

LETTER / *Cardiovascular imaging*

Acute pulmonary embolism revealing Ivemark syndrome in an adult

F. Petitpierre^{a,*}, N. Alberti^a, L. Raffray^b,
M. Durieux^a, A. Veron^a, J.-T. Perez^a, A.-S. Lasserre^a,
N. Frulio^a, T. Carteret^a, P. Balageas^a, C. Salut^a,
F. Bonnet^b, H. Trillaud^a

^a Radiology Department, Saint-André Hospital, CHU de Bordeaux, Bordeaux Segalen University, 1, rue Jean-Burquet, 33000 Bordeaux, France

^b Internal Medicine Department, Saint-André Hospital, CHU de Bordeaux, 1, rue Jean-Burquet, 33000 Bordeaux, France

KEYWORDS

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Ivemark syndrome is a polymalformative syndrome, the cardinal element of which is asplenia, described for the first time by Björn Ivemark in 1955. This condition is rare in adults, as the majority of children affected do not survive beyond their first year. We report the case of a 21-year-old, hitherto asymptomatic, male patient, in whom this syndrome was diagnosed.

Observation

The patient was a 21-year-old male, who, suffering from sudden thoracic pain combined with dyspnoea, had an emergency chest CTA (with abdominal slices). A proximal bilateral pulmonary embolism was found (Fig. 1) combined with signs of acute right heart dysfunction. His laboratory results were normal (including haemostasis) and in particular did not indicate thrombocytosis.

We unexpectedly found a series of heterotaxic malformations:

- left pulmonary isomerism (absence of the middle lobe);
- absence of the retrohepatic inferior vena cava with azygos continuity (direct drainage of the hepatic veins into the right atrium, and the calibre of the azygos vein exceeding 6 mm) (Fig. 2);
- right polysplenia (multiple splenic islets in the right hypochondrium) (Fig. 3);
- adysmorphic pancreas with agenesis of its corporocaudal part (Fig. 4);
- preduodenal and prepancreatic portal trunk;

* Corresponding author.

E-mail address: petitpierre.francois@gmail.com (F. Petitpierre).



Figure 1. Chest CTA: bilateral proximal pulmonary embolism (arrow). Dilatation of the azygos vein (star).

- median liver (liver symmetrical relative to the midline);
- complete common mesentery;
- minor vascular abnormalities (the presence of four renal arteries, a retro-aortic left renal vein, multiple accessory splenic arteries arising directly from the aorta) (Fig. 3).

Discussion

This patient presented a polymalformative syndrome, the cardinal element of which was asplenia, corresponding to a syndrome described by Ivemark in 1955.

In the majority of cases, major cardiac malformation reveals this condition in the infant and is the reason for the high mortality rate of 60% before reaching 1 year of age [1]. Only 5 to 10% of patients with this syndrome reach adulthood [2].

We picked out polysplenia of accessory spleens, which are always associated in the normal situation with a main spleen.

The lack of fusion of foetal lobules leads to individualisation of multiple splenic nodules [3]. They remain in a posterior location along the greater curvature of the



Figure 3. Axial slice in the portal phase centred on the liver: polysplenia in the right hypochondrium (white star) with the splenic artery originating from the aorta (white arrow). Voluminous hemiazygos venous trunk (black arrow). Median liver (black star).

stomach in the right or left hypochondrium (depending whether there is abnormal gastric rotation or not).

The most frequently found hepatic anomaly is a median liver. In the present case, there was abnormal venous drainage with four hepatic veins with no retrohepatic inferior vena cava, combined with azygos continuation [4].

The complete common mesentery consists of rotation of the mesentery being stopped at 90°. The colon remains to the left; the small intestine, duodenum and stomach are placed to the right of the midline. The duodenum does not pass through the aorto-mesenteric ‘‘pincer’’ [5].

The pancreas has a larger cephalic end and its morphology is oval [6].

The pulmonary embolism probably resulted from deep venous thromboses which could not be demonstrated, given that agenesis of the inferior vena cava is a recognised factor favouring them [7] and that no other cause of thrombosis was found in laboratory tests: hypercoagulability factors, JAK2

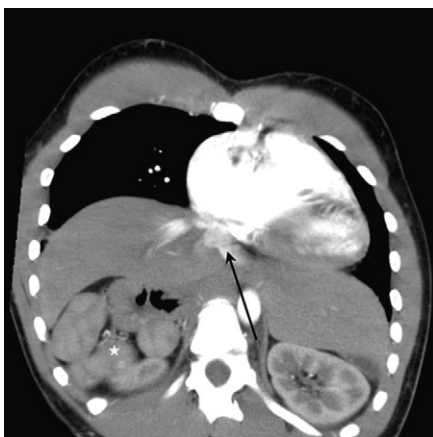


Figure 2. Oblique coronal slice centred on the hepatic veins: absence of the retrohepatic inferior vena cava (arrow). Polysplenia in the right hypochondrium (star).

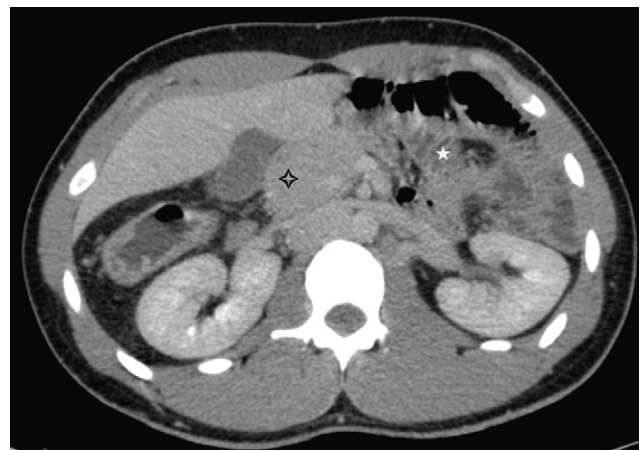


Figure 4. Axial slice in the portal phase centred on the pancreas: loop of small intestine in the left hypochondrium (white star), in relation with the complete common mesentery. Oval head of the pancreas without pancreatic body or tail (black star).

mutation, and antiphospholipid antibodies notably were all negative.

Cases of pulmonary embolism have previously helped reveal functional asplenia [8] or a venous vascular anomaly of agenesis of the retrohepatic inferior vena cava with azygos continuation [9].

In this second case, haemodynamic modification is responsible for the thrombotic phenomenon.

Conclusion

This Ivemark syndrome is an exceptional case because it has been revealed by a massive bilateral proximal pulmonary embolism. The malformations classically encountered (pulmonary, hepatic, splenic, intestinal and pancreatic) are associated with azygos continuity with agenesis of the retrohepatic inferior vena cava. By their modifying the haemodynamics, these anatomical variants are the cause of the thrombosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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