Mesenteric Manifestations of Weber-Christian Disease *

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In 1892, Pfeifer [7] described a patient who presented with tender and painful subcutaneous nodular masses of varying size scattered over the trunk and extremities. In 1916, Gilchrist and Ketron [4] had occasion to observe a similar case and were the first to call attention to the microscopic study of the subcutaneous nodules which demonstrated an ingestion of the fat cells by macrophages. Weber,[12] in 1925, referred to the condition as relapsing, nonsuppurative nodular panniculitis of the subcutaneous tissues. Christian,[2] in 1928, stressed the recurrent febrile episodes associated with the painful, inflamed nodules, and Brill [1] later referred to the entity as Weber-Christian disease.

Since these initial publications, numerous reports have appeared in the medical literature citing the clinical and microscopic findings incident to nodular subcutaneous panniculitis.[5, 9, 11]

In 1953, Steinberg [10] described for the first time the presence of perivisceral and intravisceral inflammatory fatty changes involving the tissues of various organs along with the association of subcutaneous nodular panniculitis. He was able to obtain detailed autopsy records of six cases, and in three of the six there were nodular fatty inflammatory changes involving the visceral mesentery and omentum. In addition, he cited two cases of his own, one of which demonstrated omental and mesenteric panniculitis at autopsy. The remaining patient presented inflammatory fat changes involving the myocardium, pancreas, spleen, liver, marrow and perirenal areas. In both of the latter cases there were nodular swellings also involving the subcutaneous tissues of the trunk and extremities. For lack of better descriptive terminology he referred to the wide-spread inflammatory fatty changes as systemic nodular panniculitis.

At the meeting of the Southern Surgical Association in 1959, Rives and Ogden [6] described seven cases, each of which was found at laparotomy to have involvement of the small bowel mesentery by a chronic nonspecific inflammatory process restricted to the adipose tissue of the mesentery. This group of patients was comprised of six men and one woman, whose ages ranged from 24 to 62 years. Each presented a diagnostic problem prior to operation, and each complained of moderate to severe abdominal pain of a recurrent nature. Nausea, fever, malaise and abdominal tenderness were prominent features among the group. Four of the seven patients presented with a palpable abdominal mass.

At the time of exploration the small bowel mesentery was found to be markedly thickened and presented as an irregular nodular mass in several instances. The involved mesentery varied from a reddish-brown to yellow color. These areas grossly resembled fat necrosis, with the mesentery being lobulated and puckered. The entire small bowel mesenteric root was involved in six cases; whereas, the changes were confined to the ileal mesentery in one instance. The small intestine itself was not involved

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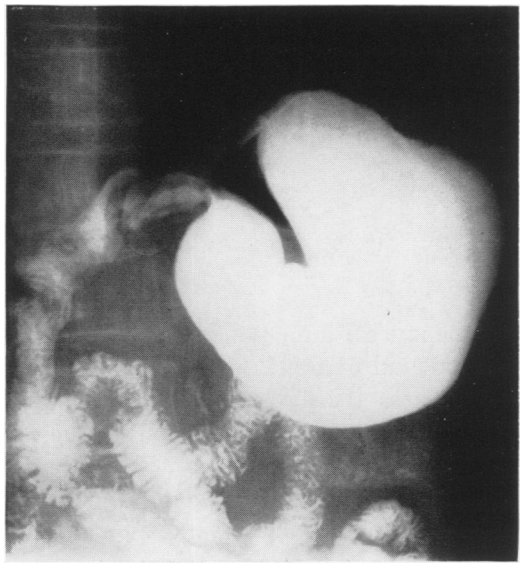

FIG. 1. X-ray showing the pressure defect on the pyloric portion of the stomach and the widened duodenal loop.

in the process. Biopsy of the mesenteric root in each case demonstrated microscopic changes identical with those observed in Weber-Christian disease. There was infiltration of the fat with macrophages, and the fat cells showed foamy cytoplasm. Macrophages were scattered about in bands, and collections of lymphocytes were present adjacent to the small blood vessels. Foreign-body giant cells, necrosis, and exudate were not present. The mesenteric blood vessels appeared normal. There was no involvement of the abdominal viscera or other perivisceral structures. Evidence of subcutaneous nodules was absent in each of the seven cases.

Our interest in this fascinating condition was aroused by the report of Rives and Ogden. Several months following their publication, we were fortunate enough to observe two strikingly similar cases within a period of several days. Both patients presented the identical clinical picture and gross surgical findings as they described. One showed microscopic pathologic changes involving the small bowel mesentery practically identical with their seven patients. The other case demonstrated somewhat a variant on microscopic pathological examination of the small bowel mesenteric root, but the findings were strikingly similar to their reported cases. Also, the pathologic process in both cases was confined to the mesentery of the small intestine, there being no evidence of associated subcutaneous tissue involvement.

Case Reports

Case 1. S. C. E. A 67-year-old man was admitted to the Vanderbilt University Hospital 10–27–60, with a 12-month history of vague generalized abdominal distress, anorexia and nausea. Weight loss had been minimal and there had been no melena or hematemesis. No fever or chills had occurred. Past history revealed episodes of pain in the dorsal and lumbar area of the spine, thought to be due to arthritic changes. The patient had suffered from coronary artery disease, with a history of two coronary attacks in the past. He had also suffered from a chronic urinary tract infection and had been on Mandelamine. On examination the blood pressure was 100/70. The skin was of normal texture and no abnormalities were present involving the skin or underlying subcutaneous tissues. There was mild diffuse tenderness over the upper abdomen, but no masses were palpable. The liver edge and spleen were not felt. Urinalysis showed a 2+ protein, 15 to 20 RBC, and 35 to 50 WBC per HPF. The WBC was 2,650 with 7% eosinophils. The PCV was 40, NPN 29, fasting sugar 77, and alkaline phosphatase 1.7 B.U. The serum amylase was 100 and serum bilirubin 0.4 mg.%.

An x-ray of the chest was normal except for eventration of the right hemidiaphragm. Barium enema was negative and IV urogram revealed reduplication of the left renal pelvis and ureter. Cholecystogram was normal. A GI series showed constant narrowing of the distal pyloric channel, but the mucosal pattern was intact. In addition, there was slight widening of the duodenal loop (Fig. 1). An EKG revealed abnormal T-waves suggestive of an old posterior wall infarction.

The pre-operative impression was possible chronic pancreatitis or a lesion of the antral portion of the stomach.

At operation the liver appeared normal. No free fluid was present in the abdomen. The entire stomach and duodenum were not remarkable. The spleen was small and showed evidence of perisplenitis. The entire root of the mesentery of the small bowel was indurated, thickened and pre-

sented as a lobular mass measuring approximately 15 × 15 cm. The overlying peritoneum was thickened and adherent to the underlying fat. The mass was a reddish-brown to yellow color and grossly resembled fat necrosis. The pancreas, kidneys, small and large intestine were normal. Several biopsies were obtained from the mass and the tissue appeared yellow on cut section. It was of a leathery consistency. The appendix was removed and the abdomen closed. The postoperative course was normal.

Pathologic Study. Microscopic section from the involved mesentery showed areas of fat necrosis with proliferation of connective tissue and accumulation of many chronic inflammatory cells and numerous foreign-body giant cells. In the giant cells and in the areas of fat necrosis were many cleft-like spaces resembling cholesterol clefts (Fig. 2).

Follow up. The patient has done fairly well since discharge from the hospital. However, leukopenia, along with a slight anemia, persists. He continues to experience intermittent nausea.

Case 2. W. M. P. A 48-year-old man was admitted to St. Thomas Hospital 10–28–60 with an 18-month history of abdominal discomfort. Transient nausea and vomiting had been present, but no anorexia or weight loss had occurred. He had un-

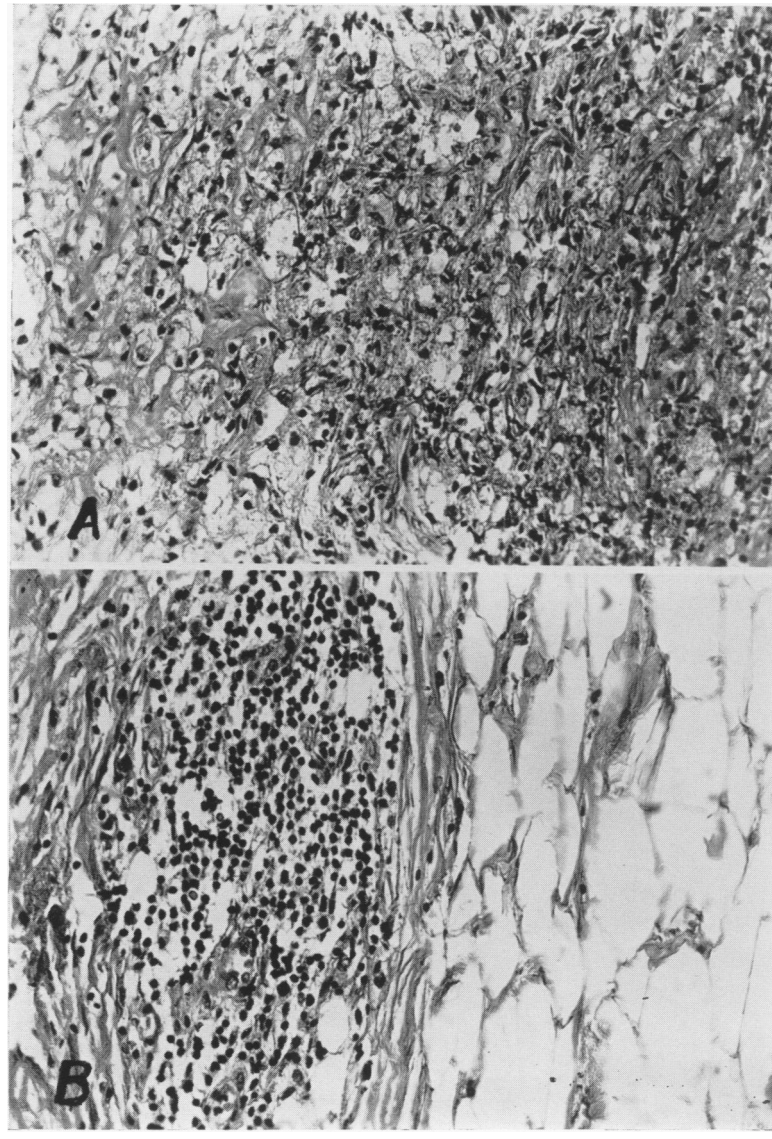

FIG. 2. A. Microscopic section showing fat with extensive areas of necrosis in which cleft-like spaces are prominent. B. Section showing the proliferating fibrous tissue with foreign-body giant cells and infiltrates of lymphocytes, macrophages, and lesser numbers of plasma cells.

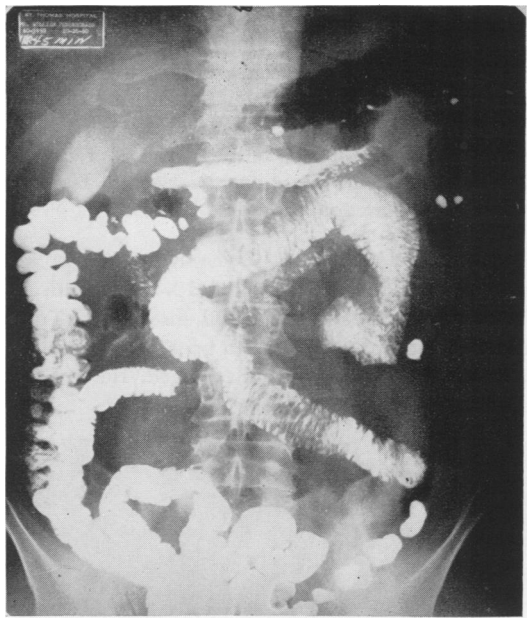

FIG. 3. Gastro-intestinal barium study showing the moderately dilated proximal jejunum.

dergone no previous abdominal surgery. No chills or fever had occurred. Examination revealed the skin and subcutaneous tissues to be normal. There was diffuse tenderness over the upper abdomen, but no masses or viscera were palpable. Blood counts were within normal limits. The alkaline phosphatase was 2.5 B.U. The cholesterol was 305 mg.%, and the blood sugar 102 mg. Bilirubin was not elevated and the BUN was 17.2. VDRL was nonreactive.

Barium enema, IV pyelogram and chest x-ray were normal. A GI series demonstrated prolonged retention of barium in a moderately dilated loop of jejunum in the left upper abdomen. No definite point of obstruction was noted (Fig. 3). The EKG showed abnormal T-waves.

At operation the liver and spleen were normal. No intrinsic lesion was noted in the small bowel, but the wall of the proximal jejunum was somewhat dilated for a distance of approximately 60 cm. Occupying the root of the small bowel mesentery, in the vicinity of the proximal jejunum, was a nodular, inflammatory-appearing mass measuring about 12 × 15 cm. The mass resembled fat necrosis and was colored a reddish-brown. The peritoneum was adherent to the underlying mass (Fig. 4). The small bowel showed no evidence of involvement. The pancreas appeared normal. Several biopsies were taken from the mass. An incidental finding was hemorrhagic infarction of an appendiceal epiploica of the sigmoid colon. This was removed, along with the appendix. The abdomen was then closed. The postoperative hospital course was normal.

Pathologic Report. Microscopic sections from the mesentery revealed evidence of fat necrosis. The wall of the small blood vessels showed congestion. Cellular infiltration was not a prominent feature (Fig. 5).

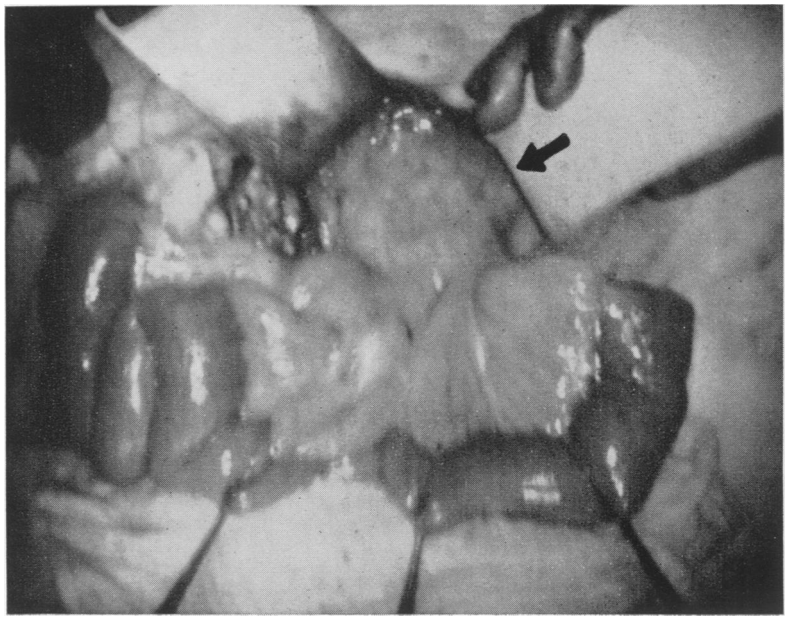

FIG. 4. Photograph at operation showing the chronic inflammatory mass occupying the root of the small bowel mesentery.

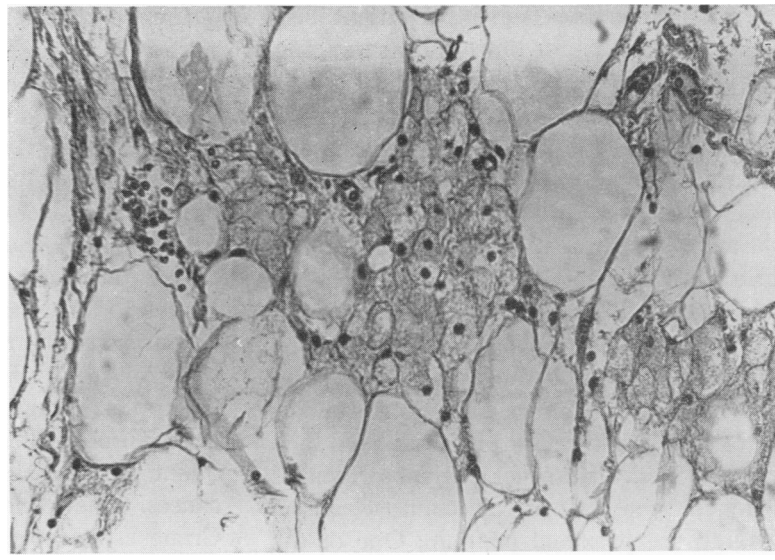

FIG. 5. Microscopic section showing adipose tissue with scattered focal infiltrates of macrophages with finely granular "ground glass" cytoplasm. Mild infiltrates of lymphocytes are also seen.

Follow up. Over a short follow up period, the patient has had no further abdominal pain. He has not received any medication since discharge.

Discussion

Steinberg described three phases in the microscopic maturation of the lesion in nodular panniculitis. The early lesion was characterized by the presence of edema, congestion and exudation between the fat cells. In the second phase, there was infiltration of lymphocytes and phagocytes within the fat cells, with the rare presence of giant cells. In the third phase, connective tissue replacement was observed, along with giant cells and lymphocytes. Evidence of periarteritis and intimal proliferation was also seen.

In the seven cases reported by Rives and Ogden, and in our two cases, the microscopic pathological findings closely paralleled those observed in Weber-Christian disease. No doubt, some variations existed in the microscopic pathology among these nine cases, but the essential features necessary to substantiate the diagnosis of mesenteric panniculitis was present in each case. The finding of foreign-body giant cells in our two cases, and their absence in the cases reported by Rives and Ogden, may be due to the fact that the sympatomatology in our two patients was of long standing, being 12 and 18 months, respectively, prior to operation.

The large blood vessels were uninvolved in each of the nine cases, but one of our patients showed congestion of the wall of the small blood vessels on microscopic study. Necrosis of the adipose tissue was present to some degree in our two cases, but suppuration was not found among the group of nine patients.

A leukopenia was recorded in two patients among Rives' series, and in one of the two personal cases observed by Steinberg. Likewise, one of our patients presented a persistent leukopenia with slight eosinophilia. The presence of recurrent bouts of fever has been a fairly constant feature accompanying panniculitis, but neither of our two cases demonstrated a temperature elevation prior to operation.

It is of interest that five of Rive's patients demonstrated additional pathologic changes elsewhere. One presented with a mass of hyperplastic lymph nodes superior to the pancreas, one an acutely inflamed appendix, another had focal hemorrhage in the omentum, and one presented with volvulus of

the small intestine. The fifth patient died following surgery for Martorell's syndrome.

It is of further interest that the entire group presented a real problem in preoperative diagnosis. The most common preoperative impression was chronic pancreatitis, pancreatic cyst, or gastro-intestinal neoplasm. Blood tests and blood chemical determinations have been of no aid in establishing a diagnosis.

The gastro-intestinal barium study showed abnormal findings in both of our cases. In one instance, a defect was noted in the distal gastric antrum and the duodenal loop was widened. The other patient presented delay in the transit of contrast media through the proximal jejunum. One of the cases cited by Rives likewise showed displacement of the stomach and descending duodenum on barium study. The overlying small intestine and omentum precluded palpation of the abdominal mass in both of our cases.

The etiology of this condition is entirely obscure. Various drugs, toxins and allergic reactions have at one time or other been thought to play a causative role in Weber-Christian's disease. In a discussion of Rives' paper Peete mentioned a case of mesenteric involvement which fulfilled the proper diagnostic criteria. His patient was of interest in that he had received severe trauma to the abdominal wall two years previously. He presented with an abdominal mass, and was thought initially to have an aortic aneurysm.

Obviously, the extent and the relationship of these mesenteric masses have rendered their removal impossible. Therefore, surgical treatment has been restricted to multiple biopsies. Surprisingly, both of our patients over a short term follow up have done quite satisfactorily. Six of the patients cited by Rives have remained asymptomatic; whereas, one patient has had cortisone and x-ray therapy for control of abdominal pain and pyrexia.

Crane,[3] referred to five patients who demonstrated at exploration massive fatty involvement of the small bowel mesentery. The clinical and gross surgical findings in his group were not altogether similar to the cases cited by Rives or to the present two cases under consideration. However, the microscopic pathologic changes in the small bowel mesentery did bear some resemblance to the cases cited above. There was alteration of the mesenteric fat cells, along with infiltrates of monocytes, lymphocytes, macrophages and rare foreign-body giant cells. Three of these cases underwent subsequent recovery and two died of unrelated causes. The condition was described by Crane as isolated lipodystrophy. More recently, Rogers [8] reported a similar case. It is of interest that fat necrosis was not a feature in the cases described by Crane or in the one patient cited by Rogers.

It is entirely possible that the cases referred to by Crane and Rogers may constitute a variant of true mesenteric panniculitis as described by Rives or they may represent the same condition but in a different stage of maturation. Very little is understood about this disease at present, and it is quite possible that it may be encountered in various stages, similar to the stages of systemic panniculitis as noted by Steinberg.

At the time of exploration, the involvement of the small bowel mesentery may present a problem in differential diagnosis. The gross findings may be confused with Whipple's disease, retroperitoneal lipomata, liposarcoma or lymphoma. A frozen section may be necessary for confirmation of diagnosis.

Apparently, chronic inflammatory adipose tissue changes involving the small bowel mesentery and without similar pathology in other abdominal organs or subcutaneous tissues constitute an idiopathic clinical entity. Mesenteric panniculitis appears to be an appropriate designation for this type of Weber-Christian disease and the recent

contribution by Rives has served to bring this clinical and pathologic entity to our attention. No doubt, other cases have been observed in the past but have not appeared in the literature.

Summary

Two cases which demonstrated nonspecific chronic inflammatory adipose tissue changes restricted to the root of the small bowel mesentery have been described.

The pathologic similarity of the gross and microscopic findings to that observed in Weber-Christian disease and to systemic nodular panniculitis as described by Steinberg have been emphasized.

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