

Pulmonary Medicine

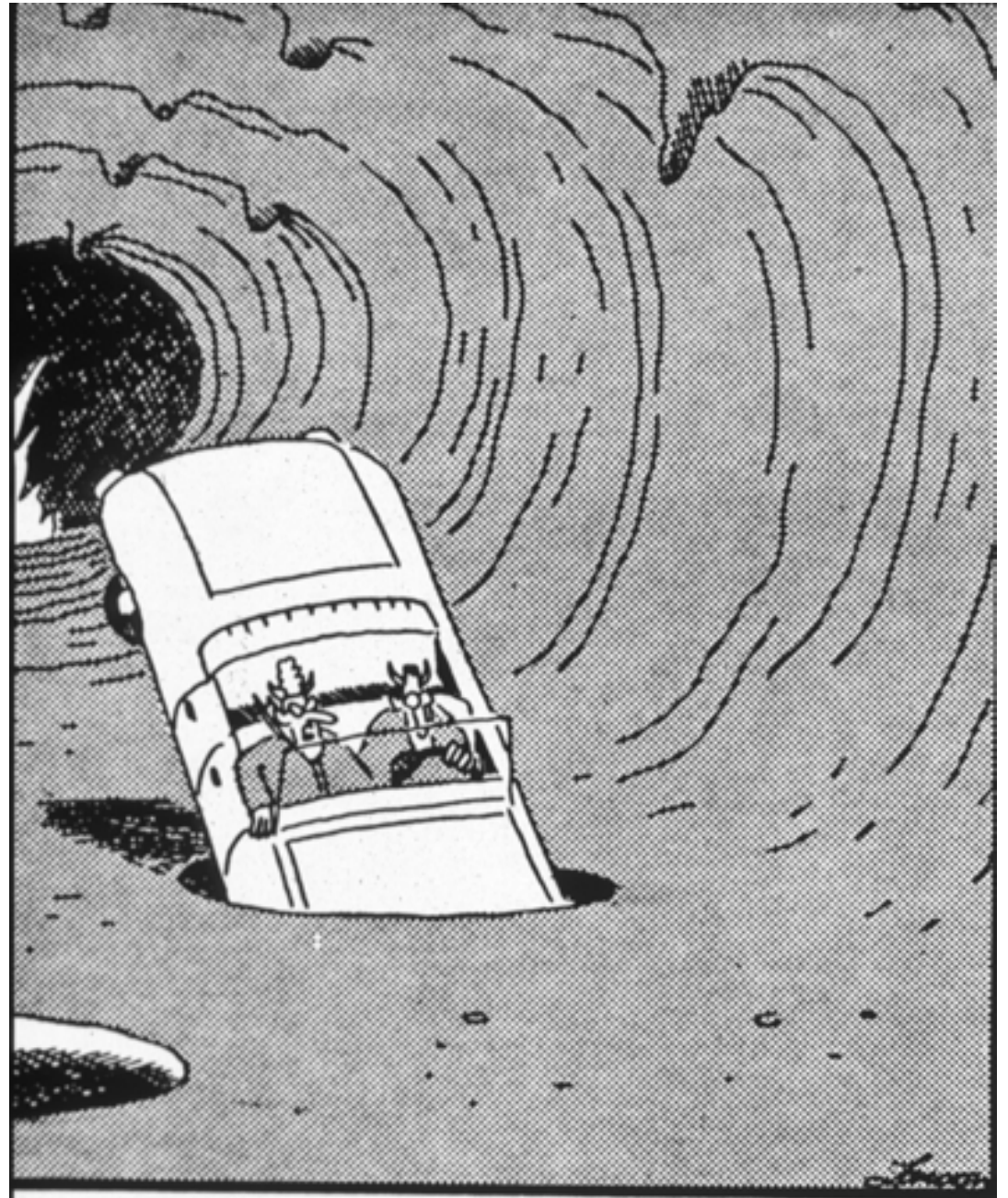
Grand Rounds

1/18/02

“Coagulopathies in Intensive
Care Medicine”

Marcus E. Carr, Jr., MD, PhD

**Nothing
Spells
Hell-Hole
Like
TTP.**



**"You moron! From a hundred yards back
I was screaming, 'Hell-hole! Hell-hole!'"**

“Coagulopathies in Intensive Care Medicine”

Thrombotic Thrombocytopenic Purpura (TTP)

Thrombotic Thrombocytopenic Purpura

- **Introduction**

- **Uncertain etiology but frequently preceded by a viral illness**
- **Characterized by**
 - **rapid onset**
 - **high mortality if not treated (>90%)**
 - **fibrin-platelet plugging of small vessels**
 - **very low platelet count**
 - **little evidence of activation of the coagulation system**

The Pentad of TTP



Clinical manifestations of the Pentad

- **Anemia - traumatic hemolytic**
 - low hematocrit
 - fragmented red cells
- **Renal Failure**
- **CNS disturbances (wide variety and degree of severity)**
 - confusion, headache, photophobia, seizures
 - focal abnormalities
- **Fever**
- **Thrombocytopenia**
 - due to consumption
 - bone marrow biopsy reveals numerous megakaryocytes

Major Differential In TTP Is DIC

- Coagulation Screens
 - Normal in TTP
 - Abnormal in DIC
- Fibrinogen Level
 - Normal in TTP
 - Low in DIC
- Fibrin Split (Degradation) Products
 - Minimal or no elevation in TTP
 - Elevated in DIC

Making the Diagnosis of TTP

- **Appropriate clinical presentation (at least part of the pentad)**
- **Laboratory data as above**
- **Biopsy**
 - typically gingival
 - reveals fibrin-platelet thrombi in small vessels
 - no evidence of surrounding inflammation
 - no associated immunoglobulin or complement

Platelet-fibrin Plug in Small Vessels on Biopsy

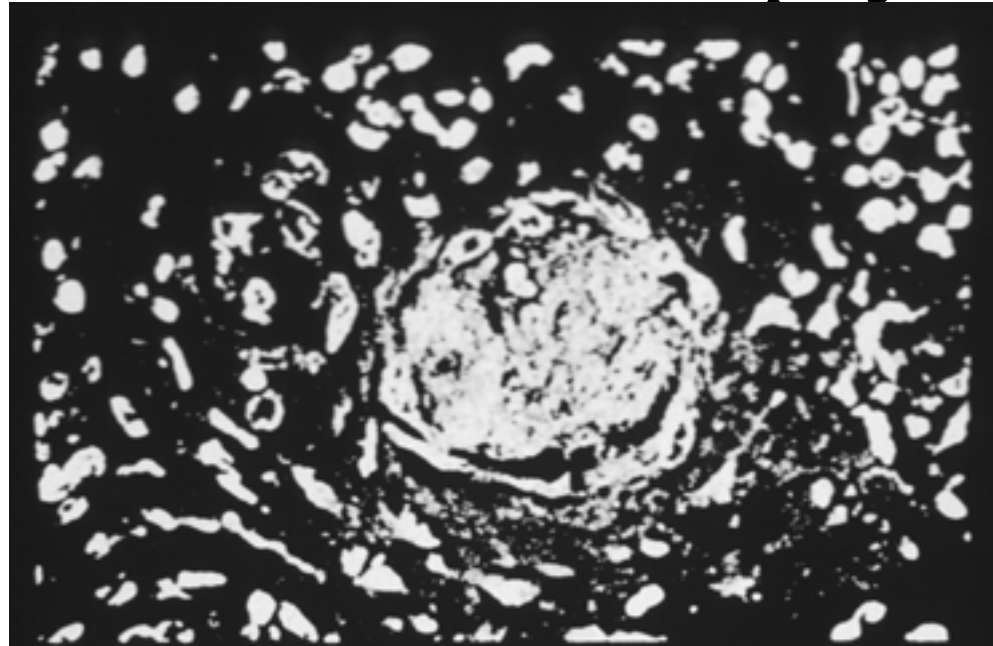


Fig. 14-3. Fibrin-plugged arteriole in gum biopsy from a patient with TTP. Note absence of inflammatory response around arteriole. (From Pisciotta, A.V. & Gottschall, J. (1980) Clinical features of thrombotic thrombocytopenic purpura. *Semin. Thromb. Hemostasis* 6:330–340.)

The Anemia In TTP Is Microangiopathic

- Red Cell Fragments Are Seen

Laboratory Evidence of TTP

Clotting abnormalities

Low platelets

Increased megakaryocytes

Clotting tests otherwise normal

Traumatic hemolytic anemia

Low hematocrit and hemoglobin

Red cell fragments on smear

Bilirubinemia

Sometimes hemoglobinuria

Treatment of TTP

- **Large scale plasmapheresis or plasma exchange**
- **Decreases mortality to <50%**
- **Unproven treatments of questionable benefit**
 - **plasma infusion**
 - **steroids**
 - **anti-platelet agents**
 - **emergency splenectomy**

Hemolytic Uremic Syndrome (HUS)

- **Usually seen in children or pregnant women**
- **Can occur in epidemic forms - almost certainly has infectious etiology**
- **Clinically looks like TTP involving only the kidney**
 - **No CNS symptoms**
 - **Marked renal impairment**
 - **low mortality (virtually none if treated)**
- **Treatment is dialysis - usually followed by spontaneous recovery**