Splanchnic vein thrombosis in patients with myeloproliferative neoplasm: little more than a blood disease in need for the evaluation of its treatment

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Valerio de Stefano and colleagues report their findings in a survey on splanchnic vein thrombosis (SVT) in 181 patients with myeloproliferative neoplasm. The major interest of this report is represented by the fact that the collaborating units were hematology units, whereas most previously reported material thus far has come from liver units. Contrasting the features according to the types of contributing units is most useful when one wants to make an idea of the disease as real as possible.

The first noticeable feature is the proportion of 42% of patients developing SVT after a diagnosis of MPN had been made, with the rest receiving a MPN diagnosis at the time of SVT or later on. This proportion is much higher than reported in surveys coming predominantly from liver units where only 26% of patients had a diagnosis of MPN prior to the development of SVT (1,2). Furthermore, mean duration from diagnosis to index thrombosis was 2.01 years (SD 3.9 years) in the recent hematology survey. The factors associated with the development of SVT following MPN diagnosis, including among others treatment and blood cell counts, will have to be determined if one is to address the issue of preventing the occurrence of these potentially severe vascular complications. Important independent confirmations include the largely predominant (97%) but unexplained role of the V617F JAK2 mutation; the absence of usual features of myeloproliferation or splenomegaly in many patients at the time of index SVT; the extremely frequent association with other thrombophilias (35%); and the rarity of local triggering factors (15%) (1, 2).

The design of the study could not ensure an accrual of consecutive patients. Therefore, the distribution of the sites of venous thrombosis and the factors that influence it can only be superficially considered. The predominance of portal vein involvement, followed by hepatic veins and mesenteric veins has thus far been a universal finding. It is interesting to note that the isolated involvement of the splenic vein was even more common than the isolated involvement of the superior mesenteric vein. This is an interesting piece of new information provided splenic vein thrombosis was not related to previous intervention on the spleen. Indeed, most cases of isolated splenic vein thrombosis thus far reported have been related to splenectomy, splenic artery embolization or pancreatic diseases.

Other findings of interest in this survey are due to the large number of patients collected, and their appreciable follow-up. It is particularly interesting to note that recurrence affected almost equally non-splanchnic territories (45%) and the splanchnic veins (55%). However, recurrence rarely implicated nonsplanchnic veins (5 patients, i.e. 16% of recurrences), and predominated in the arterial bed (10 patients, i.e. 33% of recurrences). Given the

large background difference in the incidence of thrombosis in nonsplanchnic territories and splanchnic vein in the general population, this similar rate of recurrence actually reflects the particular although unexplained propensity of thrombosis to affect splanchnic veins in patients with MPN.

These retrospectively collected data on SVT in MPN patients do not allow for as precise an evaluation as one would wish of the impact of antithrombotic therapy on the risk of recurrence or bleeding. Only 9 patients (5%) did not receive any antithrombotic treatment; only 6 (3%) received antiplatelet agents only; and only 10 patients (5.5%) received a combination of vitamin K antagonist and antiplatelet agents. Similarly, the benefits of cytoreductive therapy, an important issue for the management of such patients, could not be fully addressed as 72% of patients received such a therapy, which was combined with vitamin K antagonists in 82%. Still, there was no difference in the risk of recurrence (about 4 per 100 pt-years) according to cytoreductive therapy or combination of cytoreductive therapy and anticoagulation. These unadjusted data in non-randomized populations have to be interpreted with great caution.

Therefore one of the most interesting follow-up data in this large group of patients pertains to hematological transformation and cancer, occurring in 11 (6.1%) and 4 (2.2%) respectively, during the median 3.2 years of follow-up. Of note, no case of primary liver cancer was identified. These findings really position MPN as the predominant disease in patients with SVT. Another most interesting data in this regard is the similar rate of death among patients with BCS (1.4 per cent pt-year, 95% confidence interval 0.1–5.3) and with other SVT (2.0 per cent pt-year, 95% confidence interval 1.0–3.4). Thus far, the spontaneous outcome of BCS has always been found to be much worse that of other SVTs. The findings of the present study clearly show that current management with anticoagulation and interventional radiology, eventually complemented with liver transplantation has allowed for completely cancelling the excess mortality associated with BCS.

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