An Obstructed Malignant Inflammatory Myofibroblastic Tumour of the Rectum

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ABSTRACT

Inflammatory myofibroblastic tumour is rare but more common in children. It shows an immunophenotypic features of myofibroblastic differentiation, hence bearing neoplastic potential. The diagnosis is challenging especially if it involves rectum. Surgical resection is the mainstay of treatment if clinically obstructed. A 65-year-old gentleman presented with intestinal obstruction, which then followed by a hartmann's procedure. Final diagnosis is a rare case of inflammatory myofibroblastic tumour of the rectum. We discuss its genetic involvement with a literature review.

KEY WORDS

Gastrointestinal tract, Myofibroblastic tumour, Rectum

INTRODUCTION

Inflammatory myofibroblastic tumour (IMT) is a challenging lesion to classify and diagnose. Considered to have neoplastic predilection, it is characterized by spindle cell pseudosarcomatous proliferation in an inflammatory stroma. Multiple case reports demonstrate recurrent and constant clonal genetic alterations of IMT.[1-5] Although commonly seen in children and young adults, we present a case of a rectal inflammatory myofibroblastic tumour in a 65-year-old gentleman who presented an abdominal pain and distension with no bowel motion for two days duration with dilated small bowel seen on abdominal radiograph.

CASE REPORT

A 65-year-old gentleman presented to us with a complaint of abdominal pain and distension for two days associated with no bowel opening. However he was still able to pass flatus. It was the first time this has ever occurred in his life. He initially sought treatment at a local general practitioner's clinic but after 3 days, the medication given did not improve his condition. On further questioning, he denied any bleeding per rectum, loss of appetite or weight, had no previous surgeries and does not have any history of malignancies in his family.

Physical examination revealed a generally distended abdomen, no obvious palpable masses and no signs of peritonitis. Abdominal radiograph showed dilated large and small bowels. Blood investigation showed neutrophil predominant leukocytosis and other panels were normal. A nasogastric tube was inserted and bile was seen on aspiration of the tube. A diagnosis of intestinal obstruction was made.

Patient was rushed to the operating theatre for an exploratory laparotomy to locate and relieve the site of obstruction. Intraoperatively, there was a circumferential mass at the upper rectum without invasion to the adjacent structures. A hartmann's procedure was performed. The specimen was sent for histopathological examination and it revealed an inflammatory myofibroblastic tumour (fig. 1 and 2). Post-operatively he had recovered well. He underwent a reversal of Hartmann's procedure 6 months after the initial surgery. He is now on regular follow-up.

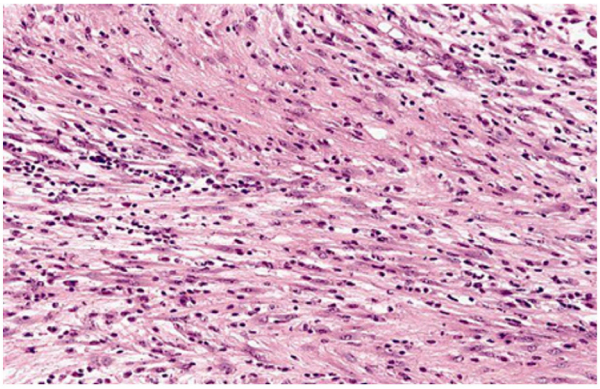

Figure 1. The myofibroblastic cells are of uniform spindle-shaped nuclei with fine nuclear chromatin (hematoxylin and eosin, original magnification x 4)

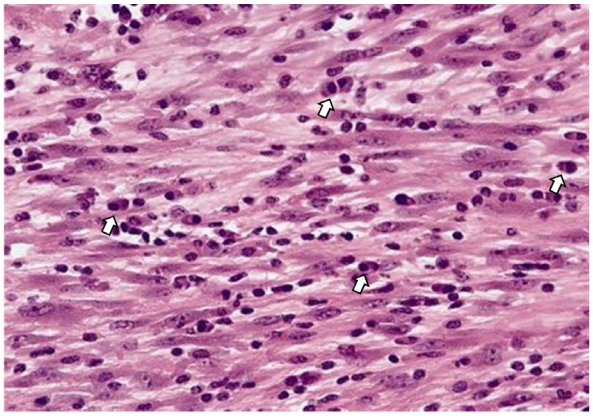

Figure 2. Typical appearance of spindled myofibroblastic cells with inflammatory infiltrate predominantly plasma cells with eccentric nuclei (arrow)

DISCUSSION

With multiple synonyms such as plasma cell granuloma or inflammatory pseudotumour, IMT remains as a rare neoplasm. Initially thought to be a benign reactive process, it has now become a malignant neoplasm. It is characterized by myofibroblastic spindle cells arising together with an inflammatory infiltrates of plasma cells. Three main histological patterns are known; nodular fasciitis-like, fibrous histiocytoma-like, and desmoid or scar tissue-type. Morphologically, IMT is circumscribed, small to large, firm, white to tan in color, single to multinodular lesions with a fleshy, myxoid cut. Although morphologically similar, IMT demonstrates a spectrum of features with varying etiologies such as Epstein-Barr virus, human herpes virus or over-expression of interleukin-6. Interestingly, they range from proliferations either reactive or regenerative, to low grade neoplasms with risk of local recurrence and having metastatic potential. This is due to recent studies that show IMT as having cytogenetic clonality, recurrent involvement of chromosomal region 2p23 and metastasis of tumor.[1-5]

Although tumor sites are commonly in the lung tissues, extra-pulmonary IMT has been reported with a myriad of presenting complaints. It may present with vague symptoms such as hematuria, non-specific abdominal pain, abdominal mass and even fever of unknown origin. IMT runs as a benign path, locally confined and mostly grows slowly over time. Aggressive behavior of the tumour can be predicted when microscopically showing presence of ganglion-like cells, cellular atypia, aneuploidy, and p53 over expression.[6,7]

Cellular features in IMT show the ultra-structural and immunophenotypic features of myofibroblastic differentiation. In spindle cell cytoplasm, diffuse cytoplasmic staining for vimentin and actin are focally to diffusely seen. IMT less commonly shows reactivity for desmin. Myogenin, myoglobin, S100, CD117 and epithelial membrane antigen are not found in IMT.8 With regard to its neoplastic potential, recurrent translocation involving 2p23, anaplastic lymphomakinase (ALK) gene site and ALK gene fusions with two tropomyosin genes (TPM3 and TPM4) usually indicate a neoplastic pathogenesis.9 Although characterizes neoplastic potential, the ALK is not related to any particular histological pattern.

Epidemiologically, IMT is more common in children rather than adults and usually presents with symptoms as mentioned earlier. It has no gender predilection although in some cases, extra-pulmonary IMT surfaces more in the young women. Common extra-pulmonary sites include mesentery, omentum, retroperitoneum, liver, spleen and bladder.10 Poor prognostic factors for IMT include ALK-negative and extra-pulmonary IMT especially in the abdomen.

Surgical resection is the mainstay of treatment. With complete excision, IMT rarely recurs, however sometimes it may be locally aggressive (depending on site and proximity) or metastatic (uncommonly to the lungs and bone).11 Usually, recurrence occurs within a year of excision. Radical surgery and adjuvant therapy are not indicated for the treatment of IMT. Metastatic disease has been reported in children but rarely in adults. No existing criteria has been put forth to determine whether distant spread are metastatic IMT or multicentric disease.

In conclusion, IMT is a unique entity and has non-specific presenting complaints. Commoner in children, it is nowadays considered to be malignant with some reports of metastatic disease. Surgical resection is the mainstay of treatment with no radical surgery or adjuvant therapy provided. More information needs to be discovered so that we can safely stratify the disease according to its cytochemical properties, thus enabling adequate planning of the treatment and long term follow-up requirements.

REFERENCES

1. Pungpapong S, Geiger XJ, Raimondo M. Inflammatory myofibroblastic tumor presenting as a pancreatic mass: a case report and review of the literature. *JOP.* 2004;5:360-7.
2. Coffin CM, Dehner LP, Meis-Kindblom JM. Inflammatory myofibroblastic tumor, inflammatory fibrosarcoma, and related lesions: a historical review with differential diagnostic considerations. *Semin Diagn Pathol.* 1998;15:102-10.
3. Freeman A, Geddes N, Munson P, et al. Anaplastic lymphoma kinase (ALK 1) staining and molecular analysis in inflammatory myofibroblastic tumors of the bladder: a preliminary clinicopathological study of nine cases and review of the literature. *Mod Pathol.* 2004;17:765-71.
4. Kapusta LR, Weiss MA, Ramsay J, Lopez-Beltran A, Srigley JR. Inflammatory myofibroblastic tumors of the kidney: a clinicopathologic and immunohistochemical study of 12 cases. *Am J Surg Pathol.* 2003;27:658-66.
5. Biselli R, Boldrini R, Ferlini C, Boglino C, Inserra A, Bosman C. Myofibroblastic tumors: neoplasias with divergent behavior. Ultrastructural and flow cytometric analysis. *Pathol Res Pract.* 1999;195:619-32.
6. Sastre-Garau X, Couturier J, Derre J, Aurias A, Klijanienko J, Lagace R. Inflammatory myofibroblastictumour (inflammatory pseudotumour) of the breast. Clinicopathological and genetic analysis of a case with evidence for clonality. *J Pathol.* 2002;196:97-102.
7. Hussong JW, Brown M, Perkins SL, Dehner LP, Coffin CM. Comparison of DNA ploidy, histological and immunohistochemical findings with clinical outcome in inflammatory myofibroblastic tumors. *Mod Pathol.* 1999;12:279-86.
8. Coffin CM, Patel A, Perkins S, Elenitoba-Johnson KS, Perlman E, Griffin CA. ALK1 and p80 expression and chromosomal rearrangements involving 2p23 in inflammatory myofibroblastic tumor. *Mod Pathol*. 2001;14(6):569-76.
9. Lawrence B, Perez-Atayde A, Hibbard MK, et al. TPM3-ALK and TPM4-ALK oncogenes in inflammatory myofibroblastic tumors. *Am J Pathol.* 2000;57(2):377-84.
10. Browne M, Abramson LP, Chou PM, Acton R, Holinger LD, Reynolds M. Inflammatory myofibroblastic tumor (inflammatory pseudotumour) of the neck infiltrating the trachea. *J Pediatr Surg.* 2004;39(10):e1-4.
11. Morotti RA, Legman MD, Kerkar N, Pawel BR, Sanger WG, Coffin CM. Pediatric inflammatory myofibroblastic tumor with late metastasis to the lung: case report and review of the literature. *Pediatr Dev Pathol.* 2005;8(2):224-9.