Fever in a female patient with a giant hepatic hemangioma - an incomplete Kasabach-Merritt phenomenon

J. Celeiro¹, B. Flueckiger², A. Schnider³, L. Aceto¹, P. Komminoth⁴, M. Weber¹

¹Department of Visceral, Vascular and Thoracic Surgery, ²Department of Rheumatology, ³Division of Infectious Disease, ⁴Institute of Pathology, City Hospital Triemli, Zurich, Switzerland

Objective:
Giant liver hemangiomas are defined by a diameter larger than 5 cm and can rarely lead to an inflammatory syndrome. The Kasabach-Merritt phenomenon (KMP) is an uncommon complication of giant hemangiomas with disseminated intravascular coagulation and can exceptionally be associated with visceral hemangiomas in adults. Clotting, fibrinolysis and anatomic necrotic changes within the hemangioma are thought to cause the coagulopathy and inflammatory response.

Methods:
We report the case of a 48-year-old female patient presenting recurrent fever, anemia, epigastric pain and substantial weight loss within a period of 6 weeks. Infections, cancer or autoimmune disease were ruled out. The laboratory testing for a disseminated intravascular coagulopathy was negative. Liver function tests and further blood cell counts were normal. The computed tomography revealed a giant hepatic hemangioma in the right lobe of 13.5 cm in size as the possible cause of the fever syndrome. An interdisciplinary conference decided surgery as a therapy.

Results:
The patient underwent a right heptectomy through a transverse upper laparotomy and a markedly enlarged liver was detected. The right heptectomy was performed without intermittent clamping (Pringle manoeuvre). The weight of the resected specimen was 1351 g. The pathological analysis confirmed the diagnosis of a benign cavernous hemangioma with evidence of thrombosis and necrotic defects. After the treatment the symptoms resolved without further complications. The patient was discharged on the postoperative day 12 afebrile and recovering from anemia.

Conclusion:
KMP is very rare, but should be regarded as one of the causes of fever of unknown origin. In this case we found an incomplete KMP with a histological proved thrombosis and necrosis in the hemangioma and recurrent fever. Several treatment options for giant hepatic hemangiomas exist: surgical resection, embolisation, treatment with blood product support and heparin. While a surgical excision is the most invasive option, it constitutes (if technically feasible) the most effective alternative and is indicated in patients with abdominal and mechanical complaints, complications or when diagnosis remains inconclusive. For complex cases, an interdisciplinary approach is crucial to sound indication and excellent outcome.