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Inflammatory Pseudotumor Occurring at a Rare Site - Female Pelvic Cavity- A Case Report

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Abstract: Inflammatory pseudotumor is a quasineoplastic lesion consisting of inflammatory cells and myofibroblastic spindle cells. Inflammatory pseudotumor most commonly involves the lung and the orbit, but it has been reported to occur in nearly every site in the body, from the central nervous system to the gastrointestinal tract. The aim of this article is to describe the various anatomic locations where inflammatory pseudotumors occur, review the pathogenesis of inflammatory pseudotumor and briefly discuss the differential diagnoses and treatment options.
Keywords: Inflammatory pseudotumor, Myoepithelial cell proliferation, Inflammatory cells.

INTRODUCTION

Inflammatory pseudotumor has been described in the literature by many different names, a fact that suggests the complexity and variable histologic characteristics and behavior of this entity. Inflammatory pseudotumor is a quasineoplastic lesion consisting of inflammatory cells and myofibroblastic spindle cells [2, 3].

Terminology

- Inflammatory myofibroblastic tumor (IMT) is terminology for neoplastic lesions
- Inflammatory lesions are often called pseudotumors.
- Also known as pseudosarcoma, atypical myfibroblastic tumor, atypical fibromyxoid tumor, plasma cell granuloma.

Understanding the pathophysiologic characteristics and natural history of inflammatory pseudotumor and considering this entity in the differential diagnosis of soft-tissue tumors may prevent unnecessary radical surgery. Inflammatory pseudotumor was first observed in the lung and described by Brunn in 1939 and was so named by Umiker *et al* in 1954 because of its propensity to clinically and radiologically mimic a malignant process [1].

CASE DISCUSSION

Clinical details: This is a case of cystic lesion from pelvic cavity of a 28 yr female, who presented with pain abdomen, fever on and off, pallor, and weight loss since 5 months, on further evaluation a cystic lesion of ?right ovary was identified. Excision biopsy was done and sent for Histopathological examination.

Grossly: Solid with central cystic mass of 7*5*4cms size. Surface smooth with congested blood vessels and thin layer of exudates. Thickness of cyst wall varying from 1-2cm.Discoloured yellowish white lesion with tiny sieve like openings. Inner cavitation had ragged appearance, with spotty yellow, white necrotic areas (Fig-1).

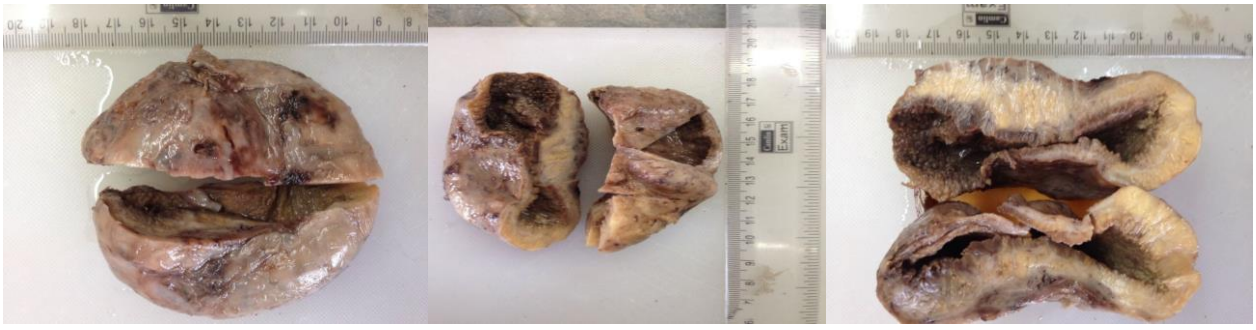

Fig-1: Gross images of the lesion

Histopathology**:** Cyst wall composed of connective tissue with irregular strands of collagen and smooth muscle. Across the wall infiltration by foamy histiocytes [xanthoma cells], histiocytes, lymphocytes, plasmacells, neutrophils and rare eosinophils. Central luminal side of the lesion shows extensive granulation tissue. No evidence of tuberculosis, malakoplakia or endometriosis (Fig-2, 3).

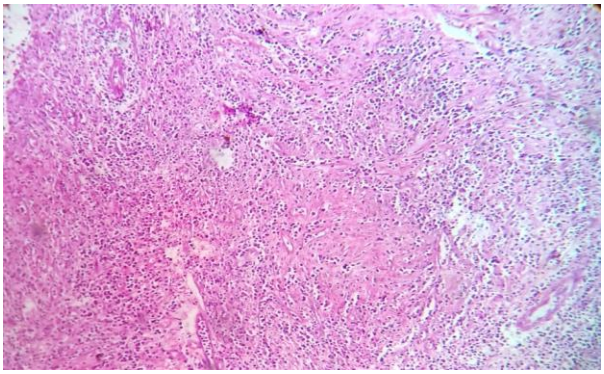

Fig-2: Spindled myoepithelial cell proliferation and lymphocytic infiltrate

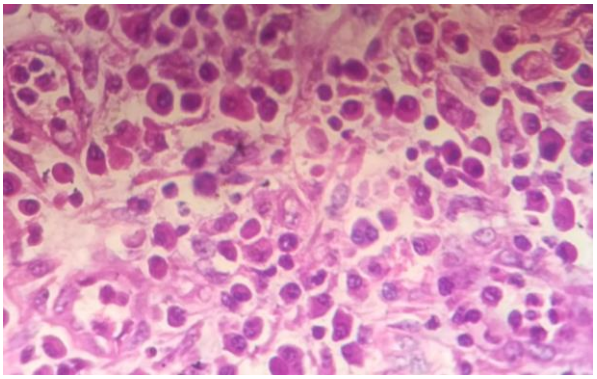

Fig-3: Cyst wall composed of connective tissue with irregular strands of collagen and smooth muscle, across the wall infiltration by foamy histiocytes [xanthoma cells], histiocytes, lymphocytes, plasma cells, neutrophils and rare eosinophils

DISCUSSION

Epidemiology

- More common in children and young adults
- More common in females

Sites

- Can occur anywhere in the body.

Etiology

- Idiopathic, no known predisposing conditions

Clinical Features: Pain, fever, weight loss, anemia, thrombocytosis, increased erythrocyte sedimentation rate and elevated gamma globulins are more common with IMTs.

Benign, but frequently misinterpreted as leiomyosarcoma or rhabdomyosarcoma. May recur locally, but doesn't metastasize.

Pathogenesis

The causes of inflammatory pseudotumor are unknown.

1. Some authors believe this tumor is a low-grade fibrosarcoma with inflammatory (lymphomatous) cells. The propensity of inflammatory pseudotumors to be locally aggressive, to frequently be multifocal, and to progress occasionally to a true malignant tumor supports this idea [3-7]. Immunohistochemical studies of T- and B-cell subpopulations may be helpful in distinguishing inflammatory pseudotumor from lymphoma. Inflammatory pseudotumors usually contain both T cells and B cells, whereas in lymphoma, a (clonal) B- or T-cell population predominates. Furthermore, the heterogeneity of the inflammatory cell population in inflammatory pseudotumor tends to exclude lymphoma.

2.In some cases, inflammatory pseudotumor is thought to result from inflammation following minor trauma or surgery or to be associated with other malignancy [8, 9]. An immune-autoimmune mechanism has also been implicated. In one case, inflammatory pseudotumor was associated with vasculitis and inferior vena caval thrombosis, with anti-C3 and antifibrinogen deposits found in the vessel wall [10].

3.There appears to be a subset of inflammatory pseudotumors that occur secondary to infection. Organisms found in association with inflammatory pseudotumor include mycobacteria associated with spindle cell tumor; Epstein-Barr virus found in splenic

and nodal pseudotumors; actinomycetes and nocardiae found in hepatic and pulmonary pseudotumors, respectively; and mycoplasma in pulmonary pseudotumors [5]. There have been case reports of inflammatory pseudotumor associated with infections caused by other organisms, including *Mycobacterium avium–intracellulare* complex, *Corynebacterium equi*, *Escherichia coli*, *Klebsiella*, *Bacillus sphaericus*, *Pseudomonas*, *Helicobacter pylori*, and *Coxiella burnetti* [3, 5, 8, 11, 12] . It has been suggested that histiocytic cells predominate in inflammatory pseudotumors associated with infection, whereas myofibroblastic cells characterize the lesions more likely to be considered true neoplasms [5].

Many of the features of inflammatory pseudotumor can be related to the production of inflammatory mediators such as cytokines and particularly interleukin-1 [13]. Interleukin-1, which is produced mainly by monocytes, by macrophages, and to a lesser extent by other cells, has a wide range of local and systemic effects. Locally, it stimulates the proliferation of fibroblasts, the extravasation of neutrophils, and the activation and increase of procoagulant activity of the vascular endothelium. Systemically, it induces production of acute-phase reactants by hepatocytes, proteolysis, and neurologic disturbances.

Treatment

- Conservative surgical excision

Gross Description

Polypoid mass with pale, firm cut surface, may be very large often gelatinous.

Micro Description

- **Essential criteria**: spindled myoepithelial cell proliferation and lymphocytic infiltrate
- **Patterns include**:
  (1) loose stellate cells with myxoid background containing scattered inflammatory cells (nodular fasciitis-like)
  (2) spindle cells with a compact fascicular pattern (fibrohistiocytoma-like)
  (3) sparse cellular, collagenous areas (desmoid-like)
  (4) mixed
- Cells are stellate myofibroblasts with abundant eosinophilic cytoplasm, elongated nuclei
- May be cellular and infiltrative with mucosal ulceration, necrosis, cytologic atypia .

Positive Stains

- Smooth muscle actin, desmin, ALK1 (75-89%), [19] vimentin, calponin, variable keratin [20]

Negative Stains

- EMA, myogenin, p53 [21] h-caldesmon [22]

Electron Microscopy Description

- Myofibroblasts: bipolar cells with eosinophilic, elongated, tapering cytoplasmic processes without striation, central oval nuclei with smooth contours, open chromatin, occasional nucleoli
- No evidence of smooth muscle or skeletal muscle differentiation

Molecular / Cytogenetics Description

Usually ALK+ or ALK gene rearrangements by FISH [14]

Differential Diagnosis

- Leiomyosarcoma: smooth muscle morphology, strong desmin staining, keratin negative [15]
- Neurofibroma: neural morphology, strong S100+
- Rhabdomyosarcoma in children: necrosis or myxoid degeneration, moderate/severe nuclear atypia, more mitotic figures, no myofibroblastic differentiation; myoD1+, myogenin+ [16]
- Sarcomatoid (spindle cell) carcinoma.
- Calcifying fibrous pseudotumor: calcification, no myofibroblastic proliferation, actin negative [17]
- IgG4 related sclerosing lesion: IgG4+ plasma cells and the ratio of IgG4+ / IgG+ plasma cells is lower in IMT
- Low grade myofibroblastic sarcoma: more uniform appearance with higher cellularity, more prominent hyperchromasia, more infiltrative, ALK [18]
- Nodular fasciitis: smaller size, older patients, less inflammation

Consent

Taken from the patient.

Competing interests

We have no competing interests.

REFERENCES

1. Umiker WO, Iverson LC. Post inflammatory tumor of the lung: report of four cases simulating xanthoma, fibroma or plasma cell granuloma. J Thorac Surg. 1954;28:55-62.
2. Scott L, Blair G, Taylor G, Dimmick J, Fraser G. Inflammatory pseudotumors in children. J Pediatr Surg. 1988;23:755-758.
3. Hedlund GL, Navoy JF, Galliani CA, Johnson WH. Aggressive manifestations of inflammatory pulmonary pseudotumor in children. Pediatr Radiol. 1999;29:112-116.

4. Agrons GA, Rosado-de-Christenson ML, Kirejczyk WM, Conran RM, Stocker JT. Pulmonary inflammatory pseudotumor: radiologic features. Radiology. 1998;206:511-518.
5. Dehner LP. The enigmatic inflammatory pseudotumors: the current state of our understanding, or misunderstanding (editorial). J Pathol. 2000;192:277-279.
6. Maier HC, Sommers SC. Recurrent and metastatic pulmonary fibrous histiocytoma/plasma cell granuloma in a child. Cancer. 1987;60:1073- 1076.
7. Griffin CA, Haukins AL, Dvorack C, Henkle C, Ellingham T, Perlman EJ. Recurrent involvement of 2p23 in inflammatory myofibroblastic tumors. Cancer Res. 1999;59:2776-2780.
8. Sanders BM, West KW, Gingalewski C, Engum S, Davis M, Grosfeld JL. Inflammatory pseudotumor of the alimentary tract: clinical and surgical experience. J Pediatr Surg. 2001;36:169-173.
9. Maves CK, Johnson JF, Bove K, Malott RL. Gastric inflammatory pseudotumor in children. Radiology. 1989;173:381-383.
10. Stark P, Sandbank JC, Rudnicki C, Zahavi I. Inflammatory pseudotumor of the heart with vasculitis and venous thrombosis. Chest. 1992;102:1884-1885.
11. Wood C, Nickoloff BJ, Todes-Taylor NR. Pseudotumor resulting from atypical mycobacterial infection: a "histoid" variety of Mycobacterium avium-intracellulare complex infection. Am J Clin Pathol. 1985;83:524-527.
12. Levy S, Sauvanet A, Diebold M, Marcus C, Da Costa N, Thiefin G. Spontaneous regression of an inflammatory pseudotumor of the liver presenting as an obstructing malignant biliary tumor. Gastrointest Endosc. 2001;53:371-374.
13. Hytiroglou P, Brandwein MS, Stauchen JA, Mirante JP, Urken ML, Biller HF. Inflammatory pseudotumor of the parapharyngeal space: case report and review of the literature. Head Neck. 1992;14:230-234.
14. Sukov WR, Cheville JC, Carlson AW, Shearer BM, Piatigorsky EJ, Grogg KL, Sebo TJ, Sinnwell JP, Ketterling RP. Utility of ALK-1 protein expression and ALK rearrangements in distinguishing inflammatory myofibroblastic tumor from malignant spindle cell lesions of the urinary bladder. Modern pathology. 2007 May 1;20(5):592-603.
15. Mills SE, Bova GS, Wick MR, Young RH. Leiomyosarcoma of the urinary bladder: a clinicopathologic and immunohistochemical study of 15 cases. The American journal of surgical pathology. 1989 Jun 1;13(6):480-9.
16. Watanabe K, Baba K, Saito A, Hoshi N, Suzuki T. Pseudosarcomatous myofibroblastic tumor and myosarcoma of the urogenital tract: Immunohistochemical characteristics and differential diagnosis. Archives of pathology & laboratory medicine. 2001 Aug;125(8):1070-3.
17. Hill KA, Gonzalez-Crussi F, Chou PM. Calcifying fibrous pseudotumor versus inflammatory myofibroblastic tumor: a histological and immunohistochemical comparison. Modern Pathology. 2001 Aug 1;14(8):784-90.
18. Sun B, Qie S, Zhang S, Sun T, Zhao X, Gao S, Ni C, Wang X, Liu Y, Zhang L. Role and mechanism of vasculogenic mimicry in gastrointestinal stromal tumors. Human pathology. 2008 Mar 31;39(3):444-51.
19. Cook JR, Dehner LP, Collins MH, Ma Z, Morris SW, Coffin CM, Hill DA. Anaplastic lymphoma kinase (ALK) expression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study. The American journal of surgical pathology. 2001 Nov 1;25(11):1364-71.
20. Lundgren L, Aldenborg F, Angervall L, Kindblom LG. Pseudomalignant spindle cell proliferations of the urinary bladder. Human pathology. 1994 Feb 1;25(2):181-91.
21. Iczkowski KA, Shanks JH, Gadaleanu V, Cheng L, Jones EC, Neumann R, Nascimento AG, Bostwick DG. Inflammatory pseudotumor and sarcoma of urinary bladder: differential diagnosis and outcome in thirty-eight spindle cell neoplasms. Modern Pathology. 2001 Oct 1;14(10):1043-51.
22. Watanabe K, Kusakabe T, Hoshi N, Saito A, Suzuki T. h-Caldesmon in leiomyosarcoma and tumors with smooth muscle cell-like differentiation: its specific expression in the smooth muscle cell tumor. Human pathology. 1999 Apr 30;30(4):392-6.