Case series: patients with JAK2 positive myeloproliferative neoplasms associated with thrombotic events

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Objective. To identify patients with chronic myeloproliferative neoplasms with JAK2 mutation positive and the presence of thrombosis. To analyze the presence of some symptoms and laboratory parameters, their relation with diagnosis and the presence of thrombosis.

Methods. We reviewed the clinical records of these patients, to extract information for analysis of different variables such as age, gender, hemoglobin, platelets and leukocytes, general symptoms, type of myeloproliferative neoplasms and outcome of bone marrow.

Participants. Cancerology Unit, Hospital Pablo Tobón Uribe, Medellín (Colombia) and Emergency Room, Hospital Pablo Tobón Uribe, Medellín (Colombia).

Results. Of the seven patients 6 (86%) were women, showing a mean age of 57 years (SD+ / -10), hemoglobin 12 g/dl (SD+ / -2) with a count of platelets and leukocytes 662,714 (SD+ / -266,887) and 20,286 (SD+ / -11,521) respectively. Only 1 (14%) patient had splenomegaly, none referred sweats, weight loss or fever. Arterial thrombosis was documented in 3 (43%) patients, of which 100% was thrombosis of the middle cerebral artery; regarding venous thrombosis there were 5 cases documented (71%), of which 2 (40%) events thrombotic committed only the portal vein, 1 (20%) case committed the hepatic vein, 1 (20%) case of porto-mesenteric thrombosis, 1 (20%) patient had multiple events including thrombosis porto-mesenteric, pulmonary embolism, and venous thrombosis of upper limb. One of these patients presented arterial and venous thrombosis. The types of neoplasms were: polycythemia vera 2 (29%) patients, essential thrombocythemia in 3 (43%) patients and primary myelofibrosis in 2 (29%) patients.

Conclusion. Most of these events occurred in female patient, with high counts of leukocytes and platelets. The most frequent sites of thrombosis were the middle cerebral artery and portal vein.

We found in most bone marrow fibrosis, with increased cellularity and megakaryocytic hyperplasia.

Secondary prophylaxis with anti-inhibitor coagulant complex (AICC): from anecdote to reality

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Background. Evidence of prophylaxis benefit in patients without inhibitors to reduce hemarthrosis and severe bleeding have not been demonstrate in patients with hemophilia A (HA) and inhibitors, however, we could assume the reduction in bleeding episodes. Several publications have shown good results of secondary prophylaxis with AICC agents to reduce safely the number of bleeding events. We report the results in hemophilia treatment centers of two cities in a developing country, with access difficulties related to the high cost of this therapy to the health system.

Objective. To describe the frequency of hemarthrosis and other bleeding episodes in patients with HA and inhibitors under AICC (Feiba®) prophylaxis.

Methods. Case series study. Inclusion criteria: HA patients with high inhibitor title (>5 UB), severe bleeding or frequent hemarthrosis or target joint, at least 6 months of prophylaxis with AICC.

Results. Seven patients, median age 19 years (7-38), 6 patients with severe HA, one moderate. Inhibitor title at diagnosis between 4 and 104 U/B, peak title 1178 U/B, median 80 U/B (25.6 to 1178). Average time with AICC prophylaxis 2.8 years (7 months-6 years), range dose 40-75 U/kg 2 to 3 times/week. Before AICC prophylaxis: all type bleeds 5/year, target joint 6/7 patients, severe bleeding 2/7, hemarthrosis 7/7 patients. Associate factors with occurrence of inhibitors: not established 4/7, severe mutation 1/7, surgery 1/7, trauma 1/7. Bleedings after starting AICC: 1.5/year. No patient had thrombotic events during the time of prophylaxis with Feiba. Compliance with AICC schedule was over 80%.

Conclusions. Use of AICC prophylaxis showed a 70% reduction in all bleeding episodes. The cost can be considered a limitation for its use in a country with limited economic resources for health. Questions such as how long prophylaxis should be maintained, when to adjust therapy schedule while attempting eradication of inhibitors wait to be resolved?