

Large Vessel Vasculitis as a Rare Paraneoplastic Effect of Endometrial Cancer

Erin C. Saricilar*

Department of Surgery, Liverpool Hospital, Sydney, Australia

Abstract

A case of large vessel vasculitis as a paraneoplastic syndrome of endometrial cancer is presented which is the first documented case in the literature. Presenting initially as undifferentiated abdominal pain, routine tests presented no clear diagnosis beyond aortitis. PET scan and biopsy showed a non-Mullerian origin endometrial cancer, with the aortitis a likely paraneoplastic syndrome. This syndrome has been demonstrated in other cancers such as myelodysplastic syndromes albeit rarely, but this case opens up differential diagnoses for unexplained large vessel vasculitis cases.

Keywords

Aortitis • Large vessel vasculitis • Paraneoplastic syndrome • Endometrial cancer

Introduction

Many cancer syndromes have paraneoplastic sequelae that present as a vasculitis. Myelodysplastic syndromes can rarely present with large vessel vasculitis as a paraneoplastic syndrome but this has not been documented in the case of endometrial cancer. While there is limited understanding of the pathophysiology of paraneoplastic syndromes as vasculitis it is suspected to be due to immune dysregulation. An understanding of this would present the opportunity for a targeted approach to large vessel vasculitis.

A case of endometrial cancer masquerading with an aortitis is presented in order to bring attention to a previously undocumented paraneoplastic syndrome for endometrial cancer. The patient's family have consented to the publication of this report.

Case Report

A 69 years old woman presented with a four day history of nausea, vomiting and loss of appetite, with non-peritonitic generalised abdominal pain. There were no changes in bowel habits. Relevant previous medical history includes a diagnosis of polymyalgia rheumatica (PMR) two years prior, and a recent history of complex migraines (temporal arteritis was not diagnosed) and treated appropriately with long term steroids. Vital signs demonstrating tachycardia of 112, tachypnoea of 24 and low grade fever at 37.6°C. Blood pressure and oxygen saturation was normal. The abdominal tenderness was generalised, but worst in the epigastrium.

Initial investigations were non-specific, though a CT scan with contrast

**Address for Correspondence:* Erin C. Saricilar, Department of Surgery, Liverpool Hospital, Sydney, Australia, Tel: +610433305302; E-mail: erin.c.saricilar@gmail.com

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showed inflammation of the abdominal aortic wall consistent with aortitis (Figure 1). While initially considered as infective in aetiology, empirical antibiotics did not lead to improvement in symptoms. A PET scan showed aortitis, but more significantly a previously asymptomatic uterine cancer with vertebral and liver metastases (Figure 2). The patient was ultimately placed on a palliative pathway. Interestingly, the CT scan also showed a unique coeliac trunk with a splenogastric trunk and common hepatic artery coming directly off the aorta—a Morita type III anomaly only seen in 0.23% of patients (Figure 1) [1].



Figure 1. CT scan with contrast showing inflammation in wall of aorta and Morita class III coeliac trunk anomaly.

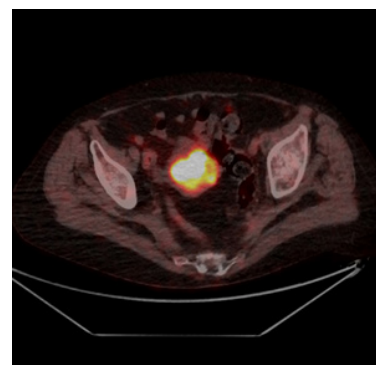


Figure 2. PET scan showing endometrial cancer.

Privity to the presence of cancer, tumour markers were added on with elevations in CA 15.3 (45.4 kU/L), CA 19.9 (45 kU/L) and CEA (7.3 µg/L). The CRP at this point was 94.9 mg/L, with the patient positive for cryoglobulins, predominantly alpha 1 globulins being elevated at 2.9 g/L. Rheumatoid factor was below 13 IU/mL while ANCA levels were below 20.

Blood film staining and microscopy presented a leukoerythroblastic film. Tissue curettage described a poorly differentiated carcinoma of endometrial origin with foci of squamous differentiation. It was notably PAX8 negative, suggesting a non-Müllerian origin, but was positive for MNF116 and p40.

Results and Discussion

Aortitis and other large-vessel vasculitis' are documented albeit rare paraneoplastic syndromes in the context of myelodysplastic syndrome. Small vessel vasculitis' are more thoroughly reported [1]. Although the pathogenesis of these conditions is unclear, they are often associated with observable immune system dysregulation. There has never been documentation of a large vessel vasculitis as a paraneoplastic syndrome of endometrial or uterine cancer.

Typically, endometrial cancer presents with irregular uterine bleeds, particularly in postmenopausal women where it is a slowly progressing malignancy often insidious until terminal [2]. Other symptoms can include menorrhagia, anaemia, and constitutional symptoms of cancer. While usually defined as an oestrogen-dependent proliferative endometrioid adenocarcinoma [2], there are reports of rarer subtypes such as adenosquamous and small cell carcinoma [3-5]. Small cell carcinoma in the endometrium may have ocular abnormalities as a paraneoplastic syndrome [4,5]. Adenosquamous carcinoma may cause hypercalcaemia due to prostaglandin-mediated stimulation of osteoclastic bone resorption [3]. Paraneoplastic neurologic syndromes may also occur in gynaecological and breast carcinomas, albeit rarely [6].

The unclear pathophysiology behind large vessel vasculitis as a paraneoplastic syndrome limits a targeted therapeutic approach. There have been suggestions of deficiencies in the immune response from lymphocytes, natural killer cells and phagocytic cell function or due to persistent immune stimulation (7). Despite this, long term steroid treatment is successful in yielding a positive response (8,9). This may be due to a generalised dysfunction in immune cells, as well as the occult production of cytokines such as interferon-alpha and interferon regulatory factor 1 [7-9].

A PET scan was used for the diagnosis in this case, but in future, tumour markers in the context of unexplained vasculitis (particularly as small vessel vasculitis is more often documented in other malignancies, specifically myelodysplastic syndrome) may be used to identify malignancy as a potential primary cause.

Conclusion

Vasculitis is a complex and rare paraneoplastic syndrome of some cancers, notably myelodysplastic syndromes. This case presents the first documented case of a large vessel vasculitis (specifically aortitis) as a paraneoplastic syndrome of endometrial cancer. It suggests the option to consider tumour markers in unexplained vasculitis. A PET scan can also be used as a diagnosis modality as it will help support an infective cause for the diagnosis as well.

Patient Consent

As the patient had unfortunately passed away prior to this report being written, signed consent was given by her family for the report to be written and published in an effort to educate the greater medical community.

Competing Interests

Authors have declared that no competing interests exist. All authors contributed towards the completion of this manuscript. All authors have read and approved the final manuscript.

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