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Significance of isolated vasculitis in the gynecological tract: what clinicians do with the pathologic diagnosis of vasculitis?☆

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ABSTRACT

Vasculitides includes a heterogeneous group of disorders with the common histologic findings of vascular wall inflammation. Systemic or localized disease (eg, renal vasculitis) has serious consequences. The incidence of isolated gynecologic vasculitis diagnosed on pathology specimens and its significance is little known. We performed a 20 year retrospective review including 53 cases with vasculitis diagnosis affecting the female genital tract identified in pathology reports. None had prior symptoms or were diagnosed with generalized vasculitis, while one patient had prior diagnosis of fibromyalgia. Most patients presented with abnormal bleeding and were treated for conditions unrelated to vasculitis. The different types of vasculitis were: predominantly lymphocytic (nonspecific) 30 cases, necrotizing 17 cases and granulomatous 6 cases. Only 2 patients had additional serologic tests. None of the patients with isolated gynecologic vasculitis received corticosteroids or additional treatment related to the vasculitis. None of the patients developed systemic vasculitis at follow-up (2 months-19.5 years; mean, 5.5 years). Isolated gynecologic vasculitis diagnosed on pathology slides is rarely associated with systemic vasculitis. Potential isolated gynecologic vasculitis causes include: previous surgical interventions and vascular inflammation secondary to local neoplasm. In almost all cases, clinicians did not perform a thorough laboratory analysis to exclude systemic vasculitis and therapy was not required in any case, suggesting minimal clinical significance.

1. Introduction

Vasculitides are chronic inflammatory diseases in which blood vessel walls are targeted by an immune insult. It includes a heterogeneous group of disorders with the common histologic findings of inflammation of the vessel wall. The assumption is that the endothelial injury is the leading event followed by inflammatory infiltrates within and surrounding the vessel wall [1]. The inflammatory infiltrate varies widely with predominance of neutrophils, lymphocytes or a mixed population, and can include eosinophils and/or giant cells. The end result is structural damage to the vessel wall, including necrosis or fibrosis [2].

Vasculitis may affect many organs, including the gynecological tract. Gynecologic vasculitis (GynV), has been reported as single-organ vasculitis (isolated) (IGynV) and, less frequently, in the context of systemic vasculitis [3–5]. The significance of isolated vasculitis and whether isolated cases represent part of true systemic vasculitis or a local inflammatory process is unclear [2].

In this study we reviewed 53 cases of isolated female genital tract vasculitis diagnosed on pathology specimens and correlate with clinical findings and outcome aiming to assess the significance of isolated vasculitis identified histopathologically in the gynecological tract.

2. Materials and methods

A search of cases with vasculitis involving the gynecological tract was performed at the Department of Pathology and Immunology, Washington University in St. Louis from January 1st 1990 until March 31st 2010. The pathologic definition of vasculitis included inflammation that involves the wall of the vessel. Focal necrosis or fibrin deposition within the wall may be present. Adventitial inflammatory infiltrates alone do not represent vasculitis. A review of the pathology reports and available slides was performed. Pertinent morphologic features, including organ involved by vasculitis, type of vessel involved and distribution (focal, diffuse), type of inflammatory infiltrate in and surrounding vessels, presence of eosinophils and/or giant cells or granulomatous changes and the presence of necrosis involving the vessels were recorded from the pathology reports. We extensively reviewed the clinical history, including presenting symptoms, reasons for the surgery and pertinent clinical findings.

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Laboratory data prior and after surgery were recorded to exclude systemic vasculitis. Follow-up information was collected to determine outcome.

3. Results

3.1. Clinicopathologic findings

Fifty-three cases of vasculitis affecting the female genital tract were identified. Data is summarized in Table. The age of the patients ranged from 27 to 85 years (mean, 58.9 years; median, 54 years). All patients had hysterectomy while 37 in addition had bilateral salpingo-oophorectomy and 5 had unilateral salpingo-oophorectomy.

Most of the patients presented with excessive or abnormal vaginal bleeding, while 11 presented with symptomatic pelvic mass; 4 had prior biopsy with high-grade cervical dysplasia or invasive squamous carcinoma. All patients were treated for conditions unrelated to usual vasculitis symptomatology, including 33 patients with uterine tumors, 10 with ovarian tumors and 1 fallopian tube tumor, 4 with cervical dysplasia or squamous carcinoma, 4 prolapse uteri, and one for complications after cesarean section. None had prior symptoms or were diagnosed with generalized vasculitis, while one patient had prior diagnosis of fibromyalgia.

Vasculitis was confined to the cervix in 24 cases, ovaries 7 cases, myometrium 6 cases, fallopian tubes 3 cases, adnexal soft tissue 3 cases and 10 cases showed multifocal involvement (involving more than one organ). In twenty cases (38%), multiple foci of vasculitis were seen in the same organ in which the main pathology was either benign lesion or tumor; in 2 additional cases, a secondary lesion (endometriosis in ovaries) was present concurrent to vasculitis. In 43 (81%) cases, single organ involvement (IGynV) was seen, while in 10 (19%) cases, multiorgan disease was identified (considering the cervix and endomyometrium as different organs based on embryological

Table
Cases of isolated gynecologic vasculitis

Age	Location	Type	Main pathology
51	Cervix	Granulomatous	Ovarian mucinous carcinoma
51	Cervix	Lymphocytic	Endometrial carcinoma FIGO 1
38	Cervix	Necrotizing	Leiomyomas
55	Cervix	Necrotizing	Endometrial carcinoma FIGO 1
69	Cervix	Lymphocytic	Endometrial carcinoma FIGO 3
47	Cervix	Necrotizing	Leiomyomas
41	Cervix	Lymphocytic	Leiomyomas, prolapse
38	Cervix	Lymphocytic	Leiomyomas
44	Cervix	Lymphocytic	Prolapse
41	Cervix	Necrotizing	Leiomyomas, prolapse
51	Cervix	Lymphocytic	Leiomyomas, prolapse
60	Cervix	Lymphocytic	Leiomyomas
64	Cervix	Necrotizing	Endometrial carcinoma FIGO 3
40	Cervix	Lymphocytic	Cervical squamous carcinoma, leiomyomas
66	Cervix	Lymphocytic	Leiomyomas
42	Cervix	Necrotizing	Ovarian cystadenoma, prolapse
61	Cervix	Lymphocytic	Ovarian mucinous adenoma, leiomyoma
49	Cervix	Necrotizing	Leiomyomas
46	Cervix	Necrotizing	Leiomyomas, prolapse
55	Cervix	Granulomatous	Endometrial carcinoma FIGO 3
68	Cervix	Necrotizing	Endometrial carcinoma FIGO 3
52	Cervix	Lymphocytic	CIN 3
45	Cervix	Lymphocytic	Leiomyomas
44	Cervix	Lymphocytic	Cervical/vaginal CIN 2/VAIN 2
58	Cervix and myometrium	Necrotizing	Cervical squamous carcinoma, leiomyomas
82	Cervix and myometrium	Lymphocytic	Fallopian tube carcinoma, leiomyomas
54	Cervix and myometrium	Lymphocytic	Complex atypical hyperplasia, leiomyomas, ovarian endometriosis
85	Cervix and myometrium	Necrotizing	Endometrial mixed carcinoma FIGO 3, leiomyomas
75	Cervix, myometrium, Ov and Ft	Granulomatous	Leiomyomas
37	Cervix and Ft	Necrotizing	Leiomyomas
85	Myometrium	Lymphocytic	Ovarian serous borderline tumor
37	Myometrium	Lymphocytic	Leiomyomas
77	Myometrium	Lymphocytic	Simple hyperplasia, chronic endometritis, leiomyoma
68	Myometrium	Lymphocytic	Carcinosarcoma endometrium
71	Myometrium	Necrotizing	Endometrial carcinoma FIGO 1, leiomyoma
27	Myometrium	Lymphocytic	Placenta accreta
60	Myometrium and ovary	Lymphocytic	Endometrial carcinoma FIGO 2, ovarian serous borderline tumor
69	Myometrium, ovary and Ft	Granulomatous	Ovarian serous carcinoma
63	Myometrium, ovary, Ft and cervix	Lymphocytic	Ovarian mucinous adenoma
74	Myometrium, ovary and vagina	Necrotizing	Prolapse
41	Ovary	Lymphocytic	Leiomyomas
49	Ovary	Necrotizing	Prolapse
44	Ovary	Lymphocytic	Leiomyomas
65	Ovary	Lymphocytic	Leiomyomas
44	Ovary	Lymphocytic	Prolapse
46	Ovary	Necrotizing	Endometriosis ovary and fallopian tube, adenomatoid tumor myometrium
45	Ovary	Lymphocytic	Leiomyomas and ovarian endometriosis
73	Ovarian soft tissue	Granulomatous	Ovarian carcinosarcoma
64	Ovarian soft tissue	Lymphocytic	Ovarian carcinosarcoma
81	Ft soft tissue	Lymphocytic	Metastatic carcinoma to ovary and fallopian tube
61	Fallopian tube	Lymphocytic	Ovarian cystadenoma
39	Fallopian tube	Necrotizing	Leiomyomas
70	Fallopian tube	Lymphocytic	Endometrial serous and clear cell carcinoma extending to bilateral Ov and Ft

FIGO, International Federation of Gynecology and Obstetrics; CIN, cervical intraepithelial neoplasia; VAIN, vaginal intraepithelial neoplasia; Ov, ovary; Ft, fallopian tube.