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CASE STUDY

INFIAMMATORY MYOFIBROBLASTIC TUMOR OF THE SMALL INTESTINE

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ABSTRACT

Inflammatory Myofibroblastic Tumor (IMT), previously called inflammatory pseudotumor and plasma cell granuloma, belongs to a class of rare spindle cell lesions showing a rather unpredictable biological behavior with occasional tendency toward invasion to the surrounding tissue and Local recurrence. The lesion, as primarily described by Bahadori and Liebow in 1973, is a reactive/inflammatory process in the pulmonary system mostly occurring in children and young adults. Since then, many cases have been reported in older patients in addition to extra-pulmonary sites. Inflammatory myofibroblastic tumor (IMT) emerges as a pseudotumor with malignant manifestation (1). This inflammatory tumor is usually seen in children and adolescents. It can affect all the body organs (2). The most common localization of this tumor is in the lungs, Mesentery comes in the second place (3). The pathogenesis of this inflammatory tumor cannot be accurately recognized (4). The manifestation of disease is variable with respect to the Involved organ, and the compressive effects of tumor are generally important (5). The definitive diagnosis of this tumor is possible through surgery and pathology, and the removal of symptoms usually requires there section of the mass (6).

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INTRODUCTION

Inflammatory Myofibroblastic Tumor (IMT), previously called inflammatory pseudotumor and plasma cell granuloma, belongs to a class of rare spindle cell lesions showing a rather unpredictable biological behavior with occasional tendency toward invasion to the surrounding tissue and Local recurrence. The lesion, as primarily described by Bahadori and Liebow in 1973, is a reactive/inflammatory process in the pulmonary system mostly occurring in children and young adults. Since then, many cases have been reported in older patients in addition to extra-pulmonary sites. Inflammatory myofibroblastic tumor (IMT) emerges as a pseudotumor with malignant manifestation (Day *et al*., 1986). This inflammatory tumor is usually seen in children and adolescents. It can affect all the body organs (Malkhlouf and Sobin, 2002). The most common localization of this tumor is in the lungs, Mesentery comes in the second place (Leuschner, 2010). The pathogenesis of this inflammatory tumor cannot be accurately recognized (Bahadori and Liebow, 1973). The manifestation of disease is variable with respect to the Involved organ, and the compressive effects of tumor are generally important (Cerfolio *et al*., 1999). The definitive diagnosis of this tumor is possible through surgery and pathology, and the removal of symptoms usually requires there section of the mass (Pettinato *et al*., 1990).

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Case report

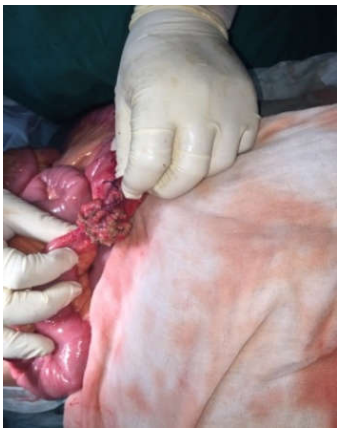
A 36 years old male presented with a complaint of dullaching abdominal pain, low grade fever and non-bilious and non-projectile vomiting of 1 month duration. There was no history of not passing stool, any bladder abnormality and any other complains. Patient have addiction of tobacco chewing and alcohol. General and systemic examinations were essentially normal. On examination, abdomen was distended. Tenderness and gaurding were present all over abdomen. Rigidity was absent. There was no organomegaly and there was no fluid in abdomen. There were increased bowel sounds on auscultation. On per rectal examination, stool was present. There was no active bleeding. There were no perianal bruises. Anal sphincter tone was normal. Proctoscopy was normal. X ray abdomen standing revealed few abnormal air fluid levels. There was no sign of pneumoperitoneum. USG abdomen whole suggestive of visualized bowel loops in abdomen appears dilated measuring 32 to 33mm and shows sluggish peristalsis of its

contents. There was cavernous transformation of portal vein. There was 3mm calculus present at lower pole of left kidney. There was minimal free fluid present in interbowel space. CECT abdomen with pelvis was suggestive of Jejunal loops are seen on right side and ileal loops seen on left side of abdomen. DJ flexure seen on right side of spine. Cecum, ascending colon and ileo-ceccal junction seen on right side of abdomen. Superior mesenteric vein seen anterior to the superior mesenteric artery-s/o intestinal malrotation. Significant narrowing of distal jejunam seen in umbilical region on left side with resultant dilatation of jejuna loops proximal to the narrowing---? Secondary to adhesions. No obvious wall thickening or mass lesion seen. Ileal loops appear collapsed. Non visualization of main portal vein represent chronic thrombosis. Multiple periportal, GB wall, perigastic and omental collaterals seen.

Management

Emergency exploratory laparotomy with resection of jejunoileal segment which was send for histopathological examination jejunoileal anastomosin with through peritoneal lavage done under general anaesthesia. Histopathology report show inflammatory myofibroblasitc tumour and both surgical margin and other areas show changes of serositis. Aspirated peritoneal fluld's culture and sensitivity report suggested of gram negative bacilli i.e. proteus. Intraoperative period was remain uneventful. Postoperatively, wound infection was occurred which was managed conservatively by daily dressings and antibiotics. Which was followed burst abdomen which was managed conservatively by daily saline dressing, abdominal binder and antibiotics. Than patient was discharged on daily dressing.

Intraoperative Photoes



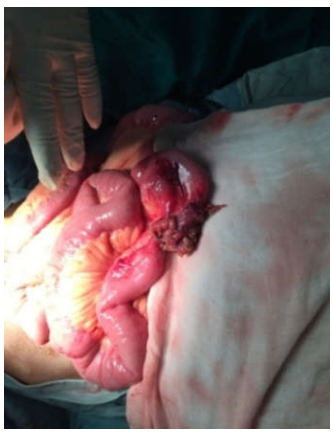
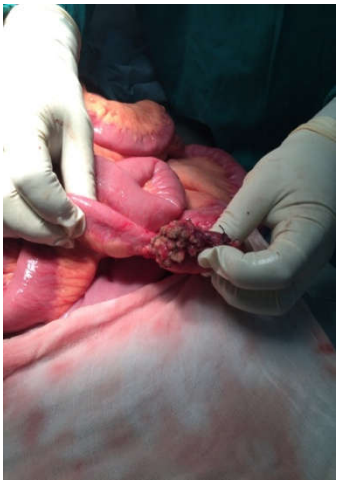


Figure 1- Sex distribution in both groups



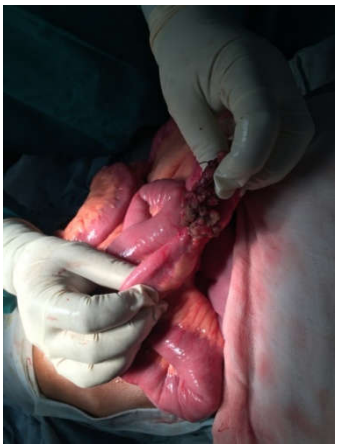


Figure 2. Age distribution in both groups

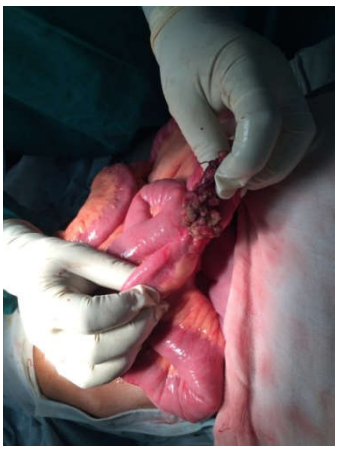


Figure 3. Comparison of mean serum cholesterol of both groups

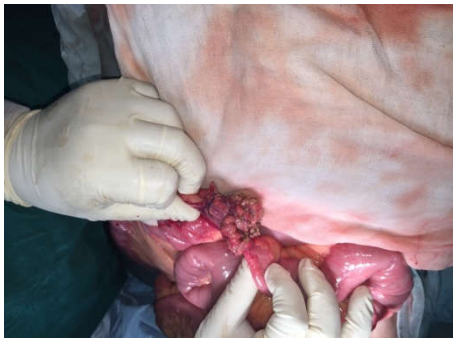


Figure 4. Comparison of mean bile cholesterol of both groups

DISCUSSION

IMT is a rare, but distinctive spindle cell tumor that contains a variable number of inflammatory cells, including plasma cells. This is the reason for previous designation of mass as plasma cell granuloma. Plasma cell granuloma as well as other (Bahadori and Liebow, 1973) alternative terms are discouraged to be used including inflammatory pseudotumor, inflammatory myofibroblastoma and inflammatory myofibrohistioblastic proliferation. (Day *et al*., 1986; Malkhlouf and Sobin, 2002; Bahadori and Liebow, 1973; Cerfolio *et al*., 1999; Leon *et al*., 2006) Multiple studies have shown expression of p80 and the clonal rearrangement of the anaplastic lymphoma kinase (ALK) gene on chromosome 2p23 leading to the over expression of the oncoprotein in the spindle cell components of some of these tumors. Abnormalities in chromosome 2p is seen in up to 60% of patients younger than 10 years of age. This finding indicates a true neoplastic nature for the tumor cells. (Coffin *et al*., 2001; Lawrence *et al*., 2000) dNA aneuploidy and association of the lesion with oncogenic viruses such as epstain-Barr virus, Human Herpes virus type 8 and over expression of IL-6 have also been demonstrated and proposed to be involved in the pathogenesis of the tumor by some investigators. (Park *et al*., 2008; Attili *et al*., 2005; Snyder *et al*., 1995) The lesion usually occurs in children and young adults but can develop in older ages with no predilection for any sex. The lung is the most common site of involvement but the lesion has also been reported in other organs including the stomach, mesentery, omentum, retroperitoneum as well as the kidneys, renal pelvis, liver, spleen, esophagus and lymph nodes. (Cerfolio *et al*., 1999; Pettinato *et al*., 1990; Leon *et al*., 2006; Attili *et al*., 2005; Karnak *et al*., 2001) In a case series of 38 tumors, stomach was found to be the most common extra pulmonary site observed in 34% of the cases. Intra-abdominal lesions usually present with non-specific signs and symptoms including abdominal pain, gastric and intestinal mass with occasional obstruction and growth retardation in children. (Day *et al*., 1986; Malkhlouf and Sobin, 2002; Demirkan *et al*., 2001) Constitutional symptoms may occur and include fever, night sweats, weight loss and malaise. Laboratory abnormalities are rarely present and could include anemia, thrombocytosis, an elevated ESR and hypergammaglobulinemia. These abnormalities often resolve with excision of the lesion. IMT has a well known tendency for local invasive behavior and recurrence. Only a small risk of distant metastasis has been reported by some authors. (Pettinato *et al*., 1990; Karnak *et al*., 2001; Gleason and Hornick, 2008; Spencer, 1984)

Characteristic histopathological findings in typical IMT are a fasciitis-like, compact spindle cell proliferation with areas of myxoid change and hypocellularity showing a collagenous back ground. Various numbers of mixed inflammatory cells including polyclonal plasma cells, lymphocytes, eosinophils and rarely foamy macrophages are invariably seen. (Gleason and Hornick, 2008) Typically the spindle cells of the IMT express vimentin, smooth muscle actin and other markers which correspond to the myofibroblastic nature of these cells. Histological features usually cannot predict the biological behavior of the tumor. The presence of aneuploidy may, however, indicate the possibility of a local aggressive behavior and recurrence. In pulmonary lesions atypia of the spindle cells are believed to indicate aggressiveness. (Coffin *et al*., 2001) The treatment of choice is believed to be complete surgical excision and long term follow up with physical examination, imaging techniques and serial monitoring of the erythrocyte sedimentation rate. Radiotherapy and chemotherapy (cisplatin, doxorubicin and methotrexate) have also been tried as an adjunct to surgery with no evidence for substantial benefit for the patient. (Dishop *et al*., 2003) Steroid therapy achieved regression of a case of renal IMT. The benefits of non-steroidal anti-inflammatory medications are controversial. (Hagenstad *et al*., 2003; Sanders *et al*., 2001) The case we present showed some of the typical features of IMT, namely the clinical presentation with intestinal obstruction. The tumor also showed the typical histomorphology of proliferating spindle cells and a considerable number of inflammatory cells, including plasma cells.

Conclusion

IMT is a neoplastic lesion with the tendency for local aggressive behavior and recurrence. This illness should be considered in the list of the differential diagnoses of spindle cell tumors in any part of the body of patients at any age, particularly children and young adults.

REFERENCES

Abraham SC, Montgomery EA, Singh VK, Yardley JH, Wu TT. 2002. Gastric adenomas : intestinal – type and gastric - type adenomas differ in the risk of adenocarcinoma and presence of background mucosal pathology. *Am J Surg Pathol*., 26:1276-85.

Attili SV, Chandra CR, Hemant DK, Bapsy PP, RamaRao C, Anupama G. 2005. Retroperitoneal inflammatory myofibroblastic tumor. *World J Surg Oncol*., 3:66.

Bahadori M. and Liebow AA. 1973. Plasma cell granuloma of the lung. *Cancer*, 31:191-208.

Cerfolio RJ, Allen MS, Nascimento AG, Deschamps C, Trastek VF, Miller DL, *et al*. 1999. Inflammatory pseudotumors of the lung. *Ann Thorac Surg*., 67:933-6.

Cessna MH, Zhou H, Sanger WG, Perkins SL, Tripp S, Pickering D, Daines C, Coffin CM. 2002. Expression of ALK1 and p80 in inflammatory myofibroblastic tumor and its mesenchymal mimics: a study of 135 cases. *Mod Pathol.,* 15:931-8.

Coffin CM, Patel A, Perkins S, Elenitoba-Johnson KS, Perlman E, Griffin CA. 2001. ALK1 and p80 expression and chromosomal rearrangements involving 2p23 in inflammatory myofibroblastic tumor. *Mod Pathol*., 14:569-76.

Coffin CM, Watterson J, Priest JR, Dehner LP. 1995. Extra-pulmonary inflammatory myofibroblastic tumor (in-flammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol*., 19:859-72.

Cook JR, Dehner LP, Collins MH, Ma Z, Morris SW, Coffin CM, *et al*. 2001. Anaplastic lymphoma kinase (ALK) ex-pression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study. *Am J Surg Pathol*., 25:1364-71.

Day DL, Sane S, Dehner LP. 1986. Inflammatory pseudotumor of the mesentery and small intestine. *Pediatric Radiol.,* 16:210-5.

Demirkan NC, Akalin T, Yilmaz F, Ozgenc F, Ozcan C, Alkanat MB, *et al*. 2001. Inflammatory myofibroblastic tumor of small bowel wall in childhood : report of the case and a review of the literature. *Pathol Int*., 51:47-9.

Dishop MK, Warner BW, Dehner LP, Kriss VM, Greenwood MF, Geil JD, *et al*. 2003. Successful treatment of inflammatory myofibroblastic tumor with malignant transformation by surgical resection and chemotherapy. *J Pediatr Hematol*., 25:153-8.

Gleason B, Hornick J. 2008. Inflammatory myofibroblastic tumours: where are we now? *J Clin Pathol*., 61:428-37.

Hagenstad CT, Kilpatrick SE, Pettenati MJ, Savage PD. 2003. Inflammatory myofibroblastic tumor with bone marrow involvement. A case report and review of the literature. *Arch Pathol Lab Med*., 127:865-77.

Karnak I, Senocak ME, Ciftci AO, Cağlar M, Bingöl-Koloğlu M, Tanyel FC, *et al*. 2001. Inflammatory myofibroblastic tumor in children: Diagnosis and treatment. *J Pediatr Surg*., 36:908-12.

Karnak I, Senocak ME, Ciftci AO, Cağlar M, Bingöl-Koloğlu M, Tanyel FC, *et al*. 2001. Inflammatory myofibroblastic tumor in children: diagnosis and treatment. *J Pediatr Surg*., 36:908-12.

Kovach S, Fischer AC, Katzman PJ, Salloum RM, Ettinghausen SE, Madeb R, *et al*. 2006. Inflammatory myofibroblastic tumors. *J Surg Oncol*., 94:385-9.

Lawrence B, Perez-Atayde A, Hibbard MK, Rubin BP, Dal Cin P, Pinkus JL, *et al*. 2000. TPM3-ALK and TPM4-ALK oncogenes in inflammatory myofibroblastic tumors. *Am J Pathol*., 157:377-84.

Leon CJ, Castillo J, Mebold J, Cortez L, Felmer R. 2006. Inflammatory myofibroblastic tumor of the stomach, an unusual complication after gastrectomy. *Gastrointest Endoscopy*, 63:347-9.

Leuschner I. 2010. Inflammatory myofibroblastic tumor. *Pathologe*., 31:106-8.

Malkhlouf HR. and Sobin LH. 2002. Imflammatory myofibroblastic tumors (inflammatory pseudotumors) of the gastrointestinal tract: How closely are they related to inflammatory fibroid polyps? *Hum Pathol*., 33:307-15.

Park SH, Kim JH, Min BW, Song TJ, Son GS, Kim SJ, *et al*. 2008. Exophytic inflammatory myofibroblastic tumor of the stomach in an adult woman: A rare cause of hemoperitoneum. *World J Gastroenterol*., 14:136-9.

Pettinato G, Manivel JC, De Rosa N, Dehner LP. 1990. Inflammatory myofibroblastic tumor plasma cell granuloma). Clinicopathological study of 20 cases with immunohistochemical and ultrastructural observations. *Am J Clin Pathol*., 94:538-46.

Sanders BM, West KW, Gingalewski C, Engum S, Davis M, Grosfeld JL. 2001. Inflammatory pseudotumor of the alimentary tract: Clinical and surgical experience. *J Pediatr Surg*., 36:169-73.

Sastre-Garau X, Couturier J, Derré J, Aurias A, Klijanienko J, Lagacé R. 2002. Inflammatory myofibroblastic tumour (inflammatory pseudotumour) of the breast. Clinicopathological and genetic analysis of a case with evidence for clonality. *J Pathol*., 196:97-102.

Snyder CS, Dell'Aquila M, Haghighi P, Baergen RN, Suh YK, Yi ES. 1995. Clonal changes in inflammatory pseudotumor of the lung ;a case report. *Cancer*, 76:1545-9.

Spencer H. 1984. The pulmonary plasma cell/histiocytoma complex. *Histopathology*, 8:903-16.

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