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Direttore Nicola Picardi

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Paolo Panaccio*/**, Manuela Bellobono*, Ludovica Esposito*, Marco Ricciardiello*, Sebastiano Cinalli*, Roberto Cotellese*

**Department of Medical and Oral Sciences and Biotechnologies, University of G. D'Annunzio, Chieti, Italy*
***Unit of Surgical Oncology, "Annunziata", Chieti, Italy*

Peritoneal sarcoidosis mimicking peritoneal carcinomatosis

AIM: *Describtion of a rare case of intestinal obstruction due to peritoneal sarcoidosis mimicking peritoneal carcinomatosis, and of the literature review about its surgical management.*
MATERIAL OF STUDY: *A 69 year-old woman was referred to our emergency department with diffuse abdominal pain, enteric vomiting and constipation. Abdominal CT showed a dilatated small bowel loops extended to the jejunum with a mechanical obstruction. Given the failure of a non operative management, an explorative laparotomy was performed.*
RESULTS: *Intraoperative evaluation showed an omental cake with extensive adhesions between small bowel and abdominal wall. The adhesion band determining occlusion was identified and cut. Furthermore, several peritoneal and omental biopsies were performed. Postoperative period was uneventful. Unfortunately, one month later, the patient died following an episode of spontaneous pneumothorax and respiratory complications.*
DISCUSSION: *Sarcoidosis is a chronic multisystemic disorder of unknown aetiology with granulomatous inflammation. Peritoneal involvement is a rare presentation of sarcoidosis. Clinical presentation depends on the extent of organ involvement. In some cases,symptoms are no specific and uncommon findings have been reported so far.*
CONCLUSION: *Despite a peritoneal carcinomatosis was suspeted, this case shows that abdominal sarcoidosis might be considered as a differential diagnosis when a lesion suspected of being peritoneal carcinomatosis shows non-typical clinical presentations.*

KEY WORDS: Peritoneal sarcoidosis, Small bowel obstruction

Introduction

Sarcoidosis is a disease involving abnormal collections of inflammatory cells that form lumps known as granulomas. The disease usually begins in the lungs, skin, or lymph nodes. Less commonly affected are the eyes, liver, heart, and brain. Any organ, however, can be affected. Clinical and radiological presentation varies depending on the organ involvment. We present a rare case of intestinal obstruction due to peritoneal sarcoidosis mimicking peritoneal carcinomatosi providing a literature review about its surgical management.

Case Report

A 69-years-old female Caucasian patient arrived at our emergency department with acute abdomen. Physical examination showed: a temperature of 37.2°C, diffuse abdominal pain, bloating, several episodes of enteric vomiting and constipation. The patient's medical records included sequelae of poliomyelitis, previous endoscopic removal of uterine myomas, no previous surgical operations. Laboratory reports showed: white blood cell count

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Correspondence to: Paolo Panaccio, MD, Department of Medical and Oral Sciences and Biotechnologies, University of G. D'Annunzio, Chieti and Pescara, Chieti, Italy - Unit of Surgical Oncology, Ss Annunziata, Chieti, Italy (e-mail: paolo.panaccio@gmail.com)

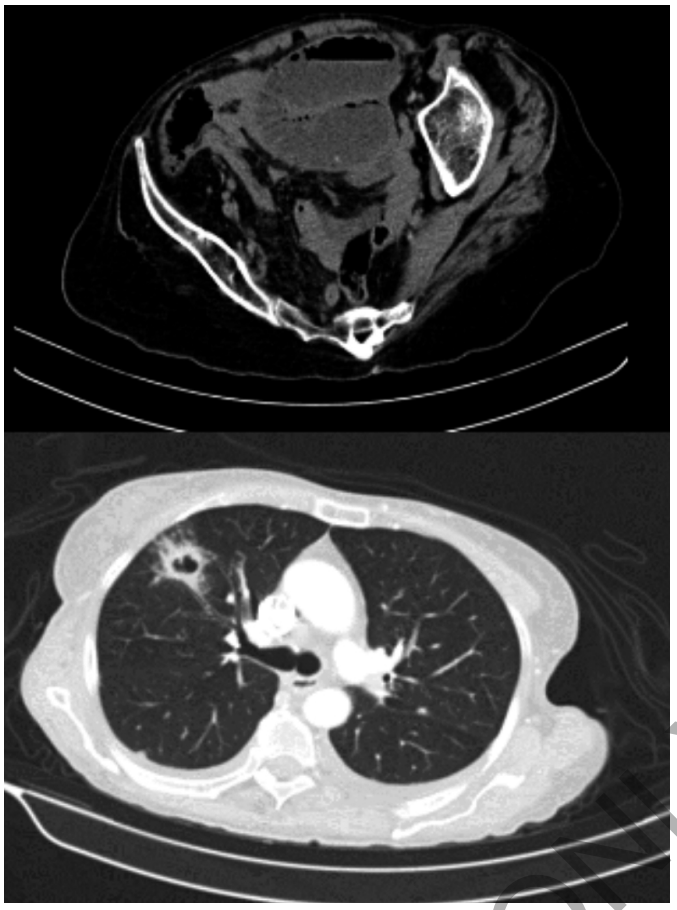

Fig. 1: Abdominal + Chest CT scan: dilated bowel loops to the jejunum and excavated consolidation areas.

10.04×103/uL (neutrophil 88.1%); creatinine 2.1 mg/dL. The other values were normal. Abdominal CT showed a dilated small jejunal loops, compatible with a mechanical small bowel obstruction; there were no pelvic masses or free fluid. Chest CT revealed several bilateral airspaces with consolidation areas, some of them excavated, and multiple micronodular opacities with mediastinal and hilar adenopathy (Fig. 1).

Results

Given the failure of a non-operative management, an explorative laparotomy was performed. Intraoperative evaluation showed an omental cake with extensive adhesions between small bowel and abdominal wall. The adhesion band determining occlusion was identified and cut. Furthermore, nodules biopsies and partial omental resection were performed. Macroscopically the most likely diagnosis appeared to be peritoneal carcinomatosis or tuberculosis (Fig. 2). The morphological appearance and histochemistry revealed epithelioid noncaseating granulomatous disease consistent with peritoneal sarcoidosis (Fig. 3). The postoperative course was uneventful. One month later, the patient returned to the emergency department with diagnosis of right pneumothorax and died a week later for the pulmonary complications.

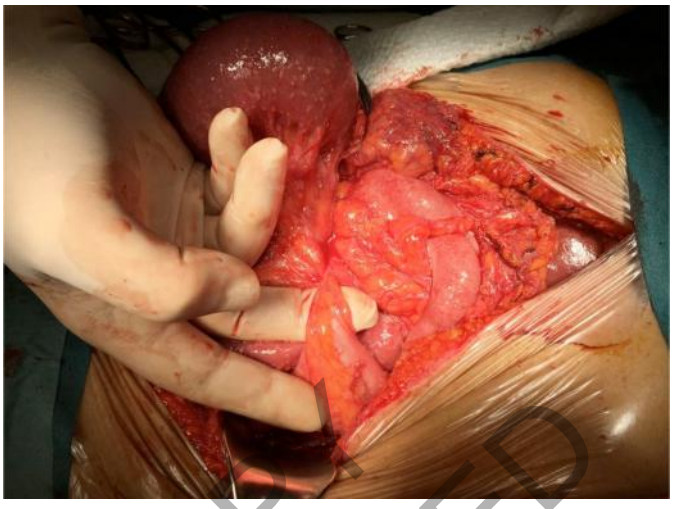

Fig. 2: Intraoperative findings: single band adhesion determining occlusion and peritoneal nodules.

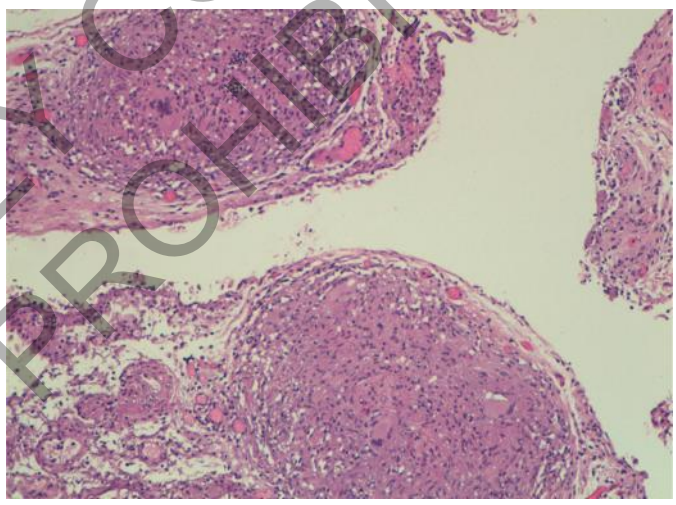

Fig. 3: Slide stained with ematoxylin, 10x magnification. Granulomas composed of aggregates of tightly clustered epithelioid cells with many Langhans-type and foreign-body-giant cells without central necrosis.

Discussion

Sarcoidosis is a chronic multisystemic disorder of unknown aetiology characterized by a noncaseating granulomatous inflammation. It affects people worldwide with a prevalence of about 4.7-64 per 100.000. Pulmonary involvement with dyspnoea and cough is demonstrated in approximately 90% of patients and it represents the principal cause of morbidity and mortality in patients affected by sarcoidosis. Gastrointestinal (GI) involvement is quite rare and it counts less than

1% of cases, but the incidence of subclinical involvement may be much higher. However, extra thoracic localizations may occur in absence of intrathoracic disease. Other tissues commonly involved are skin, eyes, reticuloendothelial system, musculoskeletal system, exocrine glands, heart, kidney and central nervous system. Sarcoidosis rarely affects the peritoneum. The first case of peritoneal sarcoidosis was published in 1954[1]. The most frequent clinical presentation is abdominal pain caused by granulomatous peritoneal nodules with bloody or non-bloody exudative ascites, but often it is asymptomatic. The onset of the pathology with abdominal complications without pulmonary is quite rare[2]. Generally, the diagnosis of peritoneal sarcoidosis is established when clinical and radiographic findings are supported by typical histopathological findings in peritoneum. Noncaseating granulomatous inflammation and other causes of granulomas, such as infectious disease (e.g., tuberculosis, histoplasmosis, brucellosis and toxoplasmosis), autoimmune disorders (e.g., Wegener's granulomatosis, primary biliary cirrhosis and Crohn's disease), occupational and environmental exposures (e.g., beryllium, talc and fungi), delayed-type hypersensitivity to foreign agents and neoplasia should be excluded[3]. Sarcoidosis is best defined in histopathological term as a disease characterized by the presence in all of affected tissues of non-caseating epithelioid cells granulomas. In contrast to the granulomatous reaction formed in response to infectious disease, a sparse population of lymphocytes in the mantle zone around the granuloma is hallmark [4]. Laboratory evaluation is often unrevealing, peripheral lymphopenia with CD4 depletion, hypercalcemia and hypercalciuria can be found. Elevated CA125 levels have been noted in some cases of peritoneal sarcoidosis, but it's not well known whether CA125 would be a reliable marker for sarcoidosis activity [5]. Despite CT scan doesn't have a high specificity, it represents an excellent method to identify the normal anatomy of the peritoneum and to detect and characterize a peritoneal disease [6]. Radiologic descriptions of abdominal sarcoidosis are very rarely reported. The most common imaging manifestations of abdominal sarcoidosis are hepatomegaly, lymph nodes and spleen involvement. In our case, CT showed neither peritoneal nodules nor omental cake. The main radiologic feature was the mechanical adhesive small bowel obstruction and an operative approach was necessary due the strangulated bowel. Most cases of peritoneal sarcoidosis usually have a benign course, evolving spontaneously or with a short course of drug therapy. Corticosteroids and immunosuppressive agents represent the therapy of choice. Prednisone is administered 1mg/kg, for four to six weeks, followed by a slow taper over two or three months [3]. In case of peritoneal sarcoidosis in which there is no gastrointestinal complications (e.g. obstruction or perforation) medical treatment with corticosteroids is useful for clinical resolution.

Conclusions

Peritoneal sarcoidosis should also be considered in the peritoneal disease diagnostic workup. Laparotomy or laparoscopy is often useful to confirm diagnosis and disease's extension.

Riassunto

La sarcoidosi è una malattia multiorgano caratterizzata dalla presenza di granulomi non caseosi. L'interessamento polmonare con dispnea e tosse è presente in circa il 90% dei pazienti e rappresenta pertanto la principale causa di mortalità e morbidità. L'interessamento gastrointestinale è raro e rappresenta circa l'1% di tutti i casi di sarcoidosis, ma il coinvolgimento subclinico sembra essere molto più frequente. La sarcoidosi peritoneale è stata raramente descritta ed il primo caso risale al 1954. Le manifestazioni cliniche principali sono il dolore addominale, l'occlusione intestinale con o senza essudazione ascitica. Riportiamo il caso di una donna di 69 anni, con anamnesi patologica remota e prossima silente per patologie polmonari, che giunge alla nostra osservazione con un quadro di addome acuto, diffusa dolenzia e alvo chiuso a feci e gas. Il quadro TC addome ha messo in evidenza la presenta di anse digiunali distese e con livelli idroaerei in assenza di masse pelviche o fluido libero in cavità peritoneale. L'esame TC del torace ha segnalato la presenza di alcune aree di consolidazione, talune escavate, e multiple nodularità con adenopatia mediastinica. A causa del fallimento del trattamento conservativo, la paziente è stata sottoposta ad una laparotomia esplorativa che ha evidenziato un quadro occlusivo sostenuto da sindrome aderenziale complessa e dalla presenza di multiple nodularità peritoneali sulla parete addominale e sul grembiule omentale che si è provveduto a bioptizzare. Il decorso post operatorio è risultato regolare con la dimissione della paziente in buone condizioni cliniche. Sfortunatamente la paziente è deceduta a distanza di un mese per le sopraggiunte complicanze respiratorie dopo un episodio di pneumotorace spontaneo. La tipizzazione istopatologica ha messo in evidenza la presenza di granulomi non caseosi a componente epitelioide compatibile con sarcoidosi. In considerazione dell'aspetto intraoperatorio sospetto per carcinosi peritoneale a partenza da organi pelvici o extrapelvici, la sarcoidosi peritoneale dovrebbe essere sempre posta in diagnosi differenziale e pertanto considerata nel workup diagnostico della carcinosi peritoneale.

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