**Introduction**—Atypical hemolytic uremic syndrome (aHUS) secondary to vasculitis is a rare but serious complication.

**Patients**—We described two patients and review previous 32 reported cases of patients with aHUS associated to small vessel renal vasculitis (SVV).

**Results**—Case 1.—A 70-year-old male with a diagnosis of microscopic polyangiitis (MPA) presented because of worsening kidney function to end stage renal disease (ESRD) and appearance of aHUS with C3 serum reduction. Kidney biopsy showed SVV. Steroids, cyclophosphamide and PE were started. Hemolysis recovered. After 20 months follow up he started dialysis.

Case 2.—A 79 years old male with a recent diagnosis of MPA presented for aHUS. He had C3 serum severe reduction. Kidney biopsy showed SVV, acute thrombotic microangiopathy associated and C3 positive immunofluorescence. PE, steroids and cyclophosphamide were started. Hemolysis recovered but kidney function never improved and the patient died because of pneumonia.

All the patients were negative for genetic mutations in the complement pathway (CFH, CFB, MCP, CFI) and for anti-CFH antibodies. ADAMTS 13 activity was normal.

**Discussion**—aHUS is associated to the most severe cases of renal vasculitis; despite the addiction of PE to burn out hemolysis the outcome was negative. In the literature review we identified 32 cases (Supplementary data). Similarly to our cases, 81% of the patients were treated with PE; nevertheless 61% of the patients died or presented ESRD. Alternative complement pathway (AP) hyperactivation seems to play an important role in SVV; indeed disease progression could be prevented by C3 depletion in animal models and human neutrophils involved in ANCA vasculitis.

**Conclusion**—All the patients with SVV with sudden deterioration of kidney function, anemia and thrombocytopenia should be suspected to have aHUS. Starting an early appropriate treatment with PE or complement blocking agents such as eccluzamab could improve patient’s morbidity and mortality.

**Table I Cases report of vasculitis syndrome aHUS associated**

<table>
<thead>
<tr>
<th>Case</th>
<th>Details</th>
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<tbody>
<tr>
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**Case 2**

A 70-year-old male with a recent diagnosis of MPA presented for aHUS. He had C3 serum severe reduction. Kidney biopsy showed SVV, acute thrombotic microangiopathy associated and C3 positive immunofluorescence. PE, steroids and cyclophosphamide were started. Hemolysis recovered but kidney function never improved and the patient died because of pneumonia.

**Discussion**

All the patients were negative for genetic mutations in the complement pathway (CFH, CFB, MCP, CFI) and for anti-CFH antibodies. ADAMTS 13 activity was normal.

**Conclusion**

Severe AH was more commonly associated with PR3-ANCA (vs. MPO-ANCA) and strongly correlated with renal vasculitis. Current treatment of severe AH leads to remission but long-term mortality remains high. Concurrent renal failure and older age were associated with higher mortality.

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