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Inflammatory Myofibroblastic Tumor of the Spleen: A Report of Two Cases

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Authors' contributions

This work was carried out in collaboration among all authors. Author VKB was involved in data collection and preparation of manuscript. Authors VI and SJA were involved in literature searches and proofreading of the manuscript. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Owing to the lack of typical clinical features and imaging characteristics, inflammatory myofibroblastic tumors (IMT) of spleen can cause diagnostic dilemmas. Here, we report two such cases of splenic IMT. Our first patient was a lady who was detected to have an incidentally splenic lesion characterized as angiosarcoma on CT scan. Splenectomy was done, Immunohistochemistry (IHC) was suggestive of IMT. After four years, follow-up imaging revealed a lesion in the liver, core biopsy was suggestive of metastasis from IMT. She was started on steroids and is on follow-up. Our second patient was an 83 year old gentleman who was detected to have adenocarcinoma sigmoid colon. CT scan revealed a splenic lesion, suggestive of lymphoma or metastasis. Anterior resection and splenectomy was done. IHC of splenic lesion was suggestive of IMT. He is asymptomatic at 9 months follow-up. The two cases of splenic IMT presented us with different challenges in management. In the first case, the patient developed a metachronous lesion in the liver four years after splenectomy, which is a rare occurrence as per literature. In the second case, the co-existence of splenic IMT with adenocarcinoma colon led to suspicion of metastatic disease, we could not find any similar case reported in literature. Thus, although splenic IMT is rare, it can cause significant diagnostic and therapeutic challenges. Surgery is mostly curative, but follow-up is essential in view of possibility of local recurrence and metastasis.

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1. INTRODUCTION

The terminology of inflammatory myofibroblastic tumor (IMT) was first officially used in 2002, when the World Health Organisation (WHO) included this entity in the classification of tumors of soft tissue and bones [1]. Prior to this, a number of terms including inflammatory pseudotumor, plasma cell granuloma and plasma cell pseudotumor were used to describe this entity. IMT was classified as intermediate malignant tumor with less than 5% risk of metastasis [1], as opposed to initial reports which often classified it as a benign tumor. The exact etiology and pathogenesis is unclear. The proposed theories include infections, autoimmune cause, vascular pathology, trauma, neoplastic process and viruses such as Epstein Barr virus [2,3,4].

IMT can affect any age group, and a slight female preponderance has been reported [5]. Initially described in the lung [6], a number of extrapulmonary anatomical sites have been subsequently reported, including the spleen [7]. Histologically, these tumors are composed of proliferation of spindle cells, the myofibroblasts, with variable inflammatory cell infiltrates composed of lymphocytes, plasma cells, histiocytes and granulocytes [8,9,10]. Generally, these tumors are considered to have low to intermediate malignant potential, with relatively low rates of local recurrence and metastasis, although cases with high malignant potential have also been reported [11].

The diagnosis of IMT of the spleen can be challenging. Clinically, the patient may be totally asymptomatic, or may have nonspecific symptoms, as was the case with both the patients in our study. The clinical presentation may include non-specific left upper quadrant pain, anaemia, weight loss, low grade fever and splenomegaly, however, these symptoms are not specific to IMT [12]. The imaging modalities used for characterization of these lesions include USG, CT, MRI and PET scan. IMT usually present as a well circumscribed single lesion on cross sectional imaging, a central satellite area corresponding to a fibrous plaque may be seen on contrast enhanced images [13,14]. MRI demonstrates low to iso-intensity on T1-weighted imaging, and high intensity with surrounding low intensity on T2-weighted images [14]. PET uptake is variable, occasional high uptake may be seen [15]. However, no typical findings or definite diagnostic criteria have been established which allows differentiation from other lesions, thereby making misdiagnosis a very common occurrence [14,16]. Core biopsy can yield the diagnosis, especially when combined with immunohistochemistry; but this can be a difficult procedure in the spleen especially when the findings on imaging are atypical. Hence, histopathology of the resected specimen often remains the only reliable gold standard modality to establish the diagnosis [15].

2. PRESENTATION OF CASE

- **1.** Mrs. X, 78 year old lady was referred to us with a space occupying lesion in the spleen detected on ultrasonogram (USG) when she was worked up for anaemia by a physician. Contrast Enhanced CT (CECT) scan of the abdomen revealed a 12.7 x 9 cm mixed density calcified mass in the spleen which showed intense peripheral enhancement in the arterial phase and minimal centripetal enhancement in the venous and delayed phase (Fig. 1, 2). Large necrotic component was present, CT suggested a differential diagnosis of angiosarcoma and hemangioendothelioma. She underwent splenectomy. Histopathology showed a well encapsulated tumor composed of irregularly oriented bland spindle shaped cells with intermixed with lymphocytes, plasma cells and occasional neutrophils. Immunohistochemistry (IHC) revealed that the spindle cells were positive for Vimentin and smooth muscle actin, and the lymphocytes were positive for CD3 or CD20; other markers were negative. Final IHC report was suggestive of inflammatory myofibroblastic tumor. She had an uneventful postoperative recovery and was asymptomatic. On a routine follow-up USG four years after surgery, she was detected to have a liver lesion. Triple phase CECT abdomen showed a 5.5 x 4.5cm hyperdense lesion in segment 4a,4b showing moderate patchy enhancement in the arterial and venous phase with contrast washout in the delayed phase (Fig. 3, 4); CT differential diagnosis being metastasis and hepatocellular carcinoma. CT guided core biopsy was done. Histopathology showed

tissue fragments composed of spindle shaped cells with diffuse infiltration by medium sized lymphoid cells and plasma cells. IHC was suggestive of inflammatory myofibroblastic tumor. The case was discussed in the Multidisciplinary Team (MDT) meeting, and the options of treatment including surgery were explained to the patient and bystanders. After discussion, a conservative line of management was adopted. Hence she was started on oral steroids. Currently after one year of follow-up, she is asymptomatic, USG showed the lesion was regressing, present size being 3.9 x 3.3 cm.

- **2.** Mr. Y, 83 year old gentleman, presented with complaints of constipation and bleeding per rectum. Colonoscopy revealed a growth in the sigmoid colon, biopsy was suggestive of adenocarcinoma. Staging CECT revealed a heterogeneously enhancing lesion in the spleen measuring 8.5 x 6.5cm and a wall thickening in the sigmoid colon with adjacent mesenteric fat stranding (Fig. 5, 6). There were no liver lesions, CECT reported the splenic lesion as ? Lymphoma / ? metastasis to spleen. PET CT showed FDG uptake in the colonic lesion (SUV Max58.3) and in the splenic lesion (SUV Max 9.6), the report suggested suspicion of primary colonic malignancy with the splenic lesion as lymphoma or metastasis (Fig. 7, 8). Anterior resection with splenectomy was performed. Histopathology of the colonic lesion was reported as adenocarcinoma, T3 N0. Sections from the spleen showed a partly encapsulated neoplasm composed of ovoid to spindle cells arranged in diffuse sheets, with an admixture of abundant plasma cells, lymphocytes and scattered histiocytes. On IHC, the cells were positive for smooth muscle actin, vimentin, CD3, CD20, kappa and lambda. Final IHC was suggestive of inflammatory myofibroblastic tumor. Currently, patient is asymptomatic at 9 months follow-up.

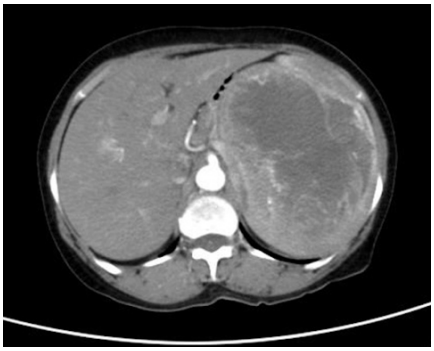

Fig. 1. Case 1 - CT (Arterial phase) image of lesion in the spleen with intense peripheral enhancement and necrotic areas

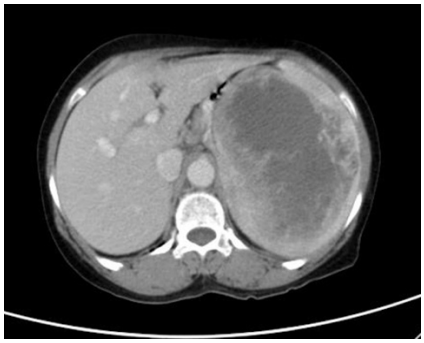

Fig. 2. Case 1 – CT (Venous phase) image of lesion in the spleen showing minimal centripetal enhancement

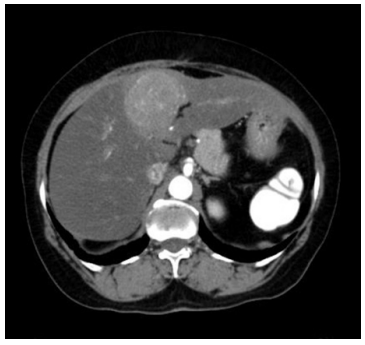

Fig. 3. Case 1 – CT (Arterial phase) image of liver lesion in segment 4a, 4b showing moderate patchy enhancement

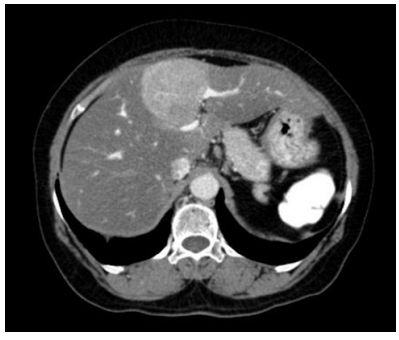

Fig 4. Case 1 – CT (Portal Phase) image of liver lesion in segment 4a, 4b showing contrast washout

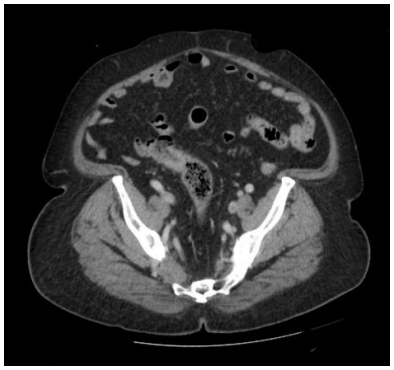

Fig. 5. Case 2 – CT (Venous phase) image of lesion in sigmoid colon

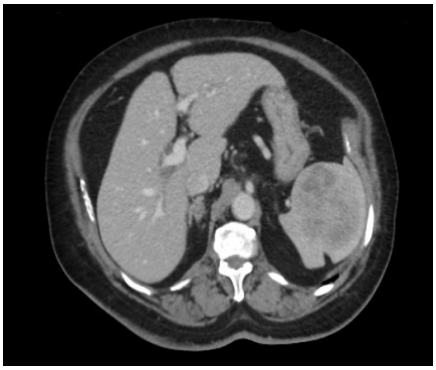

Fig. 6. Case 2 – CT (Venous phase) image of heterogeneously enhancing lesion in spleen

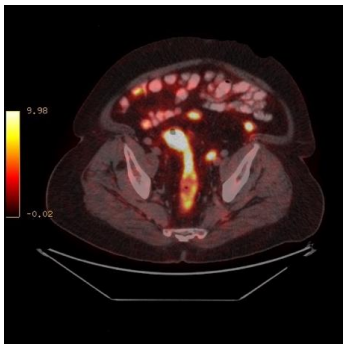

Fig. 7. Case 2 – PET CT image showing uptake in the lesion in sigmoid colon

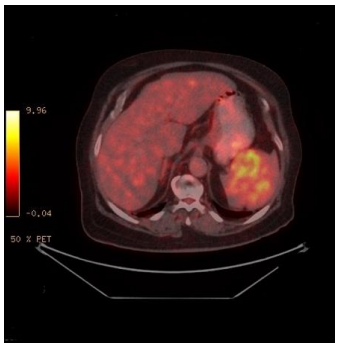

Fig. 8. Case 2 – PET CT image showing uptake in the splenic lesion

3. DISCUSSION

In this study we are reporting two cases of IMT of the spleen which have presented us with different challenges in diagnosis and management. In our first case, the incidentally detected splenic lesion was reported as angiosarcoma or hemangioendothelioma on CT. This is one of the typical situations where a preoperative definitive diagnosis is very difficult to establish. Hence, we went ahead with splenectomy. The diagnosis was clinched only on histopathology of the resected specimen, similar to most cases reported in literature. However, four years later, she developed a metachronous lesion in the liver. This is a rare occurrence, only few cases of synchronous or metachronous liver lesions from splenic IMT have been reported [17,18,19]. The previous case reports have opined that resection, if feasible, should be considered the treatment of choice for liver lesions from splenic IMT [18,19]. In our case, we decided against resection for multiple reasons. Firstly, we were able to establish a preoperative diagnosis of IMT through a core biopsy. Secondly, our patient's performance status was not considered good enough for a major hepatectomy. The non-surgical modalities of treatment described for hepatic IMT include observation, steroids, chemotherapy and irradiation [19]. We used steroids in our case, and patient is doing well on follow-up, with slight regression in the size of the lesion.

In our second case, the lesion in the spleen was incidentally detected during the staging workup of adenocarcinoma sigmoid colon. We could not find any similar report from published literature. The CECT picture suggested lymphoma and metastasis as the possible differentials for the splenic lesion. Isolated splenic metastasis, although rare, has been reported in cases with adenocarcinoma colon and rectum [20]. PET-CT was done to rule out lesions elsewhere, following which we proceeded with surgery. Core biopsy was not attempted, considering the potential complications inherent with percutaneous biopsy from the spleen in interpretation of the core biopsy samples. On exploration, since there was

no evidence of metastatic disease, we went ahead with a radical resection of the colon and spleen.

Splenectomy is the treatment for IMT spleen; in most cases, the definitive diagnosis is made after surgery. Partial splenic resection has also been described in selective cases [15], but this is rendered difficult by the lack of preoperative diagnosis in high percentage of cases. Although IMT is considered a lesion of low malignant potential, follow-up is mandatory, as further emphasized by the metachronous liver lesion detected in our patient. The treatment of local recurrence and metastasis depends on the individual case, no definite guidelines have been proposed. Surgical re-resection may be attempted if feasible, whereas other modalities including radiotherapy, chemotherapy, immunomodulators and corticosteroids have been described if surgery is not feasible [19,21].

4. CONCLUSION

Although IMT of the spleen is an uncommon tumor, it can cause significant diagnostic and therapeutic dilemma due to the lack of typical findings on clinical examination and cross sectional imaging. Surgical resection is the treatment of choice for splenic IMT, the diagnosis is often established only on histopathology of resected specimen. Although the prognosis after splenectomy is good, the patients need to be kept on close follow-up, as local and distant metastasis can occur in a small subset of patients.

CONSENT

Written informed consent was obtained from both the patients. Complete anonymity of the patients has been maintained, no identifying information or images have been included in the publication.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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