Rare Complication of Eosinophilic Granulomatosis with Polyangiitis: A Case Report

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Abstract

Mesenteric panniculitis (MP) is a rare non-neoplastic inflammatory and fibrotic disease that affects the adipose tissue of the mesentery of the small intestine. The specific etiology of the disease is unknown. The diagnosis is suggested by the computed tomography (CT). We reported a case of the mesenteric panniculitis diagnosed by CT as a complication of Churg-Strauss syndrome. MP can present as asymptomatic or as a variety of gastrointestinal and systemic manifestations, including abdominal pain, nausea and vomiting, diarrhea, weight loss and fever.

Keywords: Mesenteric panniculitis, Churg-Strauss syndrome, Complication, Diagnosis, Treatment.

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1. Introduction

Mesenteric panniculitis (MP) is a rare non-neoplastic inflammatory and fibrotic disease that affects the adipose tissue of the mesentery of the small intestine. The objective of this case report is to present the patient with mesenteric panniculitis developing 12 years after the diagnosis of Churg-Strauss syndrome. The specific etiology of the disease is yet unclear. Mesenteric panniculitis has been connected to a variety of conditions like vasculitis, granulomatous disease, malignancies and pancreatitis. Clinical manifestations are nonspecific and atypical. The disease is often asymptomatic. The most common clinical presentations include abdominal pain, vomiting, diarrhea, constipation and palpable abdominal mass or intestinal obstruction. Laboratory parameters are usually normal; however, an increased erythrocyte sedimentation rate has been noted. Abdominal CT is the most sensitive imaging modality for detecting MP, but the definite diagnosis of mesenteric panniculitis is established by biopsy. The prognosis is good in most patients and the outcome of the disease is usually benign.

2. Case Report

A 61 years American patient has been admitted to our hospital with 18 days history of severe generalized abdominal pain, which is constant and dull in nature. The patient had constipation with passage of flatus. Erythematous skin lesion in both the lower limbs and both hands were observed alongside with a painless mass in the right cheek for two months duration. He had numbness of the legs below the knees and painful swelling in the right ankle. His past medical history included the diagnosis with Churg-Strauss syndrome since 2006. Upon admission, the patient was conscious, alert and well oriented. His physical examination revealed a painless swelling in the right cheek 5 by 6 cm with normal skin color, not adherent to the bone and no lymph node enlargement. A palpable purpura of both hands and lower legs below the knees and a crusting necrotic, painful lesion over the extensor surface of the left elbow. Painful distended abdomen with the fullness of both flanks. Tenderness in joint line of the right ankle, but there is no restriction of movement.

The results for chest x-ray and the abdomen erect and supine were normal (Figures 1,2 and 3).



Figure 1: Normal chest X-ray



Figure 2: Normal Erect Abdominal X-ray



Figure 3: Normal Supine Abdominal X-ray

Ultrasound was normal apart from an incidental finding of a hemangioma of the liver DEXA score -3.2

Computed tomography (CT) for the abdomen and pelvis revealed haziness and strand-like infiltration of mesenteric fat planes and peritoneal reflections extending also to the peri-aortic, retroperitoneal and pelvic regions obscuring but not displacing or obstructing several bowel loops and visceral blood vessels (misty mesentery sign). (Figure 4)



Figure 4: CT abdomen and pelvic shows misty mesentery sign

There were no enlarged mesenteric, pelvic or retroperitoneal lymph nodes and there were few scattered colonic diverticula on the right and left. No other GI lesions or ascites were observed. MRI of the face showed ill-defined swelling and edema of the subcutaneous tissues as T2, T1, low signal intensity highlighted on STIR images, in the fat planes with good evidence to suggest the diagnosis of subcutaneous panniculitis-like infiltration of the right side of the face. (Figure 5)



Figure 5: MRI of the face showing subcutaneous panniculitis-like infiltration of the right side.

3. Discussion

Mesenteric panniculitis is a chronic and non-specific inflammation of the adipose tissue of the intestinal mesentery. In over 90% of cases, mesenteric panniculitis involves the small-bowel mesentery. Mesenteric panniculitis is a rare condition. There is no clear information about the incidence of mesenteric panniculitis in the literature. The disease has been associated with a variety of conditions such as vasculitis, granulomatous disease, autoimmune disorder, rheumatic disease, malignancies, pancreatitis, hyper-sensitivity reactions, ischemia, bacterial infection and abdominal trauma or surgery. The exact diagnosis is made by looking at the three pathologic findings: fibrosis, chronic inflammation, fatty infiltration of the mesentery. These three components can be found in various proportions. Mesenteric panniculitis is usually asymptomatic and often incidental. When symptomatic, patients may present with abdominal tenderness or a palpable abdominal mass and systemic manifestations, including abdominal pain, pyrexia, weight loss, etc. Symptoms may be progressive, intermittent, or absent. The mean clinical progression is from 2 weeks to 16 years. CT features of the disease are considered somewhat specific for this disorder. A definitive diagnosis is biopsy, but open biopsy is not always necessary for diagnosis. Recently mesenteric panniculitis has been diagnosed using CT features of the disease. Mesenteric panniculitis resolves spontaneously; however, treatment has been reserved for symptomatic cases. Some drugs are useful to medical treatment such as steroids, thalidomide, cyclophosphamide, progesterone, colchicine, azathioprine, tamoxifen, antibiotics and emetine. Surgical resection is sometimes attempted for definitive therapy and in cases of intestinal obstruction and other complications, such as ischemia.

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References

- [1] Issa, I., Baydoun, H. (2009). Mesenteric panniculitis: various presentations and treatment regimens. World J Gastroenterol, 15:3827-30.
- [2] Kipfer, R.E., Moertel, C.G., Dahlin, D.C. (1974). Mesenteric lypodystrophy. Ann Intern Med, 80:583-8.
- [3] Ferrari, T.C., Couto, C.M., Vilaça, T.S. (2008). An unusual presentation of mesenteric panniculitis. Clinics (Sao Paulo). 63:843-4.
- [4] Emory, T.S., Monihan, J.M., Carr, N.J. (1997). Sclerosing Mesenteritis, Mesenteric Panniculitis and Mesenteric Lipodystrophy, A Single Entity. Am J Surg Pathol. 21:392-98.
- [5] Akram, S., Pardi, D.S., Schaffner, J.A. (2007). Sclerosing mesenteritis: clinical features, treatment, and outcome in ninety-two patients. Clinic Gastroenterol Hepatol. 5:589-96.
- [6] McCrystal, D.J., O'Loughlin, B.S., Samaratunga, H. (1998) Mesenteric panniculitis, a mimic of malignancy. Aust NZJ Surg. 68:237-239.
- [7] Daskalogiannaki, M., Voloudaki, A., Prassopoulos, P., et al. (2000). CT evaluation of mesenteric panniculitis: prevalence and associated diseases. Am J Roentgenol. 174:427-31.
- [8] Delgado, Plasencia L., Rodríguez, Ballester L., López, et al. (2007). Mesenteric panniculitis: experience in our center. Rev Esp Enferm Dig. 99:291-297.
- [9] Mata, JM, Inaraja, L, Martin, J, et al. (1987). CT features of mesenteric panniculitis. J Comput Assist Tomogr. 11:1021-3.
- [10] Seo, M., Okada, M., Okina, S. (2001). S. Mesenteric panniculitis of the colon with obstruction of the inferior mesenteric vein: report of a case. Dis Colon Rectum. 44:885-889.
- [11] Ginsburg, P.M., Ehrenpreis, E.D. (2002). A pilot study of thalidomide for patients with symptomatic mesenteric panniculitis. Aliment Pharmacol Ther. 16:2115.
- [12] Iwanicki-Caron I., Savoye G. (2006). Successful management of symptoms of steroid-dependent mesenteric panniculitis with colchicine. Dig Dis Sci. 51:1245.