POLYARTERITIS NODOSA: TWO CASES AND A REVIEW

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Polyarteritis nodosa, though still a rare disease, is becoming more and more common. Credit is usually given to Kussmaul and Maier of Germany for the first description of the condition in 1866.[1] By 1914 only 5 cases had been reported in the USA; today hundreds of cases can be collected in the world literature.

The aetiology has remained obscure. Amongst others, Rich and Gregory[2] have stressed the importance of hypersensitivity as an aetiological factor. Selye had postulated that psychogenic factors, acting through the adrenal cortex, may also be a causal factor.[2] Rich[3] reported polyarteritis nodosa in a number of cases treated with sulphonamides. The increasing incidence of the collagen diseases, on the assumption that hypersensitivity and the antigen-antibody reaction are the primary aetiological factors, becomes explicable by the wider and freer use of the drugs, particularly serum, sulphonamides and penicillin. According to Rich and Gregory[2] these drugs are most frequently associated with, if not related to, the development of polyarteritis nodosa.

The following two cases of polyarteritis nodosa, both female, were admitted to the same ward (of only 30 cases) within less than a month of each other.

CASE 1. POLYARTERITIS NODOSA PRESENTING AS CARCINOMA OF THE LUNG

Mrs. M.S., aged 61, was admitted on 13 April 1956 complaining of sudden onset of fever, dyspnoea, haemoptysis, loss of appetite and a feeling of weakness, 7 days before admission. According to her husband, however, she had not been well for the last few months. She had been told she had influenza and given antibiotics without response. There was a history suggestive of a coronary episode 4—5 days before admission. She also complained of left-sided headaches for the last 4 years. There was no history of previous illnesses or of tuberculosis in the family.

Examination

The general examination was almost entirely negative, the only points of interest being (1) pyrexia of 100°F, (2) chest expansion greater on the left side, and (3) general wasting.

Urine: Albumen—trace. Red blood cells ++.

Blood: Haemoglobin 11·1 g.%. White blood cells 28,000 per c.mm. (92% of polymorphs).

Chest X-ray: On the right, (*a*) well-defined round area of consolidation in mid-zone, (*b*) apical fibrosis and consolidation posterior upper lobe, and (*c*) hilar and basal calcification. On the left, basal fibrosis.

ECG showed evidence of a recent and an old infarct.

Course in Hospital

During the stay in hospital the temperature was of swinging character, the highest temperature recorded being 102°F. The patient became extremely apathetic 5 days after admission, when the serum electrolytes per litre were: Na 131 mEq., Cl 93 mEq., K 5·5 mEq., CO_2 combining power 27 vols.%. The blood urea was 50 mg.%.

The chest X-ray looked like carcinoma of the lung. A thoracic surgeon was called in and it was decided to resort to bronchoscopy to elucidate the pathology. Bronchoscopy was abandoned because the patient had deteriorated very rapidly and it was found on further investigation that the blood urea had risen to 305 mg.%. Two days later, on 26 April, the patient died—13 days after admission.

Post-mortem Examination

Autopsy showed (1) consolidation of middle zone of right lung and left and right apices, thought to be tuberculous lesions, (2) myocardial infarction, both old and fresh, (3) almost complete infarction of the spleen, and (4) acute bilateral pyelonephritis.

On microscopic examination the heart, spleen, kidneys and lungs all showed necrotizing vascular lesions characteristic of periarteritis nodosa, the lung lesions being of a more chronic nature than those in the other organs.

Discussion

At the time of admission a diagnosis of pulmonary tuberculosis was favoured, until the chest X-ray presented an appearance more like that of carcinoma of the lung. The symptoms, though apparently acute on onset, would have passed for either condition. A diagnosis of carcinoma of the lung was then held clinically, the temperature and leucocytosis being assigned to secondary infection of the carcinoma.

The thoracic surgeon who was consulted agreed that the case looked like carcinoma of the lung, but before bronchoscopy could be performed the patient deteriorated rapidly and died in uraemia.

Pulmonary Involvement

Haemoptysis is a rare finding in this disease; it was recorded in only 1 of 243 cases of polyarteritis nodosa reviewed by Harris, Lynch and O'Hare.[4]

Miller and Daley[5] describe the lung involvement and its radiological aspects. The basic pathology appears to be perivascular infiltration, oedema and haemorrhage; it has been likened to thrombo-angiitis obliterans by Harkavy.[6] Although a miliary appearance, opacities of moderate density, pulmonary infarctions, pleural effusions and atelectasis may occur; the classical picture is one of a fan-like pulmonary infiltration due to increased vascular markings extending out from the hilar region.

Polyarteritis nodosa seems to spare the periphery of the lung, yet in this patient both apices were involved. The chest X-ray in this case was unusual for polyarteritis nodosa, and even at autopsy the provisional diagnosis (until microscopic examination) was tuberculosis, with the reservation of possible alveolar carcinoma or visceral angiitis, which alone could have explained the multiple organ involvement.

Other Manifestations

Besides the lung involvement there are further interesting aspects in this case:

Myocardial infarction is a rare finding in polyarteritis nodosa.[8] There is no characteristic ECG pattern, but flattening of the T waves and left ventricular strain are sometimes found. In this case the ECG showed evidence (confirmed *post mortem*) of both an old and a recent infarct. Microscopically lesions of polyarteritis nodosa were demonstrated in the heart.

Splenic infarction is found *post mortem* in less than a third of cases. This case showed gross infarction of the spleen. No thrombus was demonstrated in the splenic vessels.

Renal involvement and uraemia. The kidneys are involved in 70—80% of cases of polyarteritis nodosa, yet strangely enough clinical uraemia was observed in only 13% of a series of 177 cases.[4] This patient died in uraemia with a blood urea of over 300 mg.%.

CASE 2. POLYARTERITIS NODOSA WITH ANURIA THE PRESENTING FEATURE

Mrs. K., aged 44 years, was admitted on 1 May 1959 with the following complaints:

(1) Continuous backache and lower abdominal pain for 2 weeks. (2) Sensation of a lump in the throat for 10 days. (3) Itchy skin for 7 days. (4) Pins-and-needles and stiffness of hands and feet for 4 days. (5) Urinary output almost *nil* for 3 days, with anorexia and vomiting. (6) Poor vision in the right eye for 10 years, and in the left eye for 1 year.

The reason why the patient was sent to hospital was her anuria. She had only passed a cupful of urine in 3 days, and catheterization did not produce more than a few drops. The accompanying vomiting and anorexia were severe.

Past Illnesses

1946, blurring of vision of right eye (diagnosed as choroiditis). 1948, cholecystitis (refused operation). 1952, had 'yellow jaundice' n Johannesburg (given intravenous cortisone). 1953, further attack of jaundice—again given cortisone. Badly swollen legs and abdomen, with dyspnoea, later in 1953; also headaches for an indefinite period. 1955, pyelitis with severe backache similar to present backache; later in the year, dysentery and vomiting.

Examination

Pulse 100 per minute. Blood pressure 140/90 mm.Hg. Temperature normal. Soft systolic murmur over all areas. Varicose veins in both legs, with incompetent perforators. Firm 5-finger hepatomegaly. Fundi—vessels slightly tortuous on left side, with pale area on choroid. Ankle jerks absent.

Urine: Albumen—trace. Red blood cells ++. 4 pus cells per high-power field. On ureteric catheterization, right pelvis—albumen trace, red blood cells +++, few pus cells; left pelvis—albumen +++, red blood cells ++, more than 15 pus cells per high-power field, *B. coli* isolated.

Blood: Haemoglobin 9·2g.%. White blood cells 6,000 per c.mm. Differential count normal. Serum electrolytes (mEq. per litre)—Na 150, K 5·7, Cl 105·2. CO_2 combining power 24 vols.%.

Liver functions: Bilirubin 0·6 mg.%. Proteins 6·8 g.%, albumen-globulin ratio 1·3 : 1. Prothrombin index 92%.

Kidney functions: Blood urea 76 mg.% on admission; rose steadily to 123 mg.% after 2 months. PSP test—only 10% clearance after 2 hours.

Bone-marrow: Slight decrease in erythropoiesis. No L.E. cells.

X-rays: Chest normal. Three intravenous pyelograms showed no dye excretion after 1 hour. Retrograde pyelograms normal.

Biopsies: Liver—fatty change, increased fibrous tissue, periportal round-cell infiltration, and evidence of regeneration. Subcutaneous nodule—characteristic appearance of polyarteritis nodosa.

Course in Hospital

Soon after admission the patient commenced putting out good amounts of urine and the urinary output remained good throughout her course. The blood pressure rose from 140/90 mm. Hg. on admission to 170/100 after a few days, and remained more or less at this level. The ophthalmologist reported central scotomata in both eyes, old choroiditis in the left eye, early hypertensive retinitis in the left eye, and old exudates in both discs.

While in hospital the patient developed small subcuticular haemorrhages in her legs, arthritis of the left knee with effusion, and a few small subcutaneous nodules (biopsy characteristic of polyarteritis nodosa—see above).

Termination. The patient died about 3 months after her discharge from hospital in July 1956. She had been on cortisone therapy.

Discussion

The chief presenting symptom in this case was the low urinary output, which amounted to anuria (under 100 cc. daily) for 3 days. What little urine was obtained was loaded with red blood cells and albumen, but no casts were present.

Anuria. Although anuria as a presenting symptom in polyarteritis nodosa has been recorded in the literature (Rolnick and Davidson[8]), it is comparatively rare. The two main pathological conditions affecting the kidneys are haemorrhage and infarction and glomerulonephritis, the former in 55% of cases, the latter in 33%. Clinically this patient was not affected with glomerulonephritis. An interesting possibility is a massive infarction resulting in bilateral cortical necrosis; such a case was recorded by Wordley.[9]

Other Manifestations

Besides the anuria there are further interesting aspects in this case:

Hepatomegaly and jaundice. It is difficult to ascertain the significance of the hepatomegaly since this patient in 1952 and 1953 had two attacks of jaundice, which might have been due to infective hepatitis, and this might have left cirrhosis and hepatomegaly. Jaundice itself, however, has been reported in 12% of the 177 cases reviewed by Harris, Lynch and O'Hare.[4] It is quite possible that this jaundice was a manifestation of the polyarteritis nodosa. Hepatomegaly itself is not an uncommon finding in polyarteritis nodosa; it is associated with infarctions in the liver.

Visual disturbance. Choroiditis was diagnosed in hospital in this patient's left eye. Visual disturbance in this eye has been present for only 1 year. In contrast to this, poor vision has been present in her right eye for 10 years. There is a history of trauma over the right eye just before visual disturbance occurred, and it seems unlikely that polyarteritis nodosa would have caused this poor vision in 1946, although it probably did cause the choroiditis in the left eye 1 year ago.

Peripheral neuritis. This patient complained of pins-and-needles and stiffness of hands and feet and the ankle jerks were found to be absent. Gruber[10] reported a high incidence of peripheral neuritis in polyarteritis nodosa, and other series tend to confirm this. The basis of the neuritis appears to be involvement of the nutrient arteries to the nerves, resulting in ischaemia and degeneration.

Anaemia, subcutaneous nodules, arthritis, hypertension and subcuticular haemorrhages are all fairly common findings in polyarteritis nodosa. They occurred in the following percentages in the series of 177 cases reviewed by Harris, Lynch and O'Hare:[4] Anaemia 66%, hypertension 53%, arthritis 34%, petechiae and purpura 27%, and nodules 23%.

SUMMARY

Two cases of polyarteritis nodosa are reported on and compared with a review of the literature. Both females, they presented within less than a month in a ward of only 30 patients.

The fulminating character of polyarteritis nodosa in case 1 contrasts with a more chronic picture in case 2.

In the acute case the diagnosis was only established *post mortem*. In case 2 the diagnosis was made *ante mortem*.

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