Whipple's Disease

Report of a Case

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ONE OF THE DISEASES included in the category of malabsorption syndromes is the uncommon entity, Whipple's Disease. Described in 1907, and known also as intestinal lipodystrophy and intestinal lipogranulomatosis, it was considered for many years to be a derangement in fat metabolism and absorption. This impression was supported by the steatorrhea, the deposit of neutral fats and fatty acids in the small intestine and mesenteric lymph tissue, and the microscopic presence of "foam cells" in the intestinal submucosa. Although it was known that these cells did not take ordinary fat stains, it was not until 1945 that evidence appeared which indicated that they did stain with the periodic acid-Schiff method. This fact seemed to suggest that the material, so stained, was a glucoprotein. Because of the mucin present in the intestinal cells, and since elevated mucoprotein levels were demonstrated in patients with this disease, the idea of involvement of a mucoprotein was evolved. This led to a wholly different concept; namely, that of a general disease, a hypersensitivity state, possibly a connective tissue disease or a collagen disease. This has been substantiated by the demonstration of typical cells, not only in the small intestine and mesenteric nodes, but in multiple foci of systemic distribution. Uncommon, fascinating because of changing concepts, Whipple's Disease remains one of the diseases of unknown etiology. Recent reports have indicated that steroid therapy may be of value in the treatment of this condition, heretofore regarded as uniformly fatal. That this therapy is not specific, and may often fail, has been recently reported.

This case report concerns the clinical and pathologic aspects of a typical example of Whipple's Disease. The diagnosis was definitely established, and steroid therapy was instituted with excellent results. The authors are well aware of the natural course of the disease, including the occurrence of spontaneous remission.

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CASE REPORT

This 60-year-old farmer was first seen on Jan. 16, 1956, complaining of headache, weakness, weight loss, diarrhea, abdominal pain, and arthritis. The onset was gradual, occurring about 18 months before he was seen. In the summer of 1954, he had suffered "severe arthritis" which was migratory and involved the hands, feet, and back. Pain in the back had continued intermittently. The headache had actually been of many years' duration, but had been worse since the onset of the presenting complaints. His bowel habits had been regular until about a year before he was seen, when diarrhea began. This consisted of two to three liquid stools daily for 3–4 days at a time. He denied the presence of foamy or fatty stools and of melena. The appetite had been poor; there was nausea but no vomiting. Belching, abdominal fullness and bloating were present. There had been a weight loss of 35–40 pounds. He complained of intermittent abdominal pain in the epigastrium which was cramping and radiated to the lower abdomen. It was severe, related to change in bowel habit, and often relieved by passing flatus, which was frequently difficult. Weakness and diarrhea had increased for 4–5 months before he was seen, and he had been confined to bed for about 8 weeks of that time. A chronic cough had been present for many years, with expectoration of mucus but no blood.

The family history was negative. There were no similar symptoms in four living siblings, nor was there any familial history of mental or metabolic disease.

On physical examination there was evidence of chronic illness and wasting. The patient's height was 62 inches; weight, 109 pounds. The temperature was normal; the pulse, 84; and the blood pressure, 100/70. The skin was dry, and there was no unusual pigmentation. The spine was stiffened and there was clubbing and cyanosis of the finger tips. There was no lymphadenopathy. The vital capacity was decreased; a pleural friction rub was heard in the left base, and signs of a pleural effusion were present in the right base. The heart was normal. The abdomen was tense, somewhat distended, and no masses were palpated. The spleen was not felt. Sigmoidoscopy findings were negative.

Laboratory study revealed a hypochromic anemia. The sedimentation rate was 25 mm. in 1 hour and the urine analysis revealed a normal specific gravity and an occasional hyaline cast. The EKG was normal. X-ray of the chest revealed the presence of a pleural effusion in the right base and a smaller amount of fluid in the left base.

FIRST HOSPITAL ADMISSION

The patient was admitted to the Methodist Hospital, Indianapolis, Ind., on Jan. 19, 1956. Laboratory studies revealed a hemoglobin concentration of 11.5 gm., a red blood cell count of 3,170,000, and a white blood cell count of 4450, with a normal differential. The urine was negative. Gastric analysis revealed a fasting free acidity of 12, with a rise to 14 after an alcohol meal. Sputum examinations were negative for acid-fast bacilli. One stool specimen, of many examined, yielded a positive test for occult blood. No stool fat studies were available, but observation revealed the yellow, greasy character of the stool. The NPN was 35 mg.%, the serum calcium was 4.1 mEq., and the total serum protein level was 5.11, of which the albumin was 3.25 and the globulin 1.86. An oral glucose tolerance test revealed a fasting blood sugar of 82 mg., with a rise to 120 mg. in 1 hour, 173 mg. in 2 hours, and to 159 in 3 hours.

X-ray of the gall bladder revealed fair visualization, and no stones. Barium enema findings were negative. Chest X-ray on January 23 showed resolution of the former