BRIEF REPORT

GIANT CELL ARTERITIS AND POLYMYALGIA RHEUMATICA IN A CONJUGAL PAIR

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Despite the description of 12 pairs of first degree relatives with giant cell arteritis and polymyalgia rheumatica, including affected siblings, the occurrence of these two entities in a conjugal pair has not been described. Previous reports of familial aggregation in giant cell arteritis and polymyalgia rheumatica have emphasized a genetic predisposition. A recent extensive review of giant cell arteritis by Healey and Wilske states: "The strongest factor against an environmental or contagious etiology is that the two syndromes have not been reported to occur in conjugal pairs (1)."

In this brief report, we describe polymyalgia rheumatica and giant cell arteritis in a husband and wife and discuss possible modes of horizontal transmission.

Patients. In September 1973, a 74-year-old housewife was seen at the Internal Medicine Clinic at the University of Michigan Hospital with a 6-month history of proximal muscle weakness, bitemporal scalp pain, and daily fevers to 101°F. Because of her weakness in climbing stairs, a ramp had to be constructed to enable her to ascend the five steps to her home.

Several months before evaluation in our clinic, she had experienced a dramatic symptomatic response to an empiric course of steroids. However, she relapsed when the dose was tapered.

Extreme tenderness over both temporal arteries and marked proximal muscle weakness were noted on examination. While receiving 2.5 mg prednisone twice a day, her Westergren sedimentation rate was 92 mm/hour.

In view of her classic history and previous response to steroid therapy, a clinical diagnosis of giant cell arteritis and polymyalgia rheumatica was made. An increase in prednisone to 10 mg/day led to improvement in her symptoms and decline in the Westergren sedimentation rate to 45 mm/hour.

During the ensuing 4 years she continued to experience mild muscle weakness and fatigue; suppressive treatment with steroids was hampered by gastrointestinal side-effects. She died from intestinal perforation in 1977, 4 years after her diagnosis.

Throughout the illness, her husband remained a constant companion. The husband, a retired entrepreneur and former boatman on the Taquamenon River, was 71 years old at the time of his wife's diagnosis in 1973. They had married 3 years earlier. The marriage, the second for both partners, had followed a short courtship. They had sexual relations during the 3 years of marriage prior to the wife's diagnosis in 1973 and during the 4 years of her illness. Throughout this period, the husband remained active and healthy.

In August 1980, 4 years after his wife's death, the husband was seen at the University of Michigan Hospital with a 9-month history of marked fatigue, proximal muscle weakness, intense bitemporal scalp pain, and daily fevers to 100–102°F. Because of weakness climbing stairs, he had begun entering his house on the ramp constructed for his wife during her illness.

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On examination, moderate tenderness to palpation over the temporal regions was present. Marked proximal muscle pain and weakness were demonstrated in both upper and lower extremities. His Westergren sedimentation rate was 119 mm/hour, and both SGOT and SGPT were four times the upper limit of normal. Temporal artery biopsy showed marked disruption of the internal elastic lamina membrane and infiltration with giant cells (Figure 1). One day after starting prednisone 60 mg/day, he was ambulatory without difficulty for the first time in 6 months. On a current dose of prednisone 20 mg/day he continues to do well.

Discussion. The incidence and prevalence of giant cell arteritis in the general population vary widely. In an analysis from Olmstead County, Minnesota, Huston et al found a prevalence of 133 cases per 100,000 population aged 50 years and older (2). Thus, the chance occurrence in a second individual, assuming its presence in the first, approximates 1 per 1,000. Despite this statistical possibility, giant cell arteritis has not been reported in close associates, including conjugal pairs unrelated by blood.

Family aggregation of giant cell arteritis and polymyalgia rheumatica is well described. Sibling pairs have predominated in the literature, with Barber noting 2 sisters who developed polymyalgia rheumatica within 5 years of one another (3) and Hamrin reporting 2 sister pairs and 2 sister–brother combinations with "polymyalgia arteritica" among his 93 cases (4). Most sibships had the onset of disease separated by 4 or more years. Moreover, family members in all of the studies failed to show a common household at the time of diagnosis, and all had only casual contact with one another.

A review of 24 consecutive patients with polymyalgia rheumatica and giant cell arteritis at the University of Michigan Hospital during the last 8 years failed to demonstrate any familial aggregation or any additional conjugal pairs.

Previous reports of polymyalgia rheumatica and giant cell arteritis emphasized possible genetic inheritance patterns. However, attempts to relate these clinical entities to any genotype, including HLA tissue typing, have been unsuccessful (5).

The conjugal pair with giant cell arteritis and polymyalgia rheumatica in this report suggests a possible etiology by exposure to a common antigen or

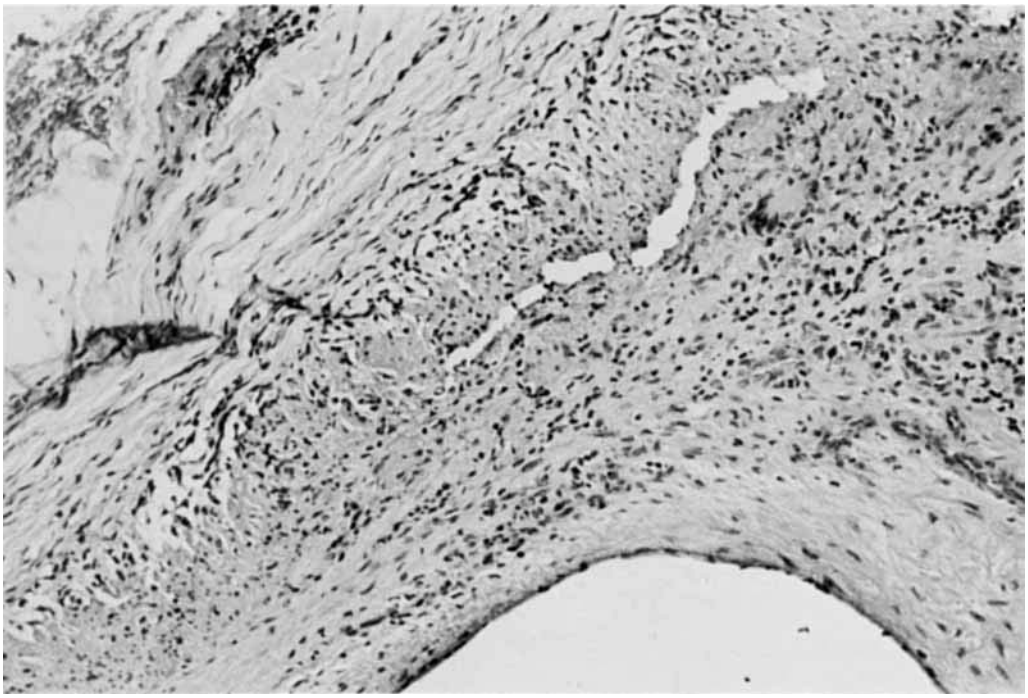

Figure 1. Section of temporal artery from the husband demonstrating disruption of media, absence of internal elastic membrane, and prominent multinuclear giant cell. (Hematoxylin and eosin; magnification ×45.)

transmission of a causative agent since they had intimate contact with one another for the 7 years of their marriage.

An infective etiology for the two illnesses has been sought, including a report of exposure to parakeets by Fessel, but this was not substantiated by further investigators (6). Mowat and Hazelman, in a series of 17 patients with polymyalgia rheumatica and giant cell arteritis, were unable to demonstrate persistent elevation of antiviral antibody titers including those to rubeola, cytomegalovirus, herpes zoster, herpes simplex, and rubella (7). Finally, a strong association between antibody to hepatitis B surface antigen and polymyalgia rheumatica was noted by Bacon et al (8). However, a specific indicator of more recent exposure to the hepatitis virus, hepatitis B surface antigen, was not detected in the sera of any of their 13 patients. The husband in this report had an elevation in SGOT and SGPT four times normal, a well-described event in giant cell arteritis; however, his sera did not contain hepatitis B antigen or antibody.

This is the first reported occurrence of giant cell arteritis and polymyalgia in a conjugal pair. Although evidence of horizontal transmission or exposure to a common source antigen remain speculative, the epidemiology is open to further investigation.

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