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| **Additional file 1** Summary including the present case and previously reported cases with preceding gastrointestinal symptoms |
|  | **Present case** | **Case 1** | **Case 2** | **Case 3** |
| **Age (y)** | 14 | 30 | 43 | 79 |
| **Sex** | male | male | male | female |
| **Chief complaints** | bloody stool, fever | bloody stool, fever, oral aphtha | bloody stool, fever, nausea, weight loss | bloody stool |
|  |
| **Location of gastrointestinal lesion** | cecum - right transverse colon | right transverse colon | cecum - hepatic flexure | total colon |  |
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| **Histological findings**  | granulomatous inflammation | non-granulomatous inflammation | non-granulomatous inflammation | non-granulomatous inflammation |  |
|  |
| **Complications** | cardiac tamponade | skin ulcer | skin ulcer, renal damage, lung lesion | renal damage, alveolar bleeding |  |
|  |
| **PR3-ANCA** | positive | positive | positive | positive |  |
| **Treatments** | PSL, AZA, MTX | PSL, AZA | PSL, cyclophosphamide | PSL, cyclophosphamide, methylprednisolone pulse, plasmapheresis |  |
|  |
| **Remission maintenance** | good | good | good | good |  |
| **Reference** |  | Yoshikawa (2017) [14] | Morchón-Simón (2011) [15] | Qian(2010) [16] |  |
| Gastrointestinal symptoms preceded in only three adult cases, and the source of bleeding in all cases was the colon. Two of three cases rapidly developed renal dysfunction and alveolar hemorrhage later. |  |