

Giant cell arteritis of the female genital tract

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Summary

Giant cell arteritis (GCA) of the female genital tract is rare with only 30 cases, including this case, documented in the literature. We present a patient who had total abdominal hysterectomy and bilateral salpingo-oophorectomy for an ovarian cyst and on histologic examination GCA involving the arteries of the myometrium, cervical stroma, ovaries and tubes was unexpectedly discovered. Upon questioning of the patient and further investigations it became clear that the patient did have symptoms and signs suggestive of systemic GCA including fatigue, low-grade fever, weight loss and elevated erythrocyte sedimentation rate (ESR). Treatment with oral corticosteroids resulted in rapid and complete recovery. It is concluded that an incidental finding of GCA in the genitalia should alert the clinician to the possibility of systemic GCA. If upon questioning of the patient and further investigations the existence of systemic GCA is confirmed, treatment with corticosteroids should be considered.

Key words: Giant cell arteritis; Temporal arteritis; Polymyalgia rheumatica; Female genital tract.

Introduction

GCA is a systematic vasculitis that primarily affects large and medium sized arteries [1-7]. Since the most frequently recognized clinical features of GCA are due to involvement of the cranial arteries, particularly the temporal arteries, GCA is also known as temporal arteritis (TA) [1, 3, 5, 7]. GCA is closely related to the clinical syndrome of polymyalgia rheumatica (PMR), which consists of diffuse muscular stiffness and aching affecting the neck, shoulder and pelvic girdles [1, 3]. Over 10% of patients with PMR also have GCA and approximately 50% of patients with GCA also have PMR [1]. The American College of Rheumatology (ACR), in 1990, proposed the following criteria for the classification of GCA [8]: 1) Age \geq 50 years at disease onset; 2) New onset of localized headache; 3) Temporal artery tenderness or decreased temporal artery pulse; 4) Elevated erythrocyte sedimentation rate (Westergren) \geq 50 mm/hour; 5) Biopsy sample including an artery, showing necrotizing arteritis, characterized by a predominance of mononuclear cell infiltrates or a granulomatous process with multinucleated giant cells. At least three of these five criteria are needed for the diagnosis of GCA. GCA is a corticosteroid-responsive disease and corticosteroid therapy (prednisone 1 mg/kg daily) results in rapid and complete control of symptoms in most cases.

GCA of the female genital tract is rare with only 29 cases previously documented in the literature [1-7, 9-23]. We describe an additional case of GCA of the female genital tract and review pertinent literature.

Case report

A 70-year-old, gravida 6, para 2, postmenopausal Ashkenazi Jewish woman was admitted to the hospital in May 1998 because of an asymptomatic left ovarian cyst detected on physical examination. Physical examination disclosed an essentially healthy appearance and normal vital signs. On pelvic examination, a left-sided, non-tender, cystic mass measuring about 12 x 12 x 12 cm was palpated, whereas the rest of the genital organs including the external genitalia, cervix, uterus and right adnexa were normal. Laboratory studies, including CBC, chemistry, serum tumor markers (CA-125, CA-15-3, CA-19-9, CEA, AFP, HCG) were normal. Erythrocyte sedimentation rate (ESR) was not examined. Chest X-ray demonstrated no abnormalities. Transvaginal ultrasound examination and computerized tomography (CT) scan confirmed a left ovarian unilocular cystic mass measuring 12 x 12 x 12 cm. At laparotomy, a left ovarian cyst measuring 12 x 12 x 12 cm with an intact and smooth capsule was identified while no other abnormalities were seen on inspection of the abdomen and pelvis. Total abdominal hysterectomy and bilateral salpingo-oophorectomy were performed.

Pathologic findings. On macroscopic examination, a simple cyst measuring 12 cm in the largest dimension was found to replace the left ovary, whereas the rest of the surgical specimen was unremarkable. Microscopic examination unexpectedly revealed GCA involving predominantly the medium-sized arteries of the myometrium, cervical stroma, ovaries (including the wall of the left ovarian cyst), tubes, paraovarian and paratubal tissues. The inflammatory infiltrate was composed of lymphocytes, histiocytes, epithelioid giant cells and a few plasma cells (Figure 1). Elastic tissue stains underlined the disruption and loss of the elastic fibers of the arteries. Luminal narrowing and complete obstruction was observed in some arteries. Immunohistochemical staining for CD-3, CD-20 and CD-68 confirmed the presence of numerous T cells and histiocytes and a few B cells in the walls of the arteries.

The patient made an uneventful postoperative recovery. In view of the histologic report on the surgical specimen, the patient was specifically asked about symptoms and signs known

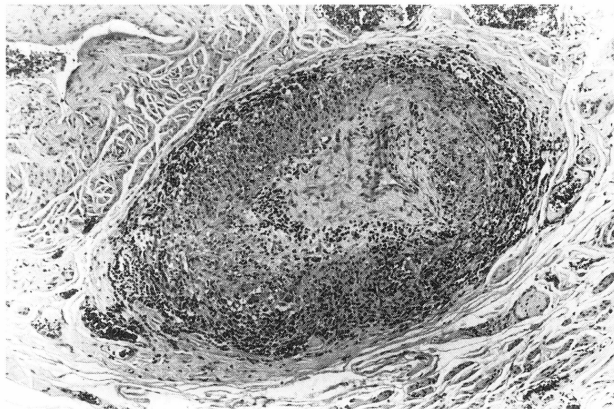


Figure 1. — Giant cell arteritis. Artery in the myometrium showing an inflammatory infiltrate of giant cells, lymphocytes, histiocytes and a few plasma cells, predominantly involving the media. The lumen is compromised by intimal proliferation (hematoxylin-eosin x 100).

to be associated with systemic GCA and/or PMR. She then gave a history of having symptoms suggestive of systemic GCA and/or PMR, including fatigue, low-grade fever and weight loss of three kilograms over the previous six weeks. The ESR was 135 mm/h (Westergren) and the hemoglobin 8 gr %. The rest of the laboratory studies including WBC, platelet count, serum chemistry and protein electrophoresis were normal. Examination of the fundus of both eyes did not reveal abnormal vessels. Both temporal arteries were normal on palpation. The patient refused biopsy of a temporal artery. Treatment with oral corticosteroids (Prednisone 1 mg/kg daily) was started. The patient has tolerated corticosteroid therapy well and ESR has returned to normal. To date, 60 months after detection of GCA of the genital tract, she is alive and well and without symptoms or signs of systemic GCA and/or PMR.

Discussion

To date, only 30 cases of GCA of the genital tract, including this case, have been reported in the literature [1-7, 9-23]. The age of the patients ranged from 40 to 82 years (mean, 67.9) with 23 patients (76.6%) over age 60, five (16.6%) between age 50 and 60, and two (6.6%) below age 50. In one patient with a known systemic GCA, GCA involving the ovaries was discovered at autopsy [19]. In 29 patients (96.6%), GCA of the genital tract had unexpectedly been discovered on histologic examination of surgically removed genital organs for gynecological reasons unrelated to GCA. The reasons for gynecological surgery were: uterine prolapse - six patients [1, 4, 10, 15, 17], fibroid uterus - four [6, 13, 17, 21], ovarian serous cystadenoma - four [7, 9, 16, 18], simple ovarian cyst - four [5, 14, 22, this case], endometrial adenocarcinoma - two [12, 22], cervical carcinoma - two [2, 3], adnexal mass - two [20, 23], endometrial atypical hyperplasia - one [17], ovarian fibroma - one [14], ovarian mucinous cystadenoma - one [14], paraovarian cyst - one [13], and incarcerated vaginal pessary - one [11]. In only one patient, who underwent surgery for ovarian serous cystadenoma, the preoperative severe

lower abdominal pain could retrospectively be attributed to ovarian infarction due to arterial occlusion caused by the GCA [9]. In the rest of the patients, GCA involving the arteries of the genital tract did not appear to lead to any local symptoms. Questioning of the patients and further investigations had elucidated that 25 (83.3%) of the 30 patients had symptoms and signs suggestive of systemic GCA and/or PMR [2-7, 9-17, 19-21, 23, this case] and five had not [1, 17, 18, 22]. Symptoms and signs suggestive of systemic GCA and/or PMR included weakness, myalgia, arthralgia, fever, dyspnea, chest pain, anemia, weight loss, loss of vision, temporal pain, decreased temporal artery pulse and headache. In 15 patients, the symptoms and signs suggestive of systemic GCA and/or PMR were accompanied by high ESR (mean, 102 mm/hour; range, 51 - 135) [1, 3, 5-7, 9, 11, 12, 14-16, 19, 20, this case]. In the current patient, at least three criteria required by the American College of Rheumatology (ACR) for the classification of systemic GCA (age \geq 50 years, ESR \geq 50 mm/hour, and biopsy sample showing GCA of an artery) have been fulfilled [8]. Seven patients (23.3%) had biopsy of the temporal artery confirming temporal arteritis; in six patients the biopsy was performed after gynecological surgery [4, 5, 7, 16, 19, 20] and in one patient the biopsy was taken six months before gynecological surgery [18].

In patients already known to have systemic GCA and/or PMR, the finding of GCA in the genital tract poses no additional problem in management since it may serve to confirm active systematic disease as well as the requirement for continued treatment with corticosteroids [17]. In patients with vague or nonspecific symptoms and signs weakly suggestive of systemic GCA and/or PMR, the symptoms and signs take on additional significance in view of the finding of GCA in the genital tract and further investigation to confirm systemic GCA and/or PMR is prompted [16]. In patients without symptoms and signs suggestive of systemic GCA and/or PMR, the unexpected finding of GCA of the genital tract should alert the clinician to the diagnosis of systemic GCA and/or PMR and questioning of the patient and further investigations should be undertaken [15, 2, 3]. If questioning and further investigations do not reveal systemic GCA and/or PMR, it may be assumed that GCA is limited exclusively to the female genital tract.

In conclusion, GCA affecting the small to medium-sized arteries of the female genital tract of elderly women is a rare finding that should alert the clinician to the possibility that it may be a manifestation of GCA involving multiple sites. If upon questioning and further investigations it is confirmed that GCA of the female genital tract is part of systemic GCA and/or PMR, treatment with corticosteroids should be considered.

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