Haemophilic pseudotumours treatment in patients with inhibitors

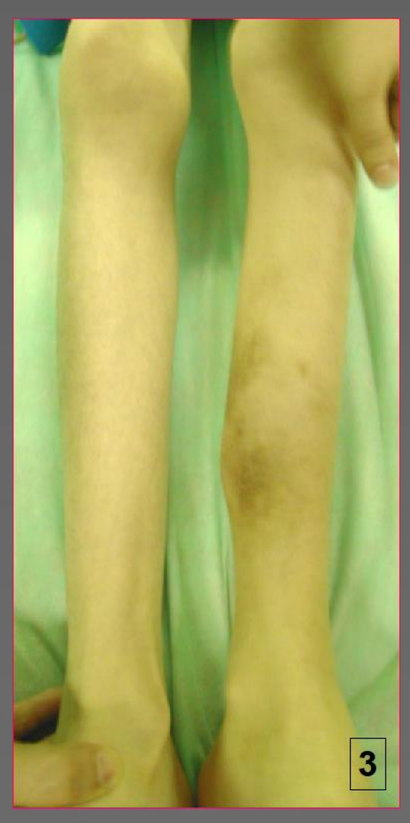
Caviglia HA[1,2] , Douglas Price AL[1], Candela M [2], Galatro G [1,2] , Moretti N.[2], Neme D.[2] ,Landro ME [1]

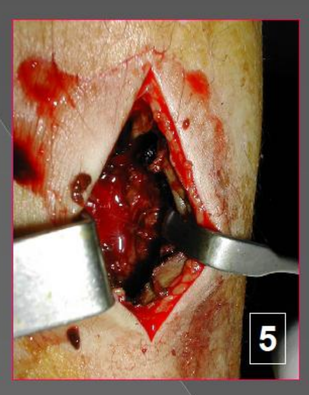
1- Hospital General de Agudos Juan A. Fernández, Buenos Aires, Argentina
2- Fundación de la Hemofilia, Buenos Aires, Argentina

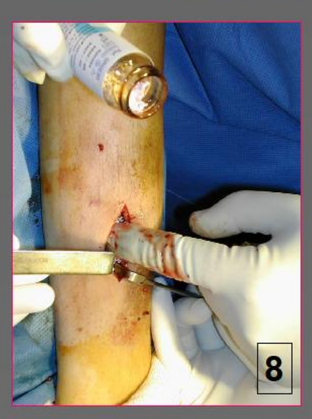
Introduction: The haemophilic pseudotumour (PS) is a serious complication in patients with haemophilia [1], that can lead to disability and even death. [2] The new mini-invasive treatments have reduced the number of amputations and disabilities. Development of inhibitors against factor VIII (FVIII) or IX (FIX) is the most serious complication of replacement therapy in patients with haemophilia.

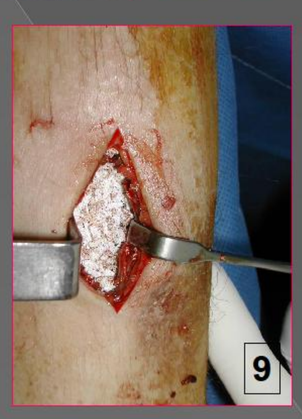
Objectives: The purpose of this study is to show our experience in the treatment of 9 pseudotumours in 7 patients with inhibitors treated by the same multidisciplinary team.

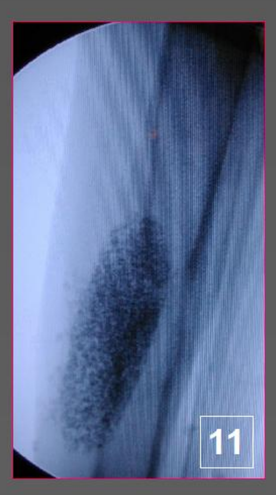
Methods: 7 patients with 9 PS, 7 had bone location: 2 in femur, 2 in tibia, 2 in calcaneus and 1 in cuboid, and 2 in soft tissues, in the arm and in the thigh. The average age was 21.4 years old (12-61 years old). Median time from onset of PS and presentation at our centre was 39 months (3-67 months). All patients were haemophilia A severe and had inhibitors to factor VIII, and underwent radiography and magnetic resonance imaging (MRI) at baseline to assess the size and content of the lesion (photos 1-2). In 2 patients, skin necrosis was observed in the lateral thigh, one had a bone PS (femur) and the other a soft tissue PS (thigh). The patients received Buenos Aires protocol as conservative treatment of their PS for 6 weeks. After 6 weeks treatment, the patients underwent to a new MRI of each PS to assess the response to treatment.
Patients that no respond to Buenos Aires protocol underwent to surgery (photo 3). The PS was identified with image intensifier (photo 4), then by aspiration was empting the cavity (photos 5-7), then the cavity was filled with hidroxiapatite (photos 8-9). Skin closure was performed and Rx made after surgery (photos 10-12).

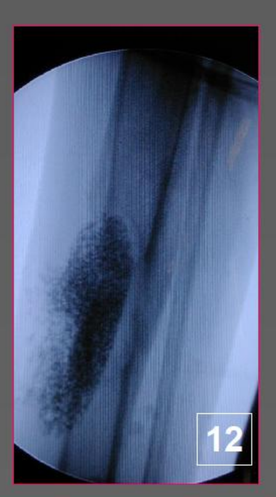












Results: Of the 7 patients with 9 PS only one responded to conservative treatment (11, 1%), it was a soft tissue PS in the arm with lowest time of evolution (3 months). Continued with Factor VIIa treatment for 6 weeks more with half dose of FVIIa and definitely cured of the PS. The others 6 patients where the PS was reduced less than 50% of its original size, surgery was performed , using the suction and refilling technique with granulated hydroxyapatite. (Table 1)All procedures were performed by the same multidisciplinary team. Four patients (57%) had some complication and 2 patients required a second surgery (28.5%). Skin necroses were observed in two PS (22.2%), in the thigh, one in the bone and other in the soft tissue. Time of evolution were 58 and 67 months respectively. Necrosis plate is produced by the excessive growth of the PS, it must be considered of poor prognosis [2, 3] . The patient with PS of the femur and skin necrosis died after necrotizing fasciitis.
Pre-surgery embolization proved to be very effective in reducing intra and postoperative bleeding.
In the postoperative period of 5 patients with 7 PS, intermittent mechanical compression with vaccum system were used reducing the possibility of bleeding into the evacuated cavities. [4]

Table 1

Patient	Age (years old)	Localization	Treatment	Complications	Vaccum System	Cured
1	12	Femur and calcaneus	Embolization +mini invasive	2nd toe Thrombosis	Yes	Yes
2	61	Tigh	Mini invasive	Wound dehiscence	Yes	Yes
3	13	Tibia	mini invasive + osteosynthesis	No	No	Yes
4	29	Femur	Pseudotumor and pseudocapsule resection	Necrotizing fasciitis, sepsis	No	Died
5	13	Tibia	Mini invasive	No	Yes	Yes
6	13	Calcaneus and cuboid	Only Embolization	PS Growth and Emergence of a new one	Yes	Yes

Conclusions: The only effective treatment to prevent PS is the treatment in time and manner of the bleedings in the musculoskeletal system. Given proper haemostatic coverage, and applying new and adequate surgical techniques, PS surgery with inhibitor in patients with haemophilia is possible, improving the quality of life of these patients.

Bibliography

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