Lesson of the Week

Epilepsy: a presentation of the Romano-Ward syndrome

F W BALLARDIE, R P MURPHY, J DAVIS

Primary cardiac arrhythmias presenting as epilepsy are a recognised phenomenon.¹ The outcome of such episodes is dependent on the underlying disorder. The potentially fatal and heritable cardiac disorder, the Romano-Ward syndrome, presented in a patient with recent onset epilepsy and a history of unexplained death in two siblings.

Case report

A 16 year old girl (case 1, fig 1) presented as an emergency after a nocturnal grand mal convulsion. Although she had had two similar fits in the preceding year, she had been otherwise well. This event aroused extreme anxiety in her parents, as her 20 year old sister (case 2, fig 1), who had had a two year history of infrequent fits, had died four days previously during an early morning convulsion. A necropsy had shown no airway obstruction or other specific findings. A second sister (case 3, fig 1), who was not a known epileptic, had also died in a nocturnal convulsion during a mild viral illness in 1976. Examination of our patient at presentation detected only a soft ejection systolic cardiac murmur with a normal pulse. Neurological examination and electroencephalogram showed no abnormality.

An underlying cardiac arrhythmia was considered as a possible cause of fits in view of the family history of at least one and possibly two deaths during convulsions. Electrocardiography showed a prolonged Q-T interval (fig 2a); an echocardiogram showed no evidence of hypertrophic cardiomyopathy and was normal.

Electrocardiograms were carried out on the rest of her family, and that of the sister who had recently died was reviewed. Three members of the family including the recently deceased sister (cases 1, 2, and the father) had prolonged Q-T intervals (fig 1). Her father, who did not have convulsions, had

A predisposing cardiac cause should be looked for in unexplained deaths after isolated fits

FIG 2—Electrocardiograms of (a) case 1 and (b) case 2.

a marginally prolonged corrected Q-T interval and suffered from occasional palpitations. None of the family was taking medication known to prolong the Q-T interval.

Discussion

In 1963 Romano² and in 1964 Ward³ reported families with histories of convulsions and sudden death, affected members showing prolonged Q-T intervals on electrocardiograms. Inheritance of the syndrome is autosomal dominant with variable expression. The syndrome has been reported in most Western societies. By 1975, 20 families with 85 affected members had been identified.⁴

The onset of convulsions occurs in most patients in childhood but is delayed to adolescence or adulthood in 25%.⁴ Convulsions result from cerebral anoxia during episodes of atypical ventricular tachycardia in which cardiac output is appreciably reduced. A prolonged Q-T interval is known to predispose to "torsade de pointes," which, like ventricular fibrillation, produces little or no cardiac output but, like ventricular tachycardia, may remit spontaneously⁵; continuing arrhythmia is likely to be fatal. A characteristic feature of the Romano-Ward syndrome is the precipitation of arrhythmias by bradycardias, such as occur nocturnally and during episodes of emotional stress.

Our patient's convulsions and the deaths of her two sisters are likely to have been a result of the Romano-Ward syndrome. Although noted, the importance of the prolonged Q-T interval on the electrocardiogram of her sister (case 2) was not appreciated until after her death (fig 2b). We believe that many

FIG 1—Family tree with Q-T intervals corrected for heart rate (QT_c).

North Staffordshire Royal Infirmary, Stoke on Trent ST4 7LN

F W BALLARDIE, PHD, MRCP, registrar in medicine

R P MURPHY, MB, MRCP, consultant neurologist

J DAVIS, MB, MRCP, consultant cardiologist

Correspondence to: Dr F W Ballardie.

doctors who are not specialist cardiologists are unaware of the existence of this syndrome. This is partly because of the paucity of published reports which draw attention to its presentation as epilepsy.⁶

The proper treatment for the Romano-Ward syndrome is unknown. The variety of methods of treatment suggests that most are only partially effective. Early reports of less frequent convulsions during treatment with beta-blockers have proved disappointing.⁷ Most other antiarrhythmic agents have been used, as has left stellate ganglionectomy.⁸⁻¹⁰ At present we have treated our patient with phenytoin 300 mg daily, which shortened the Q-T_c interval to 0.49 seconds (fig 3). As suggested¹¹ we intend to assess the threshold for arrhythmias by autonomic stimulation to determine if this is satisfactory treatment.^{7,12}

Unexpected death in a young epileptic, not resulting from status epilepticus or concomitant injury during a fit, is a rare event. It is unknown how many such deaths result from cardiac disorders. In a retrospective analysis of 19 deaths over 20 years in Cleveland, of the few patients who had electrocardiograms recorded, only one had prolongation of the Q-T interval.¹³ The allied syndrome of Jervell and Lange-Nielsen, where

congenital deafness is associated with lengthening of the Q-T interval, is clearly more easily recognised than is the Romano-Ward syndrome. The results of a survey of deaf children showed the prevalence of the former to be as high as 1%.¹⁴ We suspect that the Romano-Ward syndrome may not be as rare as it appears but that it is frequently unrecognised.

References

- Schott GD, McLeod AA, Jewitt DE. Cardiac arrhythmias that masquerade as epilepsy. *Br Med J* 1977;i:1454-7.
- Romano C, Gemme G, Ponsiglione R. *Clinica Paediatrica* 1963;45:656.
- Ward OC. A new familial cardiac syndrome in children. *J Irish Med Assoc* 1964;54:103-6.
- Schwartz PJ, Periti M, Malliani A. The long QT syndrome. *Am Heart J* 1975;89:378-90.
- Krikler DM, Curry PVL. Torsade de pointes, an atypical ventricular tachycardia. *Br Heart J* 1976;38:117-20.
- von Schildt E, Lehmann K. Zur problematik des Romano-Ward-syndroms. Fehlinterpretation als genuine epilepsie. *Z Gesamte Inn Med* 1978;33:776-8.
- Curtiss EL, Heibel RH, Shaver JA. Autonomic maneuvers in hereditary Q-T interval prolongation. *Am Heart J* 1978;95:420-7.
- Khan MM, Logan KR, McComb JM, Adgey AAJ. Management of recurrent ventricular tachyarrhythmias associated with Q-T prolongation. *J Cardiol* 1981;47:1301-7.
- Bashour T, Jokhadar M, Cheng TO. Effective management of the long Q-T syndrome with amiodarone. *Chest* 1981;79:704-6.
- Chaudron JM, Lebacqz EG. Romano-Ward syndrome treated by left stellatectomy and intracavity stimulation. *Am Heart J* 1980;100:131-3.
- Ratshin RA, Hunt D, Russell RO, Rackley CE. Q-T interval prolongation, paroxysmal ventricular arrhythmias and convulsive syncope. *Ann Intern Med* 1971;75:919-24.
- Rubin SA, Brundage B, Mayer W, Kanu C. Usefulness of Valsalva manoeuvre and cold pressor test for evaluation of arrhythmias in long QT syndrome. *Br Heart J* 1979;42:490-2.
- Hirsch CS, Martin DL. Unexpected death in young epileptics. *Neurology* 1971;21:682-90.
- Anonymous. Deafness, electrocardiographic abnormalities and sudden death [Editorial]. *JAMA* 1967;201:130-1.

(Accepted 14 April 1983)

FIG 3—Shortened Q-T interval of the patient in case 1 who was treated with phenytoin 300 mg daily.

Clinical curio: acute vitamin A toxicity from ingesting fish liver curry

Eleven patients (five men, six women) aged from 15 to 45 years were studied at the medical unit of the General Hospital, Colombo, Sri Lanka. The patients belonged to three different families and were taken ill after a meal that included the liver curry of the fish *Istiophorus gladius* (sailfish, Sinhala: Talapath). The quantity of fish eaten was about 113 g (4 oz) each. All patients complained of headache and drowsiness, the headache being severe and constant occurring within five to eight hours of the meal; one patient experienced nausea and five vomiting; blurring of vision and redness of eyes without diplopia was present in six cases; and a superficial peeling dermatitis was noted in nine of the patients in the face, neck, and back of chest; it appeared 48 to 72 hours after the meal and lasted from two to four days and regressed without specific treatment. Vitamin A concentrations were measured in four patients (in three of them temporal measurements were made) by a standard method using hexane extraction and using β carotene concentrations (figure).² All values obtained were higher than the normal for an age matched population (0.7-0.9 $\mu\text{mol/l}$ (20-27 $\mu\text{g}/100\text{ ml}$), mean 0.8 $\mu\text{mol/l}$ (24.7 $\mu\text{g}/100\text{ ml}$)). Highest values were noted in the first few days and all values were above the high normal range even on the seventh day. A falling off of the vitamin A values coincided with the loss of clinical manifestations and the disappearance of the dermatitis.

The patients had the typical manifestations of vitamin A toxicity² in its characteristic temporal sequence after ingesting fish liver curry. *Istiophorus gladius* is a large fish that can attain a length of 8 to 10 feet: it thus had a large liver. The liver contains variable amounts of fish oil—for example, it contains 5-7% of oil, and this oil may contain 5000 to 25 000 IU vitamin A/g. Its oil content appears to be higher than that of other large fish in Sri Lankan waters like tuna, skipjack, and shark.³ Interestingly, the tunny fish liver (Britain) also has a very high vitamin A content of about 50 000 IU/g oil. The clinical findings in

Serum vitamin A concentrations in four patients with toxicity. Conversion: SI to conventional units—1 $\mu\text{mol/l} \approx 29\text{ }\mu\text{g}/100\text{ ml}$.

our patients are comparable to those found after ingesting the liver of polar bear or seal, which also contain large amounts of vitamin A.⁴—S RAMACHANDRAN, physician, P PREMACHANDRA, biochemist, and D K D SILVA, technician.

- Cooper GR, ed. *Standard methods of clinical chemistry*. Vol 7. London: Academic Press, 1972:215-30.
- Stimson WH. Vitamin A intoxication in adults. Report of a case with a summary of the literature. *N Engl J Med* 1961;265:369-73.
- Mendis AS. *Fisheries research. Bulletin No 2*. Ceylon: Department of Fisheries, 1954.
- Rodahl K, Moore T. Vitamin A content and toxicity of bear and seal liver. *Biochem J* 1943;37:166-8.