ANSWER TO PHOTO QUIZ (PAGE 304)
PETECHIAE ON THE HEART

DIAGNOSIS

Postmortem evaluation shows an ADAMTS13 activity of only 2% (normal 30-200) and autoantibodies against vWF protease, confirming one of our originally suspected diagnoses: Thrombotic Thrombocytopenic Purpura (TTP). It was presumed that extensive clotting and hemorrhagic changes in the myocardium were the principal cause of death.

TTP is an acute life-threatening illness with thrombosis in small blood vessels. TTP is an illness of all ages, mostly seen in adults, and in about 75% of cases in women.[1] The majority of cases are based on auto-immune antibodies against an inhibitory factor of blood clotting (the ADAMTS-13). With the resulting intravascular clotting there is an immense consumption of thrombocytes, and hemolysis develops due to mechanical damage of red blood cells, among other things leading to the occurrence of schistocytes. Multi-organ failure develops, petechiae and purpura can arise, either spontaneous or after only minor trauma because of the deep thrombocytopenia.

We initially suspected that the diarrhea in this case might be caused by a shiga toxin producing bacterium, causing a hemolytic uremic syndrome. Post-mortem feces analysis did not confirm this suspicion. Whether the diarrhea was related to another type of infection or caused by gut wall damage through thrombotic complications due to the ADAMTS13 deficiency remains open for discussion.

Untreated TTP has an estimated mortality of 90%.[1] Under normal circumstances, treatment consists of plasma-exchange (PEX), which provides functioning ADAMTS13; such treatment should be started immediately. When PEX cannot be started, plasma transfusion can be considered. Presentation with the classic five symptoms (microangiopathic hemolytic anemia, thrombocytopenia, fever, acute kidney failure and neurological symptoms) probably occurs in only 5% of cases.[1]

Thanks to: dr. J.E. Boers, pathologist

REFERENCE

1. George JN, Al-Nouri ZL. Diagnostic and therapeutic challenges in the thrombotic thrombocy-topenic purpura and hemolytic uremic syndromes. Hematology Am Soc Hematol Educ Program. 2012:604-9.