Cranial Arteritis With Liver Involvement

BY DONALD C. MANN, M.D., AND JAMES F. TOOLE, M.D.

Abstract:
An elderly man with cranial nerve palsies was found to have cranial arteritis. Laboratory studies established concurrent liver disease which improved with treatment. The liver and brain stem pathology is discussed.

ADDITIONAL KEY WORDS
- giant-cell arteritis
- hepatic arteritis
- temporal arteritis
- liver disease
- alkaline phosphatase
- liver biopsy
- liver scan
- vascular disease
- steroid therapy

Cranial or giant-cell arteritis is a systemic disorder of the elderly in which the nervous system may undergo focal ischemic changes. Characteristically a deficit in the visual system develops during the illness, but there is usually a widespread inflammatory process as evidenced by fever, increased erythrocyte sedimentation rate and malaise. Coronary artery occlusion and polymyalgia rheumatica occurring in patients with biopsy-proved temporal arteritis attest to its widespread nature and point out the inadequacy of naming a disease for the site of biopsy.1

We recently had the opportunity to treat a patient with cranial arteritis who additionally had a reversible hepatopathy.

Case Report
A 75-year-old gentleman (NCBH #5193 44) had been in good health until late October, 1970, when malaise and anorexia developed which eventually caused a 15-lb weight loss. On the first of November a severe headache appeared and soon thereafter nausea and vomiting developed which lasted a week. He was hospitalized and a right lateral gaze weakness, conjunctivitis, and chemosis of the right eye were observed. Scalp tenderness was described. By the time of his transfer to North Carolina Baptist Hospital three days later on November 14, he had acquired chemosis of both eyes and facial edema. Mild headache was still present. Three days after admission here, the sixth nerve palsy had become bilateral and the patient was unable to look upward. He also acquired bilateral extensor plantar responses and his right corneal reflex disappeared. At this time convergence, optic fundi, and fields were normal and the facial and orbital swelling had subsided. The scalp was tender and the superficial temporal arteries were prominent but could be easily compressed. Caloric stimulation showed a poor response on the left side. The blood pressure was 150/80, temperature was 100°, and chest and abdominal examination were normal. Several features of his medical history were relevant. He had had progressive bilateral deafness for three years and had transient, untreated jaundice in 1945. His referring physician hospitalized him in May, 1970, for abdominal pain believed to be due to an old duodenal ulcer.

Biopsy of the temporal artery appeared normal on removal but arteritis with giant cells and eosinophils were present in sections (fig. 1). The following normal studies were obtained: brain scan, EEG, cerebrospinal fluid protein, pressure and cells, and EKG.

He was given Prednisone, 40 mg per day, with immediate improvement in headache. Over the next several days he regained lateral ocular rotations. The alkaline phosphatase rose from 140 I.U. (normal 30 to 85) on admission (November 11) to 504 I.U. at the time of transfer (November 14), then progressively fell to 170 I.U. during the first week of Prednisone treatment. In May it had been 22 I.U. The SGOT was 80 I.U. (normal 10 to 50) on the 14th and subsequently normal. Bilirubin and LDH remained within normal limits. The serum protein electrophoresis showed

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alpha-1 of 8%, alpha-2 of 19%, and beta of 18%. The sedimentation rate was 30 mm per hour. Two liver scans (fig. 2) showed similar focal defects but a liver biopsy showed no abnormality. One month after the beginning of his illness, he went home with diplopia requiring an eye patch, no headache, and general improvement, taking 40 mg of Prednisone per day.

One month after discharge, in January, he had minimal diplopia and no longer required an eye patch, had further improvement in lateral gaze bilaterally, flexor plantar responses had returned, and he was gaining weight.

Two months after discharge, in February, he had a minimal lateral gaze palsy, normal corneal responses, unchanged loss of superior gaze, and his caloric response on the left had returned to normal. The alkaline phosphatase was 55 I.U. The protein electrophoresis now showed alpha-1 of 2% and alpha-2 of 9%, while the beta fraction remained elevated at 18%. A repeat liver scan was now normal (fig. 3) and sedimentation rate had fallen to 17.

On examination in March, 1970, four months after his illness had begun, he had an unchanged superior gaze palsy and minimal bilateral sixth nerve weakness. Prednisone had been tapered to 20 mg per day.

In October, 1971, he felt well, was without headache, had normal ocular rotations and no symptoms or signs to suggest hepatic disease. He is on maintenance doses of 10 mg Prednisone daily.

**Discussion**

The liver disorder was manifested by enzyme elevation and multiple deficits on scan, but was missed on biopsy. Since both the scan and alkaline phosphatase closely followed the clinical progression and remission of cranial vascular disease and globulin elevation, they constitute a concomitant part of the arteritis. Nonmetastatic focal defects on scan such as these are seen in cirrhosis and infarction, presumably due either to Kupffer cell failure or to diminished perfusion. Biliary system regeneration which has been described in
periarteritis nodosa could account for the elevated alkaline phosphatase. Alkaline phosphatase elevation, hepatic artery involvement, and mesenteric giant-cell arteritis have all been reported in this disease but not the abnormal scan. We can only speculate that scattered hepatic arteritis caused diminished regional perfusion.

This patient's neurological deficit is also worthy of note. Visual acuity symptoms are common, whereas extraocular palsies are less frequently observed ranging from 10% to 15%. Ocular palsies are elusive, the diagnosis in two series being confirmed by findings only half as often as the complaint of diplopia. Loss of ocular movements has been attributed to individual muscle involvement by Wagener and Hollenhorst, and to third or sixth nerve ischemia by Fisher and Meadows; superior gaze palsy has generally been explained by depressed consciousness. It would appear in this patient with the extensor plantar responses and persistent superior gaze loss that the ischemia was central. Marked cerebellar ischemic alterations and less impressive brain stem changes were the only central nervous system pathology present in the case of conjugate gaze palsies and a seventh nerve weakness described by Crompton, while frank cerebellar infarction from temporal arteritis was present in Heptinsall's case No. 13.

The feature of this disease most likely to bring about a diagnosis, blindness, is irreversible. Because early treatment is necessary and the results good, biopsy may be performed on minimal evidence. The syndrome accompanied by giant-cell arteritis remains incompletely described. The screening chemistry battery which brought the presence of liver disease to
our attention will no doubt uncover further cases.

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