Abnormal atrial rhythm in a heterotaxy syndrome

Rythme atrial anormal dans un syndrome hétérotaxie

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A 30-year-old woman was referred with epigastralgia. Physical examination was normal, electrocardiography detected a low atrial rhythm of 45 beats per minute (Fig. 1, arrow), chest X-ray showed a slight cardiomegaly and echocardiography highlighted the existence of a dilated coronary sinus (13 × 17 mm). A thoracoabdominal computed tomography scan detected the presence of heterotaxy syndrome, consisting of:

Abbreviation: LAI, left atrial isomerism.
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Heterotaxy syndromes, or situs ambiguous, are rare (fewer than 1/10,000 births) and often include complex anatomical variations involving the heart, lungs, liver, spleen, and other organs. These syndromes affect the arrangement of organs in the thoracic and abdominal cavities, with differing degrees of complexity.

Figure 1. Electrocardiogram.

Figure 2. (A) Chest X-ray; (B–D) thoracoabdominal computed tomography scan images. A: left atrial isomerism; B: left bronchial isomerism; L: liver; S: spleen; ST: stomach; V: persistent left superior vena cava.

- left atrial isomerism (LAI; Fig. 2A) and left lung and bronchial isomerism (Fig. 2C);
- anomalous venous system (dilated coronary sinus draining to a persistent left superior vena cava (Fig. 2B), absence of intrahepatic segment of the inferior vena cava and abdominal drainage pouring to the hemiazygos system and from there to the persistent left superior vena cava);
- liver placed in middle position, occupying both hypochondria but with a predominantly left lateralization (Fig. 2B) and finally;
- stomach (Fig. 2A and D) and dysmorphic spleen (Fig. 2D) placed on the right side; the rest of the study was normal.
cardiovascular malformations. LAI is a form of heterotaxy in which the two atria and appendages are morphologically left; this explains why, in the absence of a sinus node and normal specific conduction tissue, these patients often have bradyarrhythmias (atrial ectopic pacemaker, abnormal P wave axis generally greater than 90°, rhythm of the atrioventricular junction and atrioventricular block, etc.); some require permanent pacemaker implantation. In the reported case, the P wave morphology (negative in leads II, III and aVF; positive in leads I and aVL; negative in leads V3–V6) indicated that the atrial rhythm anatomical site of origin was located at the inferior vena cava/right atrial junction or around the coronary sinus ostium.

LAI is usually associated with other cardiac malformations of different complexity, which are the main determinants of prognosis; only a minority of patients have mild disease, are asymptomatic into adult life and are diagnosed incidentally. These patients also often have a lack of asymmetry (i.e. lung and bronchial isomerism and anomalies) and impaired venous drainage of abdominal viscera: the most frequent features are polysplenia, liver predominantly symmetrical or left and intestinal malrotation.

Disclosure of interest

The author declares that she has no conflicts of interest concerning this article.