Fulminating hyperlipidaemia

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Summary
A case is described of severe recurrent ischaemic heart disease associated with rapidly increasing and ultimately massive hyperlipidaemia.

The case is discussed, with suggestions for management of similar problems.

Introduction
Massive hypertriglyceridaemia has been recorded in association with acute alcohol abuse (Zieve, 1958), with oestrogen therapy (Molitch, Oill and Odell, 1974), with impaired fatty acid incorporation into adipose tissue and severe chylomicronaemia (Carlson, Eriksson and Walldius, 1974) and in glycogen storage disease and diabetic ketoacidosis (Carlson, Fröberg and Öröl, 1972). There may be associated jaundice, haemolysis and abdominal pain with or without pancreatitis. Disorders of circulation, including increased platelet adhesiveness, red cell aggregation and plasma viscosity with reduced fibrinolysis are probably a constant feature.

Although the condition usually responds well to treatment by management of any precipitating cause, and appropriate diet and drug therapy, the case reported was resistant to these manoeuvres and manifested progressive and ultimately fatal hyperlipidaemia.

Case report
The patient was a female born in 1906. She had an apparently completely successful mastectomy for carcinoma in 1955. Otherwise, she enjoyed good health until 1970 when she was seen in the medical outpatient department with angina pectoris. There was no family history of premature ischaemic heart disease. She was teetotal, but smoked 40 cigarettes/day. Her blood pressure was 170/90 mmHg, and her weight was 65 kg (ideal for height 56 kg), but there were no other physical signs. She was persuaded to stop smoking and prescribed glyceryl trinitrate with apparent benefit. Her serum cholesterol was noted to be 9·6 mmol/l. Her serum triglycerides were not measured although the serum was not noted to be turbid.

In October 1974 she was admitted as an emergency with severe chest pain, and serial ECGs demonstrated an acute inferior myocardial infarction, which was complicated by the development of left ventricular failure. Her weight was now 67 kg. Fasting serum lipids on the day following admission showed a cholesterol of 10·2 mmol/l and triglycerides of 8·8 mmol/l. Cellulose acetate electrophoresis showed a very marked increase in the pre-β-lipoprotein, a moderate increase in β-lipoprotein and a trace only of chylomicrons. Preparative ultracentrifugation followed by lipoprotein electrophoresis excluded broad-β disease, and the patient was considered to have endogenous hypertriglyceridaemia (Fredrickson Type IV). She was commenced on a 4·2 MJ low carbohydrate reducing diet. Transient glycosuria was noted during this admission.

In February 1975 she was readmitted with further severe chest pain and ECG changes of an inferolateral myocardial infarction, complicated by pulmonary embolism 10 days later. She was treated with heparin, and then warfarin subsequently. Her serum lipids remained abnormal with a cholesterol of 10·7 mmol/l and triglycerides of 7·2 mmol/l. Transient glycosuria again occurred. Since her weight had not fallen despite claimed adherence to diet, her energy intake was further restricted to 3·4 MJ daily.

A glucose tolerance test was performed in March 1975 which indicated diabetes mellitus, with a blood glucose of 14 mmol/l and glycosuria at 2 hr.

In May 1975 she was admitted with myocardial ischaemia, without further ECG changes diagnostic of infarction, and her serum lipids showed a cholesterol of 15·4 mmol/l and triglycerides 30 mmol/l. In July 1975 she was readmitted with further myocardial ischaemia and her serum lipids were again disturbed, with a cholesterol of 27 mmol/l and triglycerides >20 mmol/l, although her weight had
There was a small haemorrhagic pericardial effusion and massive bilateral pulmonary emboli. The liver weighed 2.17 kg with centrilobular venous congestion and uniform fatty degeneration. There was no evidence of recurrent or metastatic breast carcinoma.

**Discussion**

It is known that any acute illness may cause temporary alterations in serum lipid levels, and this makes interpretation of values difficult, since the patient reported had 6 emergency hospital admissions in 14 months. Ideally serum lipids should be sampled 3 months or more after emergency admission, and this was clearly impractical in this patient. Except where stated the values recorded were those obtained immediately after admission, when they are least likely to be affected in this way.

In a local population screen, 4% of 99 healthy women and 14% of 282 healthy men had serum triglycerides \( \geq 2.5 \) mmol/l. The present patient had the most severe hypertriglyceridaemia seen in 5 years at a lipid clinic, and fared badly on treatment. In 4 other cases with severe hypertriglyceridaemia (\( \geq 10 \) mmol/l) there has been a good response to dietary advice, with the addition of clofibrate 2 g/day in 3 cases. Their initial levels were 15, 10, 24 and 13 mmol/l, which fell to 1.8, 1.1, 2.9 and 2.9 mmol/l respectively after therapy. Severe hyperlipidaemia detected can sometimes be associated with alcoholism, glycogen storage disease, diabetic ketoacidosis and oestrogen therapy, but none of these was present in this severely affected patient. Thiazide diuretic therapy can be blamed for mild hyperlipidaemia (Ames and Hill, 1976), but not in this patient, who had been treated with frusemide.

The results of glucose-tolerance testing indicated that the present patient had mild maturity onset diabetes but this was at no time out of control. No other primary factors were identified nor any explanation of the severity of the progressive and ultimately massive hyperlipidaemia which apparently precipitated the patient's terminal illness. The levels of serum triglyceride detected would have had marked effects on platelet adhesion and aggregation (Nordøy, Strøm, and Gjesdahl, 1974) as well as increasing plasma viscosity, and these secondary phenomena could explain the accelerated ischaemic heart disease.

Conventional management with low-energy low-carbohydrate diet, clofibrate and nicotinic acid did not cure the hypertriglyceridaemia, though there was evidence of compliance with each of these measures. Normally the response to treatment for hyperlipidaemia should be assessed at leisurely intervals, to permit maximal benefit to be obtained from each therapy before further alteration. This
was clearly impossible with this patient, where a prompt response was particularly desirable. Other procedures which might have helped by reducing platelet adhesion and aggregation include unsaturated fat diet (Nordøy and Rodset, 1971) and drugs such as aspirin, dipyridamole, phenformin and ethyloestrenol. The course of warfarin did not prevent 2 further episodes of myocardial ischaemia, however.

The plasma viscosity might have been lowered by repeated plasmapheresis. This probably could only have been used to produce a temporary respite while other manoeuvres were employed, such as higher dose nicotinic acid therapy and possibly the recently proposed chenodeoxycholic acid (Bell et al., 1973; Bateson, 1976).

The sequence of events in this patient illustrates the need for vigorous treatment of severe hyperlipidaemias, despite current scepticism about the management of lesser degrees of these disorders (Coronary Drug Project, 1975; Werkö, 1976).

References


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