Myometrial Single Organ Vasculitis. A Rare Entity

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Abstract

Aim: The aim of this study is to present a rare case of Single Organ Vasculitis (SOV) of the uterine corpus.

Case Presentation: We present a 50-year-old patient who underwent TAH/BSO (Transvaginal Abdominal Hysterectomy/Bilateral Salpigoopherectomy) for a known uterine leiomyoma which had recently increased in size. Histology report described an incidental finding of vasculitis of a small myometrial vessel.

Discussion: Single Organ Vasculitis (SOV) has been described in several systems, including female genital tract, and it is a form of vasculitis affecting a single organ, without evidence of systemic disease. It is most commonly an incidental finding in specimens of surgery performed for other gynecologic conditions. Decision to treat depends on the severity and extent of the vasculitis, as its prognosis is most commonly favorable.

Conclusion: Single Organ Vasculitis (SOV) is an unusual pathologic entity of the female genital tract.

Keywords: Vasculitis; Myometrium; Hysterectomy

Introduction

Vasculitis is defined by the presence of inflammatory leukocytes in vessel walls, with reactive damage to mural structures, occurring as a primary process or it may be secondary to other conditions [1].

Noninfectious vasculitides are classified based on the predominant size of the vessels involved (large, medium and small-vessel vasculitides). Single Organ Vasculitis (SOV), previously known as "limited", "localized" or "isolated" refers to vasculitis in arteries or veins of any size in a single organ and it has no features suggesting it is a limited expression of a systemic vasculitis. It has been described in several organs such as the appendix, the skin, the gallbladder and the central nervous system. Some patients who were initially diagnosed with single organ vasculitis may develop other disease manifestations and therefore may be reevaluated for another systemic vasculitis (eg., cutaneous arteritis later becoming polyarteritis nodosa). Here we present a case of isolated uterine vasculitis discovered as an incidental finding in a patient with uterine leiomyomatosis (primer ectopic myometrial vasculitis).

Case Presentation

A 50-year-old premenopausal patient with an insignificant medical or surgical history presented at the Gynecology Clinic of our Department for surgical treatment of a known uterine leiomyoma which had recently doubled in size and caused urinary symptoms (frequency).

Further investigation with a pelvic MRI revealed multiple uterine fibroids, the bigger of which was 7.7 cm × 6.7 cm × 6.5 cm in size. Tumor markers did not depict any signs of malignancy. Patient was led to surgery and underwent a Total Abdominal Hysterectomy and Bilateral Salpingo-Oophorectomy (TAH/BSO), with an uneventful postoperative course.

Histology report was in accordance with the preoperative diagnosis of uterine leiomyomatosis. Besides a few superficial foci of adenomyosis, a notable finding was the presence of a small myometrial vessel with thrombosis, perivascular and focally intramural lymphocytic infiltration as well as necrotic nuclear debris and fibrin deposits. These morphologic features are diagnostic of vasculitis (Figures 1 and 2).

Figure 1: Wide short arrow: Myometrium Cross: Small vessel with muscular wall Long arrow: Inflammatory infiltration and destruction of the vascular wall H&E X200.
Polymorphic inflammatory infiltration (lymphocytes, neutrophils, eosinophils, nuclear dust) H&E X400.

Following, the patient underwent a thorough clinical and laboratory investigation with a Rheumatologist which proved to be negative for systemic vasculitis and she remains in the same condition a year after her surgery.

**Discussion**

Vasculitis represents inflammation of blood vessel walls. Vasculitides are classified either using the American College of Rheumatology Criteria [2] or using the 2012 Chapel Hill consensus classification scheme which is according to vessel size, presence or absence of an associated systemic disease and association with a probable etiology [3].

Systemic vasculitis can present with vague symptoms such as headache and weakness or more specifically with renal failure, arthritis, pulmonary symptoms, infarction, or hypertension [4]. Single organ vasculitis was first described by Plaut et al. [5] who reported 88 cases of incidental vasculitis in the appendix. It is defined as vasculitis in arteries or veins of any size in a single organ, with no features indicating that it is a limited expression of a systemic vasculitis. It is often incidentally diagnosed after biopsy or surgical resection because of suspicion of cancer, infections, or other unrelated abnormalities [6].

However, some of the patients originally diagnosed as having single organ vasculitis will develop additional disease manifestations that warrant reclassifying the vasculitis as one of the systemic vasculitides. Certainty about the accuracy of diagnosis and cure comes only after long-term surveillance [7]. Histopathological findings of single organ vasculitis are similar to systemic vasculitis. Single organ vasculitis may affect small, medium or large sized vessels. Vascular inflammatory patterns may be granulomatous or nongranulomatous. Granulomatous inflammation is defined by the presence of aggregates of lymphocytes and macrophages, with or without giant cells.

In non-granulomatous vasculitis cell types within the infiltrates may be predominantly lymphocytes and/or neutrophils, and vessel wall necrosis may or may not be present. Presence of lymphocytic or neutrophilic infiltrates, necrosis or fibrinoid deposits may reflect stages in the evolution of the same pathologic process [7]. Single organ vasculitis distribution may be unifocal or multifocal (diffuse) within an organ or organ system.

Focal single organ vasculitis has been reported in the breast, aorta and components of the gastrointestinal and genitourinary tracts. Examples of diffuse single organ vasculitis include diffuse cutaneous vasculitis, multifocal peripheral nerve vasculitis, central nervous system vasculitis, renal - limited vasculitis and vasculitis of coronary and pulmonary vessels [8]. Literature on single organ vasculitis of the female genital tract is limited. While it has been described in the uterine corpus, the fallopian tubes, the ovaries and the parametria, it seems that the uterine cervix is the most common site of involvement [9]. Its features may be similar to Polyarteritis Nodosa (PAN), giant cell arteritis or Wegener granulomatosis.

A large retrospective study of hysterectomy and cervical amputation specimens (n=120,000) by Ganesan et al. found that the incidence of single organ vasculitis was 0.04% (n=46), of which thirty lesions (65.2%) were found in the cervix. Of these 46 patients, only four had or developed proven systemic arthritis (polyarteritis nodosa, temporal arteritis or extra genital arteritis). It seems that gynecologic single organ vasculitis often occurs in the setting of other gynecologic comorbidities, some of which may be relevant to the pathogenesis of inflammation of blood vessels, such as inflammation or precancerous lesions. Infectious agents have been implicated in the pathogenesis of vasculitides either by direct infection of the vascular wall or by indirect immunologic pathways, such as hypersensitivity reactions [10].

In the study by Ganesan et al., the most frequent coexisting histopathologic findings were benign leiomyomas (41%) and adenomyosis (26%). In 15.2% (n=7) of the patients’ leiomyomas occurred in isolation and in 17.4% (n=8) of cases leiomyomas were associated with adenomyosis, as it was in our case described here.

Accordingly, the most common malignant disease associated with gynecologic vasculitis had been reported to be endometrial carcinoma [11]. Several other studies report on leiomyomas associated with vasculitis [12-16].

However, in the majority of these studies a GnRH analog had been administered preoperatively, which was not administered to the patient we describe here. Literature data on patients with adenomyosis and concurrent vasculitis are even more limited [17,18].

Evidence on treatment of SOV has emerged from analysis of retrospective case series, as we lack randomized trials.

No universal treatment exists, so treatment decision depends on the distribution of the lesions, underlying etiological factors and disease severity. If there is an underlying etiological factor, withdrawal of the causative medication or treatment of the associated condition may be sufficient for cure. Focal SOV is treated with surgical excision, which is adequate for cure. Diffuse SOV requires immunosuppressive therapy, the regimen depending on the severity of the disease, ranging from non-steroidal anti-inflammatory drugs, colchicine, topical glucocorticoids, dapsone, pentoxifylline and other agents [19].

Prognosis of SOV is most commonly favorable, which is a major distinctive feature of SOV and systemic vasculitis, as literature data suggest that SOV infrequently evolves into systemic disease [19].

**Disclosure of Interest**

All authors declare any financial interest with respect to this manuscript.

**Conclusion**

Single Organ Vasculitis (SOV) of the female genital tract is an unusual pathologic condition. It is most commonly an incidental finding after surgery performed for concurrent gynecologic conditions. Its treatment depends on the severity and extent of the disease and its prognosis in considered favorable.
References


