BACKSTREET BUBBLES

Aurélie Chapdelaine, PGY4
Université de Montréal
No conflict of interest
1. Investigate paradoxical embolism from an anatomical perspective

2. Describe the causes of extracardiac shunts, with an emphasis on cirrhotic patients

3. Illustrate how close intraprofessional collaboration led to understanding and addressing a complex anatomical issue
1. CASE PRESENTATION

Mr. G, ♂ 20 years old
Reason for consultation

*Multiple embolic strokes*
Past medical history

▷ Primary sclerosing cholangitis, neonatal diagnosis

▷ Cirrhosis
  - Portal hypertension (HTN)
    • Esophageal varices
    • Splenomegaly and hypersplenism
  - Liver failure
Initial presentation

▷ 2 episodes of tonic-clonic seizures
▷ T° 38.3
▷ No nuchal rigidity, normal neurological exam

Initial laboratory workup

▷ Pancytopenia
▷ Progressive liver failure
▷ Negative extensive infectious panel
▷ Normal TTE and TEE
Cirrhosis
Liver failure

Procoagulant state

Multiple embolism
Cirrhosis
Liver failure

Procoagulant state

Multiple embolism

Paradoxical embolism?
PATHWAY OF A PARADOXICAL EMBOLISM

1. Thrombotic Clot
   The formation of a blood clot inside a blood vessel (vein)

2. Intracardiac Shunt, Pulmonary Arteriovenous (AV) Malformation
   The venous blood clot crosses into arteries through a patent foramen ovale (PFO), septal defect, or pulmonary AV malformation, as a paradoxical embolism

3. Cerebral Embolism
   The embolus blocks blood flow to the brain circulatory system
   or
   Peripheral Embolism
   The embolus blocks blood flow to arms and legs

Windecker et al. J Am Coll Cardiol (2014)
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Bubbles in echocardiography

▷ Identification of R→L shunt
▷ N: contrast does not reach LV
▷ Intra♡ shunt: contrast reaches LV after 1-2 beats
▷ Extra♡ shunt: contrast reaches LV after 4-6 beats

Bubbles in echocardiography

- Identification of R → L shunt
- N: contrast does not reach LV
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Mr. G’s echocardiography

- Appearance of massive delayed bubbles in LV: arteriovenous malformation?
Differential diagnosis

▷ Hereditary hemorrhagic telangiectasia

▷ Hepatopulmonary syndrome (HPS)

+ No epistaxis
No telangiectasia
No family history
Differential diagnosis

- Hereditary hemorrhagic telangiectasia
- Hepatopulmonary syndrome (HPS)

- Portal HTN
- Cirrhosis

- No hypoxemia
- Capillary dilation > AVM
- Rarely associated with stroke
2. PORTO-PULMONARY VENOUS ANASTOMOSIS
Epidemiology

▷ Prevalence: ≈ 20%
▷ No association with the etiology of liver disease

Clinical consequences of PPVA

▷ LV volume overload leading to congestive heart failure
▷ Hypoxemia
  ○ Desaturation if shunt carries > 20% of CO
▷ Systemic embolism
  ○ Mainly described per procedure

Gonzalez et al. J Gastroenterol Hepatol (2012)
Imaging

- Contrast-enhanced CT scan
- Fluoroscopy combined with contrast TTE
  - Contrast injected in the left gastric vein

Coronary gastric vein

References:
- JP Chevrel. Anatomie clinique
If PPVA +:
- Contrast reaches the LV before the RV
- Contrast is more prominent in the LV

Coronary gastric vein

JP Chevrel. Anatomie clinique
3.

BACK TO THE CLINICAL CASE
Delayed bubbles remain unresolved

▷ PPVA is responsible for the cerebral embolism

▷ PPVA is a shunt from splanchnic system to left heart
  ○ It is not a right $\rightarrow$ left shunt

▷ Saline contrast was injected through the PICC line

▷ Hypothesis:
  ○ Backward flow of the contrast towards varices

A pathway via the azygos vein

Adapted from Netter, Atlas d’anatomie humaine (2009)
Transplant occurred 30 days after admission

The PPVA was successfully ligated
Key messages

PPVA is a rare cause of paradoxical embolism in patients with portal hypertension.

Associated risk is not well characterized, however particular vigilance is warranted during interventions.
References


Thank you