr after admission, so continuous intravenous furosemide was started and dopamine was reduced at the dosage of 3 μ g/kg/minute to increase renal perfusion and dobutamine at the dosage of 5 μ g/kg/minute was combined to maintain blood pressure. With these treatment, urine output was

increased, and plasma creatinine and blood urea nitrogen (BUN) and liver and biliary enzyme were decreased, and lung congestion was improved. On 29 August, urine output was 2000ml/day, so furosemide was reduced. But urine output was 2,500ml/day after that, plasma potassium was decreased at 2.4 mEq/l on 31 August, so potassium was replaced. At this point of time, blood pressure and heart rate were stable, and body temperature was afebrile, UTI was improved. Around 3:00 AM on 1st September, wet cough and yellow sputum progressed, so frequent suction of sputum was necessary. At 4:20, suddenly ECG monitor revealed ventricular tachycardia (VT) at the rate of 140bpm, and the patient's circulation fell into shock state and the patient died at 5:11 in spite of cardiopulmonary resuscitation. His autopsy was performed 4 hours after death.

Autopsy Findings (figure 5,6)

(1) suspect of polyarteritis nodosa

- ① fibrinoid necrotizing inflammation of small and medium-sized muscular arteries accompanied by infiltration of eosinophilic leukocyte (heart, kidney, stomach, liver, colon, duodenum, pancreas, peri-adrenal gland)
- ② myocarditis

I : diffuse vasculitis of small and medium-sized muscular arteries in the parenchyma of



Figure 5 Autopsy finding in the heart (HE stain, $\times 400$): diffuse vasculitis of small and medium-sized coronary arteries in the parenchyma of myocardium with small myocardial infarction

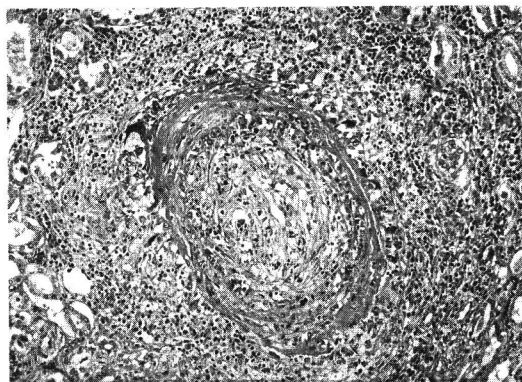


Figure 6 Autopsy finding in the kidney (HE stain, $\times 100$): vasculitis of small and medium-sized artery with glomerulonephritis

myocardium

II : myocardial infarction secondary to vasculitis

III : secondary endocarditis

IV : no apparent stenosis of coronary artery

V : pericardial effusion 30ml

③ acute renal failure with glomerulonephritis

④ cerebral infarction

⑤ liver congestion, centrilobular liver necrosis

(2) prostate carcinoma (well differentiated adenoma)

(3) pulmonary edema

(4) moderate atherosclerosis

III . Discussion

We reported a case of vasculitis. Judging from autopsy, the most possible diagnosis was polyarteritis nodosa (PN). Since classic PN was described in 1866 by Kussmaul and Maier, many classifications of vasculitis have been proposed, but it is difficult to make perfect classification¹⁾. Among vasculitis, PN is representative and divided into classic PN which is multisystemic necrotizing vasculitis of small and medium-sized muscular, and microscopic PN which can be made a diagnosis only by biopsy²⁾. The pathologies of these two disease are similar and have many common features but have a few different fea-

tures. For example, pulmonary arteries are rarely involved in classic PN, whereas pulmonary capillaritis occurs frequently in microscopic polyarteritis. And the pathology in the kidney in classic PN is predominantly that of arteritis without glomerulonephritis, in contrast, glomerulonephritis is very common in microscopic PN³⁾. PN has a striking tendency to involve medium-sized muscular arteries. It spares the aorta and its major branches, as well as capillaries and small arterioles that lack muscular coats. In contrast to many other forms of vasculitis, PN also spares the venous system. Although Kussmaul and Maier termed the disorder periarteritis nodosa, the site of the earliest lesion in PN remains a matter of debate⁴⁾. In this present case, classic PN was probable because arteritis existed in small and medium-sized muscular arteries, and this case was accompanied with glomerulonephritis. Small myocardial infarction and myocarditis associated with coronary arteritis seems to have caused low cardiac function, and the small myocardial infarction might be the origin of ectopic VT. However VT might be associated with hypokalemia. Vasculitis of small and medium-sized muscular arteries existed not only in the heart but also in the liver, kidney, intestine, pancreas, spleen. No glomerulonephritis were seen, but small infarctions existed in the kidney. Renal dysfunction

might be caused by these infarctions and low perfusion pressure of glomerulus due to dehydration and low cardiac output. In the liver, besides inflammation associated with arteritis, liver congestion might cause elevation of liver and biliary system. Hypertrophy of gallbladder wall in the AUS seems to be associated with arteritis and bacterial cholecystitis seemed to be negative because no precipitate were seen. Although brain autopsy was not performed, consciousness disturbance might be associated with arteritis of cerebral arteries.

Differential diagnosis is Churg Strauss syndrome because of infiltration of eosinophilic leukocyte and involvement of lung⁹⁾. If we had measured serum anti-neutrophil cytoplasmic antibody (ANCA), that might have been helpful to make a diagnosis. But we couldn't consider the possibility of vasculitis. Insufficient medical history and physical examination due to dementia made it difficult to make a diagnosis. But consciousness disturbance, abdominal pain, muscle pain, cardiac dysfunction, renal dysfunction, elevation of liver and biliary system enzyme, which are recognized in this case, could be caused by systemic vasculitis uniformly.

IV. Conclusion

We reported a case of PN that was diagnosed at autopsy. We should consider the systemic vasculitis as one of differential diagnosis in case of multi-organ dysfunction.

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