

Sclerosing Mesenteritis Involving the Small Bowel and Large Bowel Mesentery: A Case Report and Review of the Literature

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Mesenteric Panniculitis/Sclerosing Mesenteritis is an under diagnosed, rare chronic fibrosing inflammatory disease that affects the mesenteric adipose tissue of the small intestine and rarely the mesocolon. The disease poses diagnostic challenge due to its non- specific clinical and radiologic findings. Biopsy and histopathologic evaluation is required where there is clinical or radiological suspicion of neoplasia. Management of Mesenteric Panniculitis / Sclerosing Mesenteritis depends on the presentation, with medical therapy preferred for symptomatic patients and surgery reserved for life threatening complications. We represent a case of a 71-year old man who presented with vague abdominal symptoms. Computed Tomography and Positron Emission scans demonstrated mesenteric lesions suspicious for carcinoid. A diagnosis of Mesenteric Panniculitis / Sclerosing Mesenteritis was made on histologic evaluation. The patient's symptoms on follow up did not warrant any therapeutic interventions till the writing of this article.

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INTRODUCTION

Sclerosing mesenteritis (SM) is a rare benign inflammatory and fibrotic disorder of unknown etiology, that primarily effects small bowel mesentery.¹⁻⁷ It was first described by Sulla et al in a case series in 1924 under the name, "refractile mesenteritis and mesenteric sclerosis".^{1,2,6} Subsequently many terms have been used to describe this entity based on the predominant histology within a spectrum of three histologic features that characterize this lesion, including fat necrosis, chronic inflammation and fibrosis.^{1,5} It can present as an isolated finding or in association with other disorders predisposing to inflammation and fibrosis as non- specific infection, autoimmune disorders, paraneoplastic syndrome, previous abdominal trauma or surgery.⁸ Abdominal computed tomography is the imaging modality of choice but has low specificity for sclerosing mesenteritis.⁴ Due to varied and nonspecific presentations and findings, the disease poses great diagnostic challenge leading to misdiagnosis in the majority of cases.⁵ The rarity of this condition has limited the ability to study demographic and clinical features, natural history and response to therapy. Here of the report case of a 71y Male who initially presented with abdominal pain and loose stools. An abdominal CT scan demonstrated multiple abdominal lesions, laparoscopic evaluation biopsy with bowel resection was

performed with suspicion of carcinoid tumor and a diagnosis of sclerosing mesenteritis was made on histopathology.

CASE REPORT

A 71year old male presented with complaints of nausea, vomiting, abdominal pain and loose stools. Other medical history included cutaneous vasculitis (leukocytoclastic vasculitis) of lower extremity and unspecified joint complaints, Coronary Artery Disease, Hyperlipidemia, Diabetes Mellitus with retinopathy and neuropathy, Hypertension, Iron deficiency anemia, and bilateral mixed hearing loss. After exploring his symptoms further, a CT scan was conducted which showed possible fibrotic and inflammatory changes in the omentum, transverse colon mesentery, cecal mesentery, terminal ileum mesentery and sigmoid colon mesentery. It also showed partially calcified mesenteric masses which lead to the suspicion of carcinoid tumor. Two hepatic foci were suspicious for metastatic disease (**Figure 1-2**). Following the CT scan a PET CT was conducted to assess for carcinoid tumor. The PET scan demonstrated some tracer uptake of a left paramedian mass that was less intense than what is usually seen in carcinoid tumor, other mesenteric calcified densities were not associated with abnormal tracer uptake. The patient's laboratory values were remarkable for an elevated ESR and chromogranin-A level, but his 5HIAA, serotonin and CEA levels were normal. This was followed by a colonoscopy and upper endoscopy which did no demonstrate masses that were amenable to

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biopsy. The patient subsequently underwent an open segmental resection of his sigmoid colon and the adjacent mesentery with mass. A frozen section of the mesenteric mass showed calcified hyalinized nodule with no evidence of carcinoma. Histopathology of mesenteric mass and sigmoid colon biopsy showed lobular areas of organizing fat necrosis variably surrounded by foci of loose storiform fibrosis and associated mild chronic lymphoplasmacytic inflammation (**Figure 3-5**). Immunohistochemical stains of CD138, IgG, IgG4 demonstrated few plasma cells with IgG: IgG4 < 15%, not supporting a diagnosis of IgG4 related disease. The histopathological features supported the diagnosis of sclerosing mesenteritis, a rare, non-neoplastic inflammatory and fibrotic disease affecting the mesentery. Sclerosing

mesenteritis can affect the integrity of the gastrointestinal lumen and mesenteric vessels by a mass effect. Post-surgical follow up CT scans (**Figure 6**) noted dilated loops of large bowel, an interval increases in the size of the left mid abdominal mesenteric calcified mass without change in other masses. The patient continued to experience some indigestion, vomiting, diarrhea and constipation but his symptoms were manageable and did not bother him. He was informed that if his symptoms were significant, he would need to start immunomodulatory drug therapy. The patient declined active management and has still not been started on any medical therapy related to sclerosing mesenteritis till the time of writing this report.

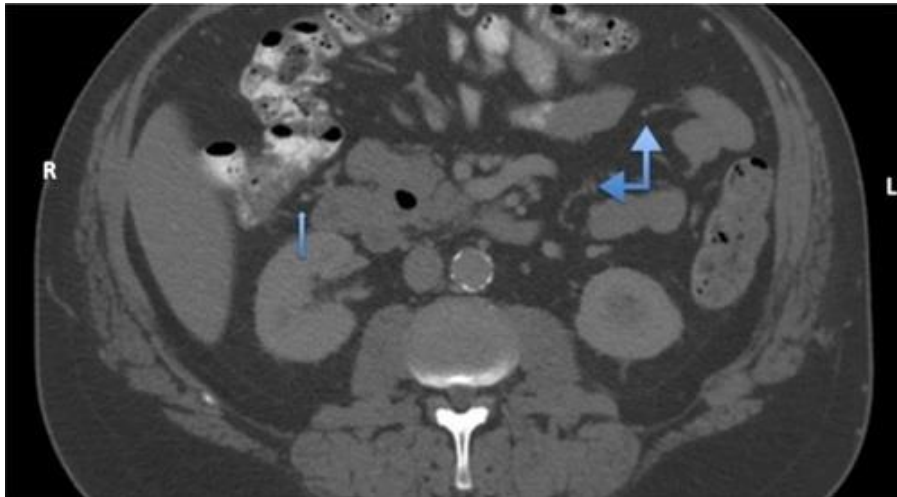


Figure 1. Five months prior to surgery signs of mesenteric panniculitis including halo sign ↑ and tumor pseudocapsule ↙ are seen. (CT with contrast).

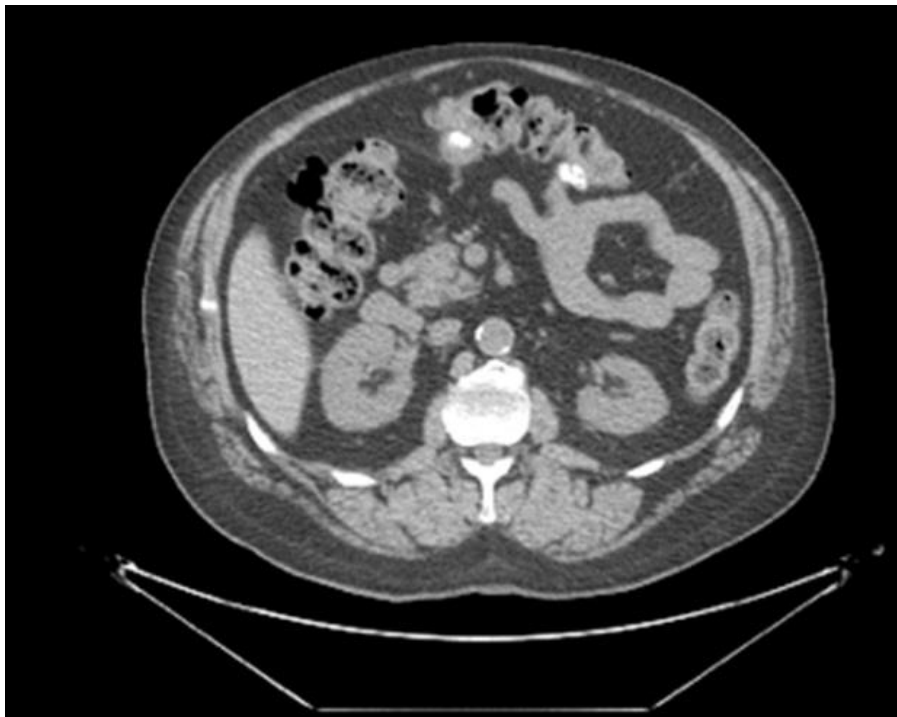


Figure 2. Two Months before surgery mesenteric masses are visible. (CT with contrast)

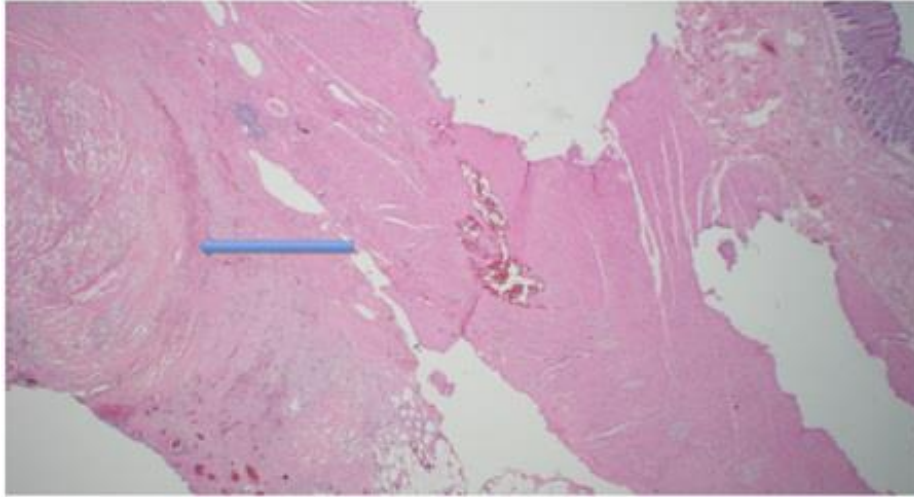



Figure 3. Unlike a Desmoid (mesenteric fibromatosis) the lesions of Sclerosing mesenteritis  involve the mesenteric adipose and do not arise from the muscularis propria of the bowel wall. (H&E x 200)

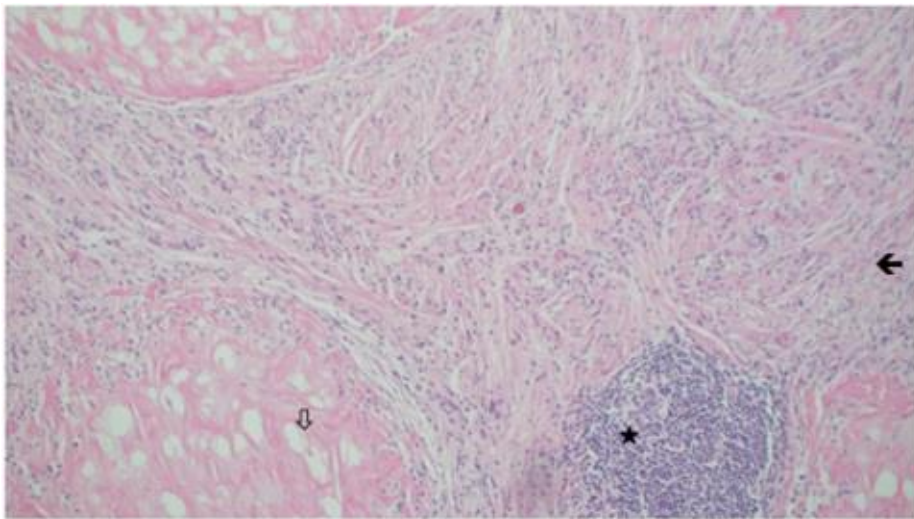




Figure 4. The triad of findings characteristic of sclerosing mesenteritis; Storiform fibrosis , organizing fat necrosis  and chronic inflammation*. (H&E X 200)

DISCUSSION

Sclerosing mesenteritis is a rare benign chronic inflammatory disorder of primarily small bowel mesentery.¹⁻⁷ Various terminologies have been used to describe the condition that include mesenteric lipodystrophy, retractile or liposclerotic mesenteritis, Weber-Christian disease, xanthogranulomatous mesenteritis, mesenteric lipogranuloma and systemic nodular panniculitis.^{5,7} Patients usually have a range of pathologic findings with varying components of chronic inflammation, fibrosis and fat necrosis, although usually one feature predominates at a given time and depending on the predominant histologic features the condition is referred to as panniculitis, retractile mesenteritis or mesenteric lipodystrophy, respectively.⁷⁻⁹ Three stages of histologic

progression have been described; the first stage shows mesenteric lipodystrophy with little or no acute inflammation; this stage is asymptomatic and has a good prognosis. The second stage of mesenteric panniculitis (MP) demonstrates plasma cells, a few neutrophils and foreign body type giant cell reaction with foamy macrophages. This stage usually presents with non-specific systemic symptoms as fever, abdominal pain and malaise. The final stage of retractile mesenteritis presents with scarring fibrosis, chronic inflammation and retraction of mesentery with formation of mesenteric masses and obstructive symptoms. All three components are present in most cases,⁷ as also seen in our patient. Diagnosis is often difficult with radiologic findings and biopsy and even surgical

excision maybe necessary in difficult and symptomatic cases where malignancy needs to be ruled out.

The prevalence of SM according to literature varies from 0.6-7.83%. True prevalence is actually estimated by some studies as 0.5-3%.^{11,14} It is most commonly seen in Caucasian males with the male to female ratio 2.3:1.^{10,11} It is usually diagnosed between ages of 3 and 88 with a median age of 65 years. The low prevalence in childhood and adolescents is attributable to smaller amounts of mesenteric fat.⁴

As mentioned above the clinical manifestations of Mesenteric Panniculitis / Sclerosing Mesenteritis (MP/SM) are largely nonspecific.^{12,13} The most frequent presenting symptoms are abdominal pain, bloating and distention, diarrhea and systemic

symptoms including fever malaise and weight loss. The presenting symptoms also depend on development of complications associated with SM which most commonly are bowel obstruction and obstructive uropathy / renal failure.^{4,10} Approximately 10-15% of patients are asymptomatic or have minimal symptoms. An abdominal mass can be palpated in about 35-50% of patients. Masses tend to be poorly defined and deep seated. Signs of peritoneal inflammation and ascites (usually chylous) are rare. Patient may also have symptoms of an underlying associated malignancy.^{11,14-16} Erythrocyte sedimentation rate and C-reactive protein level may be elevated in approximately 80% of patients. These or other laboratory abnormalities such as anemia and hypoalbuminemia are extremely nonspecific.⁴

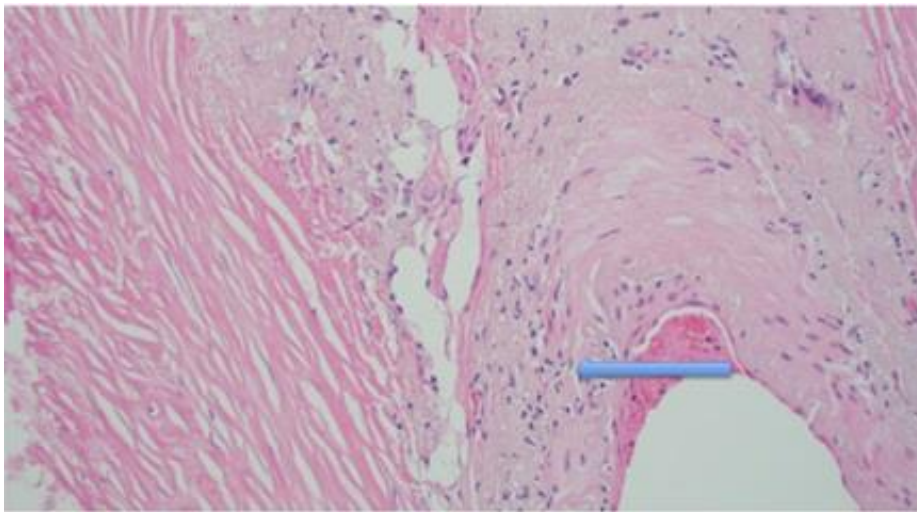


Figure 5. Spotty chronic phlebitis ← was seen. (H&E x 200)

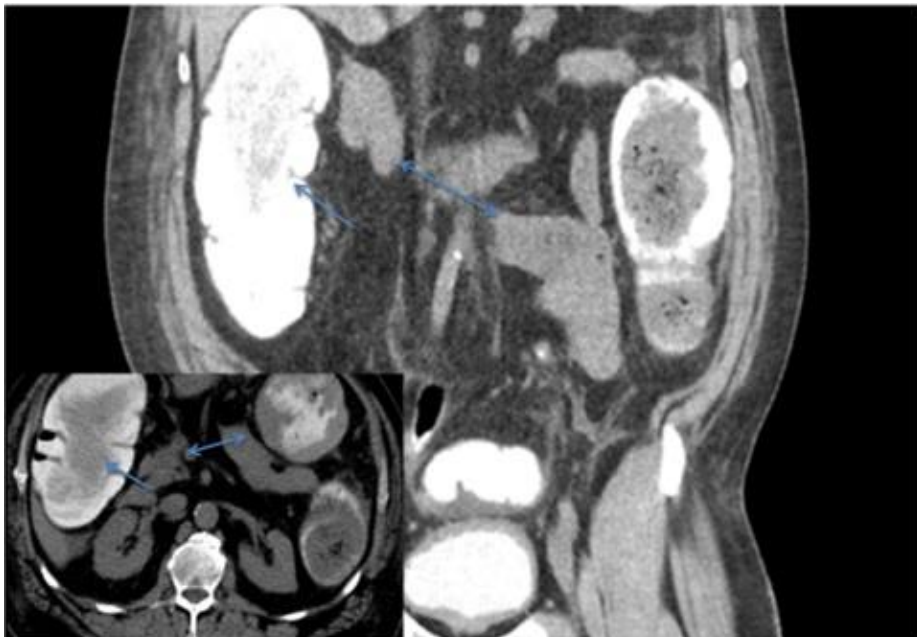


Figure 6. Some weeks post-surgical CT with contrast showing distended loops of bowel and persistent mesenteric masses.

The pathogenesis of SM is unclear however several hypotheses have been postulated. A large number of patients diagnosed with SM have a history of previous abdominal surgery or abdominal trauma and inflammation and it is hypothesized that these individuals are genetically predisposed to have abnormal healing and repair of connective tissue response to trauma.⁹ The use of powdered surgical gloves has also been implicated in the development of abdominal fibrosis as a precursor of SM.¹⁴ There have also been published reports of SM in association with immune mediated disorders such as vasculitis and fibrosing disorders as Riedel's thyroiditis, sclerosing cholangitis, orbital pseudo tumor or retroperitoneal fibrosis,^{4,17} hence clinical response to immunomodulatory medicines and glucocorticoids is frequently seen. Studies have suggested a possible role of IgG4 related immunopathologic processes in the development of SM,^{10,18} however these have not been validated. Some studies have demonstrated a characteristic histology associated with IgG4 -Related Disease, as dense lymphoplasmacytic infiltrate, with IgG4 to IgG ratio of approximately 40%, storiform fibrosis and obliterative phlebitis.^{4,6} Our patient also had a history of leukocytoclastic vasculitis and inflammatory arthropathy and IgG4 related disease was ruled out.

Case reports have also been published of sclerosing mesenteritis in patients with history of systemic or bowel infection such as Typhoid fever, Dysentery, tuberculosis, Syphilis, Malaria, influenza and Rheumatic fever.¹³ The suggested mechanism being bacterial endotoxins initiating intravascular coagulation in the mesenteric vessels with consequent mesenteric ischemia, causing angiomatoid proliferation of vessels and subsequent histologic changes of MP.¹⁹ A significant number of cases are however also described as idiopathic.²⁰

Many reports have looked at an association between MP/SM and malignancy. Earlier Studies explored the prevalence of malignant diseases in groups of patients identified with mesenteric panniculitis. Daskalogiannaki et al²¹ and others reported the coexistence of Mesenteric panniculitis with neoplastic diseases, most commonly Non-Hodgkins Lymphoma in particular follicular lymphoma^{18,22} and other solid tumors.^{2,11,22} However, the association between SM and neoplasia remains a subject of dispute. These studies are limited by selection bias due to the rarity of the disease, with most of the data being retrospective and also because diagnosis is mostly based on CT findings without a tissue diagnosis.¹¹ More recent studies^{1,14,22} with case control and match pair analysis using a large cohort of patients have found that contrary to previous reports, mesenteric SM is neither paraneoplastic nor is it associated with malignant diseases.

Since majority of lesions of SM are asymptomatic or present with non-specific symptoms, it is usually discovered incidentally. Dual phase abdominal computed tomography (CT) scan is the most sensitive imaging modality for detecting sclerosing mesenteries. A soft tissue mass in the root of the mesentery of the small bowel is present in about 60% of patients.⁹ Jejunal mesentery seems to be most commonly

involved although involvement of the mesocolon has also been described.¹⁷ Focal infiltration in the region of the pancreas may simulate pancreatic malignancy. Fat ring or Halo sign (halo sign describes the preservation of densitometric values of fat nearest the mesenteric vessels) and tumor pseudo capsule (which represents the finding of hyper attenuated stripe partly surrounding the mass) are two radiologic findings highly suggestive of MP/SM.^{4,5,15,20} Calcifications may be picked up in 20% of lesions in areas of fat necrosis. Misty Mesentery, a term used to describe the finding of increased attenuation of mesenteric fat with small lymph nodes but without evidence of a discrete mass is due to inflammation and edema and can be seen in various benign conditions including early stage MP/SM, mesenteric fibromatosis and mesenteric edema or in malignant conditions such as early stage or treated Lymphoma, carcinoid tumors or mesenteric carcinomatosis. Khasminsky et al and others^{7,11,23,24} used the size of mesenteric nodules (> 1 cm) to differentiate lymphoma versus fibrotic nodules of SM. Malignant versus benign can be further evaluated by positron emission tomography (PET) where carcinomatosis and high-grade lymphomas will have high FDG uptake, as will some inflammatory conditions. On the other hand, most benign processes including the fibrotic phase of MP/SM and sometimes lymphomas and carcinoids may have no or low uptake. In such cases where CT or Magnetic Resonant Imaging (MRI) scans are unable to differentiate Sclerosing Mesenteries from a malignant or infectious process, definitive diagnosis will depend on biopsy and histologic evaluation,⁵ as was resorted to in our patient. Another feature that can help differentiate malignant masses from MP/SM is that following treatment no change in the lesions of MP/SM is seen while malignant lesions show some response. This feature further refutes a positive association between MP/SM and malignancy.^{11,14}

The natural history of sclerosing mesenteritis is not well understood mainly due to the rarity of the condition. The management of Sclerosing mesenteritis is driven by the presence of symptomatic complications associated with the disorder or high suspicion of a malignant process that was not ruled out by radiologic findings. Biopsy findings maybe non-specific inflammatory and fibrotic changes alone⁵ and in such cases exploratory laparotomy is indicated to get a definitive diagnosis, as in our case. Approximately 20% of patients with Sclerosing mesenteritis develop complications^{9,10} and surgical therapy is attempted with primary goal of relief of life-threatening symptoms such as bowel ischemia and bowel obstruction and obstructive uropathy. Aggressive surgery may be difficult due to vascular compromise. Surgical management has not been shown to prevent progression of disease, therefore conservative measures to alleviate non-life-threatening symptoms are recommended. In cases that present without life threatening obstructive symptoms, medical therapy targeting the inflammatory phase of the disease is recommended including immunosuppressive agents as Steroids and tamoxifen, with non-responders given a trial of Azathioprine and cyclophosphamide. Once fibrotic masses form, medical therapy is no longer effective. Approximately 10-15% of patients are asymptomatic or have minimal symptoms²¹ and do

not require treatment.

CONCLUSION

Sclerosing mesenteritis is a rare benign chronic inflammatory disease which poses a diagnostic challenge for surgeons, radiologists and pathologists. Invasive and more aggressive surgical procedures are sometimes necessary to rule out malignancy in radiologically ambiguous cases. The pathogenesis, natural course and therefore management of the disorder is not well understood due to the rarity of the disease and variable course. Therapeutic decisions are therefore based on individual presentation and primarily to rule out malignancy.

CONFLICT OF INTEREST

The author has no conflict of interest to disclose.

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