Cristina Emilia Ursu, MD, PhD

Romanian Academy of Medical Sciences, Onco-Hematology Research Unit, Children Emergency Hospital "Louis Turcanu" Timisoara, European Hemophilia Treatment Centre, 2nd Dr. Iosif Nemoianu, 300011 Timisoara, Romania

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HEMOPHILIC PSEUDO-TUMORS - A SINGLE CENTER RETROSPECTIVE ANALYSIS

Cristina Emilia Ursu[1], Margit Serban[1], Eugen Boia[2], Fulger Lazar[3], Jenel Marian Patrascu[4], Cristian Jinca[5], Estera Boeriu[5], Delia Savescu[6], Teodora Smaranda Arghirescu[5]

1. Romanian Academy of Medical Sciences, Onco-Hematology Research Unit, Children Emergency Hospital "Louis Turcanu" Timisoara, European Hemophilia Treatment Centre, 300011 Timisoara, Romania
2. Department of Pediatric Surgery, "Victor Babes" University of Medicine and Pharmacy Timisoara, 300041 Timisoara, Romania
3. Department of General Surgery II, "Victor Babes" University of Medicine and Pharmacy Timisoara, 300041 Timisoara, Romania
4. Department of Orthopedics II, "Victor Babes" University of Medicine and Pharmacy Timisoara, 300041 Timisoara, Romania
5. Department of Pediatrics, Division of Onco-Hematology, "Victor Babes" University of Medicine and Pharmacy Timisoara, 300041 Timisoara, Romania
6. Laboratory Department, Children Emergency Hospital "Louis Turcanu" Timisoara, European Hemophilia Treatment Centre, 300011 Timisoara, Romania

Introduction. Hemophilic pseudo-tumors (HPT) are rare, severe, late complications of hemophilia, observed in 1% - 2% of patients with severe disease. They are the result of recurrent hemorrhage in the musculoskeletal tissue, characterized by a slowly progressive encapsulation and calcification of an enlarging hematoma with subsequent erosion of the adjacent bones, always requiring a challenging difficult, mostly surgical, therapeutical approach.
Objectives. The aim of this study was to evaluate retrospectively our experience with HPT in the real-life of our patients with haemophilia (PwH).
Methods. In the period of 2001-2021, 198 invasive surgeries have been performed on 160 PwH, 6 of them (3,75%) with HPT; 4 of them were with proximal and 2 with distal HPT, all requiring a major intervention.
Results. The mean age at diagnosis and surgery was 27.33 ±6.42 years (19–36), five patients had severe form of the disease (3 HA, 1HB, and 1 with vWD type3), and one mild HB. The iliac bone was affected in 3 patients, 2 of them developing a giant retroperitoneal HPT, burdened by recurrent bleeding episodes and super-infection with fistulization to the intestine and the abdominal wall, with a slow healing process requiring a complex surgical intervention and over 90 days of hospitalization. A giant posterior right thigh HPT in another patient and a left forearm HPT in a young adult were noticed, both with good evolution after the tumor resection. In the last case, a neglected patient, the pseudo-tumor affected the right calcaneus, along the time completely destroying the talus, reaching the posterior edge of the tibiotalar joint and eroding the peroneal distal epiphysis, finally solved unfortunately only with limb amputation.
Conclusions. In the real-life of our PwH, HPT, high cost-demanding complications, are reflecting the scarce replacement therapy, only recently improved with the introduction of prophylactic regimens in our country.