CNS Complications in Pulmonary Arteriovenous Malformations

To the Editor:

I read with great interest the review article by Cartin-Ceba et al1 in CHEST (September 2013). The authors give a concise, yet complete, overall view of pulmonary arteriovenous malformations (PAVMs), covering different aspects about the cause, clinical manifestations, diagnosis, and treatment.

One quite perplexing issue about PAVMs is how it seems like cerebral complications (ie, strokes, abscesses) are much more common in PAVMs related to hereditary hemorrhagic telangiectasia (HHT) than in those related to hepatopulmonary syndrome (HPS). A review of the literature provides just a handful of case reports of cerebral complications in PAVMs.2,3 On the other hand, there is an abundance of literature regarding cerebral complications of HHT. A recent case series from Denmark estimated a 7.8% prevalence of brain abscesses in patients with HHT and PAVMs.4 Knowing that the prevalence of HPS in patients with cirrhosis is estimated to be between 5% and 30%5,6 it is clear that there is a vast difference between these two entities and their CNS complications.

It could be argued that, mechanistically, PAVMs related to HPS are much smaller than those associated with HHT. But it is also true that patients with HPS usually have a much larger number of microscopic PAVMs (which explains why patients with HPS can have severe right-to-left shunt). In addition, patients with advanced cirrhosis often have altered mental status as a consequence of their own disease (hepatic encephalopathy), perhaps masking neurologic signs; therefore, some of the CNS complications of their HPS could be missed. But as a counterargument, many times patients with hepatic encephalopathy do have brain images taken, and one would expect that if brain abscesses were common in them, more case reports or series should be available. Local mechanisms for the clearance of microemboli and bacteria in the lung vasculature will require further study to elucidate the cause of this striking difference. At the current time, it is not clear if this is an issue related only to the size of the PAVMs or whether some other mechanisms are involved.

I truly enjoyed reading the comments of Cartin-Ceba et al1 about the difference in CNS complications between these two entities. There may be potential for research into the management of lung endotoxemia, phagocytosis, and clearance of microemboli that compare PAVMs related to HPS with those related to HHT.

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REFERENCES

Response

To the Editor:

We thank Dr Salerno for his interest in our article1 and for his insightful comments. We agree with him that the low reported incidence of brain abscesses in patients with hepatopulmonary syndrome (HPS) compared with patients with hereditary hemorrhagic telangiectasia (HHT) is intriguing. Although surveillance bias may play a role because this complication is well-known in patients with HHT, we consider that different mechanistic factors are at play.

First, although intuitively one might consider that size and frequency do matter (pulmonary arteriovenous malformations [PAVMs] are usually larger and more frequent in HHT than in HPS), the literature does not fully support this observation. Initial small studies suggested that neurologic complications in patients with HHT (brain abscess and stroke) are more common in those with more...