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Unusual association of Wolff-Parkinson-White syndrome and massive myocardial calcification in an infant with Ebstein's anomaly

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RESUMEN

El síndrome de Wolf-Parkinson-White (WPW) es una causa bien conocida de arritmias de reentrada debido a una vía atrioventricular accesoria (Arai y cols., 1990). Por otro lado, la calcificación miocárdica masiva es una reacción tisular rara y única; es el resultado de diferentes condiciones que inducen daño hipóxico-isquémico (Drut, y cols., 1998). Informamos el caso de un niño mexicano de seis meses de edad, con anomalía de Ebstein quien padecía síndrome de Wolf-Parkinson-White, arritmias cardiacas repetidas y calcificación miocárdica masiva. En la autopsia se encontraron trombosis en la aorta y las arterias renales con infartos renales secundarios. Proponemos que las arritmias fueron la causa subyacente de la calcificación miocárdica masiva.

Palabras clave: Síndrome de Wolff-Parkinson-White, arritmias, anomalía de Ebstein, vía atrioventricular, hipoxia-isquemia, infarto renal

ABSTRACT

Wolff-Parkinson-White syndrome (WPW) is a well known cause of reentry arrhythmias due to an accessory atrioventricular pathway (Arai *et al.*, 1990). On the other hand, massive myocardial calcification (MMC) is a rare and unique tissue reaction; it is the result of different conditions inducing hypoxic-ischemic damage (Drut *et al.*, 1998). We report a six months old Mexican male, with Ebstein's anomaly who had WPWS, repeated cardiac arrhythmias and MMC. Thrombosis in the aorta and the renal arteries were found at autopsy with secondary renal infarcts. We propose that the arrhythmias were the underlying cause of MMC.

Key words: Wolff-Parkinson-White syndrome, arrhythmias, Ebstein's anomaly, atrioventicular pathway, hipoxic-ischemic, renal infarcts

CASE REPORT

A six months old Mexican male patient was transfered to our hospital due to anuria, arrhythmias and sepsis. Two months before he presented with intractable diarrhea and vomiting. In a prior admission to another hospital he suffered severe dehydration, and loss of renal function. Several episodes of arrhythmia took place including ventricular fibrillation and cardiac arrest. An ECG showed the features of WPWS (figure 1). He was placed in peritoneal dialysis. An abdominal ultrasound showed several renal infarctions. The patient developed impetiginous ulcers and fascitis in the lower abdomen and genitalia positive for *Staphylococcus aureus* and *Pseudomonas aeruginosa*. The peritoneal cavity was secondarily infected and the patient died due to septic shock.

An autopsy was performed. The heart showed the features of Ebstein's anomaly; there was left ventricular hypertrophy (1 cm in thickness) and yellow-tan irregular areas of transmural myocardial discolorations under the endocardium and the papillary muscles (figures 2 and 3).

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On histology, numerous geographic areas of heavily calcified myocardial fibers were identified along with myocardial necrosis. There was no evidence of an inflammatory reaction (figures 4 and 5). There was an abdominal thrombus blocking and infarcting both kidneys. An hemorrhagic infarction was also found in the brain. The pancreas and lungs showed eosinophilic plugs of inspissated secretion in the ducts of exocrine pancreas and subbronchial glands.

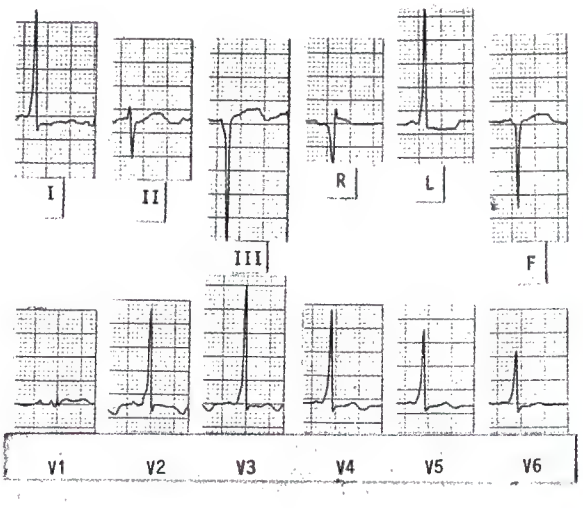

Figure 1. The patient's ECG tracing in sinus rhythm following a successful defibrillation. Notice the short P-R interval, <120 msec, the widened QRS complex, >110 msec and the delta wave.

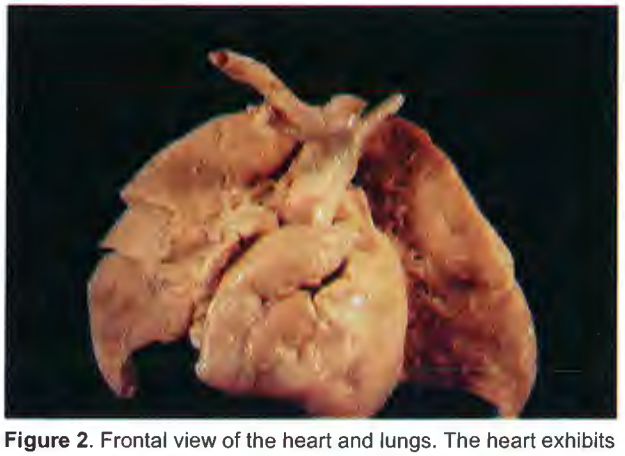

Figure 2. Frontal view of the heart and lungs. The heart exhibits dilatation of the right cavities.

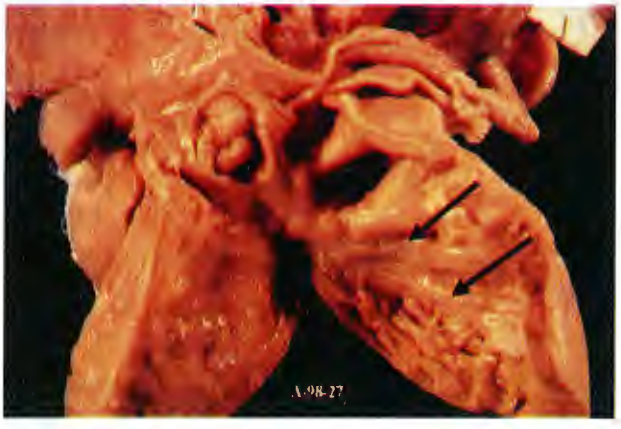

Figure 3. Right ventricle showing the abnormally low implantation of the tricuspid valve and an "atrialized" supravalvular portion of the right ventricle.

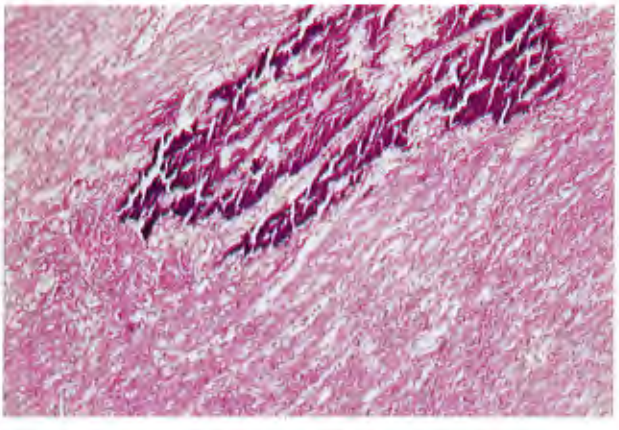

Figure 4. Histologic section of the left ventricular myocardium showing several areas of dystrophic calcification related to areas of infarction. HE 10 x.

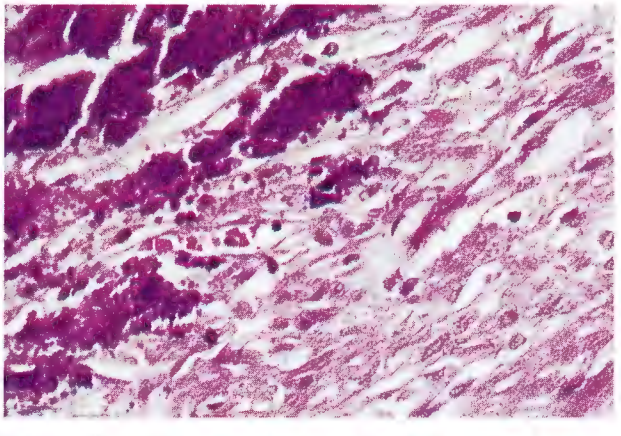

Figure 5. Enlarged portion of a peripheral calcified region showing necrotic myocardial areas. No inflammatory reaction is seen. HE 40 x.

DISCUSSION

Few articles have been published on MMC. Drut *et al.* (1998) recently wrote an extensive study on the subject. They presented seven cases in the perinatal period as a unique tissue reaction and pointed out that MMC is the end result of different conditions inducing hypoxic-ischemic damage (Topaz *et al.*, 1991). Two of their seven cases had developed cardiac arrhythmias detected *in utero*. In our case, the cause for arrhythmias was a preexcitation syndrome (WPW). In addition, the patient's condition was aggravated by the dehydration episodes and electrolyte imbalance caused by diarrhea.

Wolff-Parkinson-White syndrome (WPW) is a well known cause of reentry arrhythmias due to accessory atrioventricular pathways (Arai y cols., 1990; Smith y cols., 1982). This syndrome may be present in otherwise normal hearts, but it is often found in Ebstein's anomaly of the tricuspid valve as reported by Sodi-Pallares *et al.*, since 1955. The incidence of this association has been reported in 6 to 26% (Bialostozky *et al.*, 1972; Smith *et al.*, 1982; Iturralde, 1997). Therefore, the coexistence of these two conditions accounts for the episodes of arrhythmia of our patient. Most likely dehydration and electrolyte imbalance contributed to the occurrence of ventricular fibrillation in this child. Whereas the most common arrhythmia in patients with the WPW syndrome is supraventricular tachycardia, in cases of familial WPW syndrome, the appearance of more severe rhythm disorders such as paroxysmal atrial fibrillation and flutter are more common than in subjects with sporadic WPW syndrome (Gollob *et al.*, 2001). Furthermore, ventricular fibrillation does occur in some cases of WPW (Bialostozky y cols., 1972; Farre *et al.*, 1980) often following a bout of atrial fibrillation (Iturralde, 1997; Basson, 2001). In patients with Ebstein's anomaly and WPW, arrhythmias may occur in utero and as early as three months of age (Gaussí *et al.*, 1966).

The hereditary nature of WPW syndrome had been postulated since 1944 by Ohnell. There is recent evidence that this syndrome is an hereditary autosomal dominant disorder. The responsible gene has been mapped to 7q34-q36 (Gollob *et al.*, 2001). To our knowledge, no member of our patient's family (two parents and five siblings) had a congenital cardiopathy, WPW or episodes of dysrhythmias; one of the siblings was born with hearing loss and developed language disorders.

The associations of WPW with other congenital cardiopathies such as familial hypertrophic cardiomyopathy (McRae *et al.*, 1995; Gollob *et al.*, 2001), ventricular septal defect, tricuspid atresia, tetralogy of Fallot, etc. have been reported (Gaussí y cols., 1966).

It is a well recognized fact that WPW syndrome is due to the presence of accessory pathways, i.e., the bundles of Kent (Kent, 1893; Cárdenas, 1987, 1990), which allow the electrical impulse generated in the sinus node to establish a direct continuity between atriae and ventricles bypassing the normal path through the bundle of His. According to Gollob *et al.*, (2001) there is a mollecular defect responsible for inhibiting "the normal regression of muscle fibers" during cardiogenesis and the permanence of the future accessory pathways.

On the other hand, massive myocardial calcification (MMC) is a rare and unique tissue reaction seen in different conditions inducing hypoxic-ischemic damage [2] (Drut *et al.*, 1998).

We propose that the occurrence of repeated episodes of cardiac arrhythmias owing to the presence of a WPW syndrome in this child with Ebstein's anomaly was the cause of multiple myocardial calcification areas through hypoxic-ischemic damage to the heart.

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