PERIARTERITIS NODOSA AS A SOURCE OF SUBARACHNOID HEMORRHAGE AND SPINAL CORD COMPRESSION

REPORT OF A CASE AND REVIEW OF THE LITERATURE

HAROLD HAFT, M.D., BERNARD E. FINNESON, M.D.,* HARRY CRAMER, M.D., AND ROSA FIOL, M.D.

Neurosurgical Section, Surgical Service, Medical Service and Laboratory Service, Veterans Administration Hospital, Bronx, New York

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PERIARTERITIS nodosa has become a well recognized entity during the past 30 years. It is classified together with lupus erythematosus, rheumatoid arthritis, rheumatic fever, scleroderma and dermatomyositis in the group of collagen diseases. The following case is reported because of its unusual onset, involving subarachnoid hemorrhage and compression of the spinal cord, thus presenting an acute neurosurgical problem.

CASE REPORT

199 046. C.F., a 44-year-old white male, was admitted to the Neurosurgical Section on Aug. 25, 1955. Two weeks previously he first noted aches and pains in both thighs associated with an elevated temperature between 99°F. and 101.5°F. Within the next 2 weeks there developed in succession numbness and paresthesiae in both feet and calves, a left-sided foot drop, and paresthesiae and numbness along the C6 dermatome of the left forearm with pain in both shoulders. On the day prior to admission the patient experienced severe pain in the lower thoracic spine followed by a sudden motor, sensory and sphincteric paralysis with a level below the T9 cord segment. Lumbar puncture performed at another hospital revealed grossly bloody spinal fluid with an initial pressure of 280 mm. of water, as well as a spinal-fluid block. The history further revealed that bronchial asthma had developed 10 months before admission and that the patient was found to be sensitive to feathers, coffee, tea, cold foods, spinach and alcohol.

Examination. Blood pressure was 134/80, pulse rate 86, and temperature 101.2°F. General physical findings were normal. There was a complete motor, sensory and sphincteric paralysis with a level at the T9 cord segment, weakness in the biceps and triceps, and hypalgesia in the C6 dermatome on the left. There was no nuchal rigidity and the cranial nerves were normal.

The blood count was normal except for a white blood count of 16,200 with 78 per cent neutrophils. The urine was normal except for a trace of albumin and 1-plus glycosuria.

Emergency lumbar and cisternal myelograms revealed complete blocks at L2 and C2 respectively. The spinal fluid was grossly bloody with a total protein of 5.1 gm.

* Present address: 255 S. 17th Street, Philadelphia 3, Pennsylvania.
Operation. Laminectomy of T6–T9 was carried out on the day of admission with a preoperative diagnosis of thrombosed and ruptured varicosities. The intradural exploration revealed a solid mass of recent blood clot throughout the entire extent of the operative field; thrombosed worm-like vessels were seen and felt in the center of the exposure. Following a wide decompression the wound was closed.

Course. Throughout the immediate postoperative period the patient continued to run a temperature of 100°F.−103°F., and the neurological status remained unchanged. On Sept. 7, 1955 examination of the urine revealed 3-plus albuminuria with pyuria, hematuria and cylindruria. On Oct. 4, 1955 hemoglobin was 8.9 gm.; there were 17,000 white blood cells with 31 per cent eosinophils. This pattern continued until death, with the eosinophils varying between 19 per cent and 41 per cent. The patient was treated empirically with Gantrisin, penicillin, Terramycin and Achromycin without effect. Paralysis of the left diaphragm with pneumonitis at the left base occurred on Oct. 10, 1955. On Nov. 4, 1955 the patient complained of weakness in the right upper extremity, and paralysis of the intrinsic muscles of the hand was noted.

Because of the persistent fever, leukocytosis, eosinophilia, anemia, albuminuria, and the 10 months’ history of bronchial asthma, periarteritis nodosa was suspected. Two muscle biopsies were negative, although one showed acute and chronic myositis. Several lupus erythematosus preparations were negative. Aspiration of bone marrow revealed a marked eosinophilia. Serum electrophoresis showed decreased albumin with normal globulin. Brucellosis agglutinins, Coomb’s test, cryoglobulins and trichinosis complement fixation tests were all negative.

On Dec. 26, 1955, bulbar speech with weakness of the right lower facial muscles, the right masseter muscle and right palate was noted, as well as deviation of the tongue to the right. On Dec. 27, 1955, the patient complained of diplopia on right lateral gaze. At this time blood pressure was 160/95, and pulse rate 127. Hemoglobin was 10 gm.; there were 3.3 million red blood cells, 18,000 white blood cells with 34 per cent eosinophils, 16,500 platelets, 0.3 per cent reticulocytes, and blood urea nitrogen was 17.

On Jan. 3, 1956 the patient showed nuchal rigidity and petechiae were found on his back and chest. A Jacksonian seizure involving the left upper extremity occurred on Jan. 5, 1956. Examination revealed bilateral lower facial and palatal weakness with absent pharyngeal reflexes. Cortisone therapy was started on this date. Cheyne-Stokes respiration and difficulty in swallowing were noted on Jan. 14, 1956. There then developed a left medial rectus palsy, deafness on the right, and nystagmus on left lateral gaze. Bilateral nystagmus was present on Jan. 15, 1956 with marked impairment in left visual acuity. On Jan. 17, 1956 Cheyne-Stokes respiration recurred and the patient expired.

Autopsy. The body showed marked wasting. There was a large decubitus ulcer in the right gluteal region. There were approximately 300 cc. of yellow purulent fluid in the left pleural cavity. The vessels in the mesentery of the small bowel were hard and nodular. There were hemorrhagic spots and two large ulcerations in the gastric mucosa. There was a large ulcer in the rectal mucosa near the anus. The subcapsular surfaces of the kidneys were nodular and mottled by areas of light yellow-brown discoloration. The mucosa of the urinary bladder was severely congested. The testes were atrophied.

Microscopically there was lymphocytic infiltration into the subepicardium and hepatic parenchyma. The arterioles and smaller arteries in the heart, lungs, gastro-