Case Report

Eosinophilic Perifolliculitis Presenting as an Incidental Finding in a Patient with Leiomyomata with Perinodular Hydropic Degeneration

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Abstract

Eosinophilic perifolliculitis is a rare disease entity and is considered a possible variant of autoimmune oophoritis. Here we report a 49-year-old, nulliparous female presented with menorrhagia and markedly enlarged uterus. Incidentally, the right ovary showed numerous eosinophils infiltrating a hemorrhagic corpus luteum consistent with eosinophilic perifolliculitis. It is important to recognize eosinophilic perifolliculitis histologically because of its possible relation to autoimmune oophoritis which can cause primary ovarian failure.

Keywords: Eosinophilic perifolliculitis; Autoimmune oophoritis; Primary ovarian failure

Abbreviations

CD - Cluster of Differentiation; HPF - High Power Field; FSH - High Follicle-Stimulating Level; IL - Interleukin

Background

Eosinophilic perifolliculitis is a rare disease entity and is considered a possible variant of autoimmune oophoritis. With minimal discussion in the surgical pathology and gynecologic literature, the pathogenesis and natural history of eosinophilic perifolliculitis are not well-known. Some of the reported cases in the literature have been associated with autoimmune diseases.

Case Presentation

A 49-year-old, nulliparous female presented with menorrhagia and an enlarged uterus suspicious for leiomyosarcoma or endometrial tumor. Pelvic and transabdominal ultrasound revealed a markedly enlarged uterus equivalent to mid-term pregnancy measuring 21.0 x 11.0 x 17.6 cm. The right ovary is enlarged measuring 6.7 x 4.4 x 4 cm with no evidence of cystic change or mass. The left ovary is not visualized. Subsequently, she underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy.

Gross examination showed an enlarged uterus weighing 2150 grams and a huge mostly intramural mass with multinodular white-tan fleshy areas with myxoid and cystic change measuring 17.0 x 9.0 x 4.5 cm. The cut-surface had multiple smaller nodules; some of the nodules were separated by mucoid gelatinous soft tissue. There were no areas of hemorrhage, calcification, or necrosis present. The endometrial cavity was distorted, compressed and lined by unremarkable endometrium with a small benign endometrial polyp. The bilateral ovaries and fallopian tubes appeared grossly unremarkable. Histologically, the uterus showed benign leiomyomata with perinodular hydropic degeneration and myxoid change. There was no cytologic atypia with rare mitotic activity (<1 mitosis/10HPF). The hydropic change within leiomyoma with had some thick-walled blood vessels and thin cords of residual smooth muscle cords in with an edematous pale background. The presence of extensive hydropic change may raise the suspicion for myxoid leiomyosarcoma in some difficult cases. This may also explain the clinical-radiologic suspicion of leiomyosarcoma. Incidentally, the right ovary showed abundant inflammatory cells predominantly eosinophils infiltrating a hemorrhagic corpus luteum only. There are no prominent primordial follicles present. The ovarian stroma was not involved by the inflammatory infiltrates.

Figure 1&2: Hemorrhagic corpus luteum surrounded by infiltrate of abundant eosinophils, lymphocytes, macrophages and plasma cells. (H&E, original magnification x 20 [1] and x 200 [2]).

The pathogenic mechanism of autoimmune oophoritis has not been well studied [3]. The immunophenotype of the ovarian inflammatory cells infiltrates revealed a mixture of B cells, plasma cells, T cells, macrophages and natural killer cells suggesting a complex immune process with interaction of humoral and cellular mechanisms is involved in the pathogenesis [4]. There is some evidence that CD4 type 1 helper cells are the major pathogenic T-cell [3]. In addition, there is evidence that CD4 T helper 2 cells also elicit autoimmune oophoritis with autoreactive T cell producing IL-4 and IL-5 which are key mediators in eosinophil activation [3]. This mechanism elucidates the mechanism of eosinophilic perifolliculitis and its relation to autoimmune oophoritis. It also explains the few cases with predominant of eosinophilic infiltrates [5,6] including this case. Interestingly, enteric nematodes (rodent pinworm) which can cause primary ovarian failure and its frequent association other autoimmune diseases and the eosinophilic perifolliculitis is an incidental finding. The perifolliculitis may be explained by the presence of leiomomas. Her ovaries were grossly unremarkable as may be seen in the early stage of autoimmune oophoritis.

Autoimmune oophoritis typically occurs in the setting of autoimmune polyendocrine syndromes and is commonly associated with Addison’s disease, hypothyroidism, hyperparathyroidism and diabetes mellitus [1]. Although our patient clinical history is not significant for other autoimmune diseases and the presence of circulating autoantibodies is unconfirmed, this case may represent a variant autoimmune oophoritis. Her father having celiac disease is of interest given that autoimmune diseases have a strong hereditary component and may cluster in families as different illnesses. It is essential to recognize eosinophilic perifolliculitis histologically because of its possible relation to autoimmune oophoritis which can cause primary ovarian failure and its frequent association other autoimmune diseases.

References