Granulomatosis with polyangiitis (GPA, formerly Wegener granulomatosis) is a vasculitis with various organ involvements. Involvement of the cervix, urethra and vagina was reported in less than 1% of patients [1].

We here report a lady 7 months post-Cesarean section presented with lower abdominal pain, and abnormal uterine bleeding. She underwent total hysterectomy due to mass-like lesion of the uterus and last diagnosis of GPA presenting with cervical necrotizing vasculitis. So far, only 10 cases have been reported with this rare manifestation [2] (Table 1).

Table 1
Cases of gynaecological involvement in granulomatosis with polyangiitis.

<table>
<thead>
<tr>
<th>Ref</th>
<th>Age</th>
<th>Initial presentation</th>
<th>Diagnosis delay</th>
<th>ANCA Titer</th>
<th>Treatment</th>
<th>Diagnostic method</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>45</td>
<td>Unspecified</td>
<td>Undisclosed</td>
<td>C-ANCA 1/160</td>
<td>Cyclophosphamide and prednisolone</td>
<td>Pap smear and biopsy</td>
<td>Resolved in 3 months</td>
</tr>
<tr>
<td>2</td>
<td>82</td>
<td>Cervix then lung</td>
<td>4 years</td>
<td>C-ANCA 1/320</td>
<td>Cyclophosphamide and prednisolone</td>
<td>Lung and cervix relapse</td>
<td>Favourable</td>
</tr>
<tr>
<td>3</td>
<td>32</td>
<td>Nose and pulmonary</td>
<td>1 year</td>
<td>C-ANCA 1/640</td>
<td>Cyclophosphamide and prednisolone</td>
<td>Biopsy</td>
<td>Lost to follow-up</td>
</tr>
<tr>
<td>4</td>
<td>80</td>
<td>Dialysis</td>
<td>16 months</td>
<td>C-ANCA 1/800</td>
<td>Not available</td>
<td>Hysteroscopic biopsy</td>
<td>Died day after biopsy</td>
</tr>
<tr>
<td>5</td>
<td>64</td>
<td>Unspecified</td>
<td>3 weeks</td>
<td>Anti-PR3 14 IU/mL</td>
<td>Cyclophosphamide and prednisolone</td>
<td>Cervix biopsy, lung disease</td>
<td>Resolution</td>
</tr>
<tr>
<td>6</td>
<td>42</td>
<td>ENT and cervix</td>
<td>5 years</td>
<td>Anti-PR3 &gt; 100 IU/mL, PR3+</td>
<td>Cyclophosphamide and prednisolone</td>
<td>ENT involvement</td>
<td>Two year remission</td>
</tr>
<tr>
<td>7</td>
<td>71</td>
<td>Not available</td>
<td>1 year</td>
<td>C-ANCA 1/160</td>
<td>Surgery and azathioprine</td>
<td>ENT disease</td>
<td>Surgery ineffective, good result with azathioprine</td>
</tr>
<tr>
<td>8</td>
<td>61</td>
<td>ENT</td>
<td>Undisclosed</td>
<td>Not available</td>
<td>Prednisolone</td>
<td>Biopsy re-read after radiotherapy</td>
<td>Died five years later, unknown radiotherapy cause</td>
</tr>
<tr>
<td>9</td>
<td>55</td>
<td>Nose and URT</td>
<td>First work-up</td>
<td>C-ANCA 1/160</td>
<td>Cyclophosphamide and prednisolone</td>
<td>Biopsy and ENT symptoms</td>
<td>Resolved in one month</td>
</tr>
<tr>
<td>10</td>
<td>34</td>
<td>Skin, joints, sinus</td>
<td>19 months</td>
<td>Anti-PR3 101 IU/mL</td>
<td>Rituximab</td>
<td>Conization</td>
<td>Near resolution in six months</td>
</tr>
<tr>
<td>Our case</td>
<td>33</td>
<td>Lower abdominal pain, abnormal uterine bleeding</td>
<td>2 months</td>
<td>C-ANCA (u/mL)+: 4.98 (nl: 0.0–0.5)</td>
<td>Cyclophosphamide and prednisolone</td>
<td>Hysterectomy and skin biopsy</td>
<td>Partial remission in 6 months</td>
</tr>
</tbody>
</table>

ENT: ear, nose and throat; URT: upper respiratory tract.

In May 2015, a 33-year-old lady was transferred from general surgery ward to medical ward with cough, fever, and oral and skin lesions. The patient was well until about 2 months prior to admission when developed progressive lower abdominal pain and abnormal uterine bleeding. Abdominopelvic sonography and computed tomography (CT) showed enlarged uterus due to a heterogeneous lesion measuring 106 in 77 millimetre with fluid attenuation that occupied almost entire endometrial body. At operation the uterus was friable, necrotic and contained a malodorous fluid collection, so total abdominal hysterectomy was performed. Two days after the operation, on broad-spectrum antibiotic, pussy discharge from the site of operation was observed and she developed progressive cough and dyspnea. Subsequent pelvic CT showed multiple fluid collections in the pelvic cavity. After two days, she developed oral ulcer, painful papule and plaque lesions predominantly in the lower extremities. Patient was transferred to the medical ward.

On past history, she denoted nose bleeding and crusty nasal discharge following cesarian section about 7 months ago; it was diagnosed as rhinitis. Also, she had a history of infertility for 10 years and a successful in vitro fertilization was done for her 1.5 years ago.

http://dx.doi.org/10.1016/j.jbspin.2015.12.006
1297-319X(© 2016 Société française de rhumatologie. Published by Elsevier Masson SAS. All rights reserved.)
On arrival, she was febrile, acutely ill and had respiratory distress. There were multiple large deep oral ulcers. Multiple papule and bullous lesions were seen on both lower extremities (Fig. 1a).

Chest radiography (Fig. 1b) and computed tomography (CT) showed complete right upper lobe necrotic consolidation, two peripheral round necrotic mass involving the left and right lower lobes. Also, a few smaller nodules were seen through the lung parenchyma. Sinus CT showed bilateral ethmoid and sphenoid sinusitis. C-ANCA titer was positive at high titer. ESR was 140 mm/h and she had 2.7 gram proteinuria in 24-hour urine collection. Review of the pathologic slides showed chronic endometritis, abscess formation with severe acute and chronic inflammation of the serosa and myometrium consistent with pelvic inflammatory disease. The cervix showed stromal medium-sized vessels necrotizing vasculitis rich in neutrophil and eosinophil with fibrinoid necrosis of the vessel wall (Fig. 1c and d). Pathologic result of skin lesions showed neutrophilic vasculitis of small blood vessels.

GPA was diagnosed. Standard intravenous methyl prednisolone pulse and cyclophosphamide was administered. Two weeks later she was generally well, afebrile without respiratory symptom.

It seems that GPA has predilection to involve the uterine cervix rather than the corpus [1,3,4]. In our case, review of available slides revealed only chronic endometritis, abscess formation with severe acute and chronic inflammation of serosa and myometrium consistent with pelvic inflammatory disease. Although in cervix necrotizing vasculitis of medium size vessels along with fibrinoid necrosis was seen.

Therefore, the goal of the this report was to present a rare, misdiagnosed manifestation of GPA presenting with unusual menometorrhagia to help earlier diagnosis of this disease and prevent unnecessary total hysterectomies.

Disclosure of interest

The authors declare that they have no competing interest.

Acknowledgements

The authors would like to thank Center for Development of Clinical Research of Nemazee Hospital and Dr. Nasrin Shokpour for editorial assistance.

References


![Image](image1.jpg)
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Accepted 2 December 2015  
Available online 26 May 2016