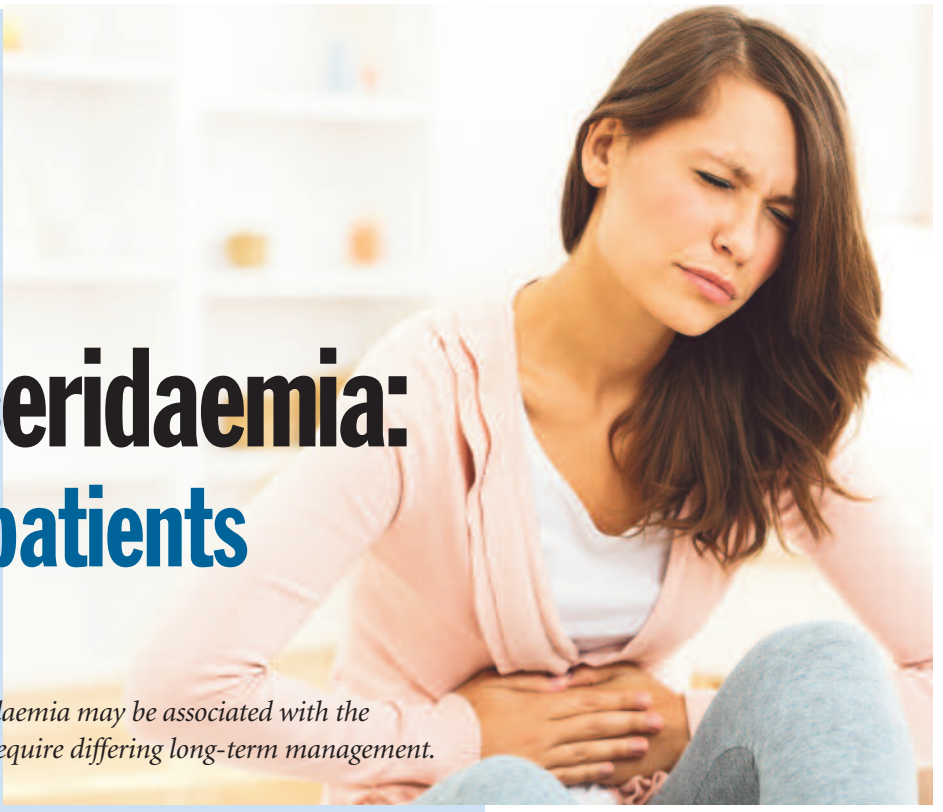
CASE STUDY

Extreme hypertriglyceridaemia: a tale of two patients



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The phenotype of extreme hypertriglyceridaemia may be associated with the onset of acute pancreatitis, patients may require differing long-term management.

Case scenario 1

Ms BM is a 28-year-old, previously well, woman who presented to a district hospital with a 14-hour history of acute upper abdominal pain radiating through to her back. This was associated with fever and vomiting.

Based on her history, physical examination and abdominal signs, a provisional diagnosis of acute pancreatitis was made and this was rapidly confirmed by highly elevated serum amylase and lipase readings. Other key blood test results were as follows:

- *sodium 129 mmol/L (desirable 135 to 145 mmol/L)*
- *potassium 3.3 mmol/L (desirable 3.4 to 5.4 mmol/L)*
- *creatinine 60 µmol/L (desirable <110 µmol/L)*
- *glucose 4.1 mmol/L (desirable <5.5 mmol/L)*
- *alanine aminotransferase 49 U/L (desirable <45 U/L)*
- *aspartate aminotransferase 48 U/L (desirable <40 U/L)*
- *gamma glutamyl transferase 130 U/L (desirable <55 U/L)*
- *total cholesterol 16.6 mmol/L (desirable <5.0 mmol/L)*
- *triglycerides 78 mmol/L (desirable <2.0 mmol/L)*
- *HDL-cholesterol 0.7 mmol/L (desirable >1.0 mmol/L).*

Her plasma appeared very creamy with a pink tinge, which was indicative of haemolysis.

Ms BM was managed with nil-by-mouth plus an intravenous antibiotic, electrolytes, glucose and pain relief. Within three days, she had made a good clinical recovery, with serum amylase levels returning to the reference range, and a low-fat diet was gradually introduced from day four. A CT abdominal scan on day three showed an oedematous pancreas and evidence of fatty liver, but no evidence of gallstones or fluid accumulation in the lesser peritoneal sac.

Her pancreatitis seemed to be attributable to extreme hypertriglyceridaemia. Her lipid levels were monitored over the nine days of hospitalisation (Figure).

Consultant comments

This is a well-documented scenario in patients with extreme hypertriglyceridaemia, especially when triglyceride levels persistently exceed 10 mmol/L. Other causes of pancreatitis, such as gallstones or alcoholism, were excluded in this case. The low serum sodium level is an example of 'pseudohyponatraemia' due to the volume effect of extreme lipaemia, which will have resolved as lipid levels were reduced. Fatty liver is likely to be due to extreme hypertriglyceridaemia because other likely causes are absent and it will slowly resolve as lipids levels fall. Beyond the immediate acute management, long-term treatment of Ms BM will revolve around management of the extreme hypertriglyceridaemia.

Key points

- **Extreme hypertriglyceridaemia may be associated with and even cause acute pancreatitis.**
- **Extreme hypertriglyceridaemia generally has a genetic basis but with possible exacerbation by factors such as diabetes, alcohol consumption, obesity, nephrosis, hypothyroidism, exogenous oestrogen or other drugs.**
- **Although there may be other causes of acute pancreatitis present, one goal of treatment is to achieve and maintain triglyceride levels well below 10 mmol/L.**
- **Long-term management of extreme hypertriglyceridaemia generally requires lipid-lowering drugs and/or attention to exacerbating factors.**

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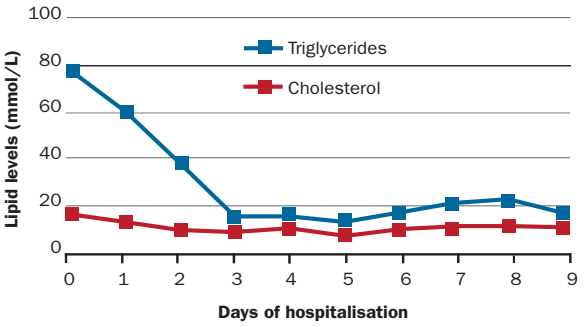

Figure. Graph showing Ms BM's lipid levels over her nine day hospital stay.

Extreme hypertriglyceridaemia generally has a genetic basis but is possibly exacerbated by factors such as diabetes, alcohol consumption, obesity, nephrotic syndrome, hypothyroidism, exogenous oestrogen exposure or use of some other drugs.

With Ms BM's triglyceride levels now averaging about 20 mmol/L in the 'recovered' state, this may be representative of her basal level on a low-fat diet. Why she drifted to much higher levels is unclear, but this may be related to a higher fat intake. The metabolic problem here is usually a clearance defect in triglyceride-rich lipoproteins (reduced activity of lipoprotein lipase). In the absence of other specific exacerbating factors, her extreme hypertriglyceridaemia will need ongoing drug therapy with a fibrate or perhaps high-dose omega-3 fatty acids or high-dose niacin.

Case scenario 1 continued

Ms BM's doctor was concerned about her residual cholesterol problem and discharged her on a high-dose statin, namely atorvastatin 40 mg/day, plus a low-fat diet, requesting follow-up review within six weeks.

Six weeks after discharge Ms BM remained clinically well and her lipids seemed 'improved', with the following results:

- *total cholesterol 8.1 mmol/L*
- *triglycerides 10 mmol/L.*

Her dose of atorvastatin was increased to 80 mg/day and she was reviewed again six weeks later. Lipid levels had now 'deteriorated', with the following findings:

- *total cholesterol 9.6 mmol/L*
- *triglycerides 15 mmol/L.*

She was then referred for my consultant opinion.

Consultant comments

Considerable variability in this lipid disorder can complicate interpretation of any response to treatment. Ms BM has not improved with this treatment. As suggested above, a fibrate is an important option in this setting, although many such patients often require dual statin/fibrate therapy (in addition to a low-fat diet).

Case scenario 1 concluded

Ms BM was started on fenofibrate 145 mg, one tablet daily in combination with a reduced dose of atorvastatin of 10 mg/day and she was reviewed eight weeks later. Her lipid levels seemed improved, with the following findings:

- *total cholesterol 6.2 mmol/L*
- *triglycerides 5.9 mmol/L*
- *HDL-cholesterol 0.9 mmol/L.*

Atorvastatin was increased to 40 mg/day and she was reviewed several times over the next six months. Her best lipid results were:

- *total cholesterol 5.9 mmol/L*
- *triglycerides 4.8 mmol/L*
- *HDL-cholesterol 1.0 mmol/L.*

However, there was only minor variations in these results from visit to visit. Her compliance with drug intake was 100% and the ongoing therapy was then supplemented with fish oil capsules 1000 mg, three capsules twice a day (delivering 1.8 g/day of omega-3 fatty acid). Subsequent follow up showed no further genuine improvement in her lipid profile.

Given the heavy nature of her lipid therapy and the good degree of improvement in her lipid profile, it was time to declare a compromise in her lipid management and she was continued permanently on atorvastatin 40 mg/day and fenofibrate 145 mg/day but without fish oil. She remained clinically well over the next 18 months and to the present time and with essentially unchanged lipid results.

Consultant comments

Most patients with extreme hypertriglyceridaemia can avoid acute pancreatitis if their triglyceride levels are kept below 10 mmol/L, although there are exceptions to this generalisation.

Case scenario 2

Mr HC is a homeless 58-year-old man and a self-confessed alcoholic. He was admitted to my university hospital with acute pancreatitis. His test results were as follows:

- *triglycerides 38 mmol/L*
- *total cholesterol 10.1 mmol/L.*

He was given standard management and required five days of nil-by-mouth and intravenous fluids before he was well enough to resume food intake. (He was also given treatment to manage his alcohol withdrawal.)

After five days of nil-by-mouth and zero alcohol intake, Mr HC's results were as follows:

- *triglycerides 3.9 mmol/L*
- *total cholesterol 4.6 mmol/L*
- *HDL-cholesterol 1.5 mmol/L*
- *LDL-cholesterol 1.3 mmol/L.*

Other investigations confirmed the presence of early cirrhosis. He was eventually discharged to attend follow up at a clinic for alcohol and drug addiction. Not surprisingly, he failed to attend.

Consultant comments

Mr HC is an example of a case of extreme hypertriglyceridaemia largely due to alcohol induction. Based on extensive past experience with this type of situation, lipid drugs are not generally very effective if heavy alcohol intake continues. The prognosis for Mr HC is poor and he is likely to return to hospital or die from complications of alcoholism.

Conclusion

We see here two patients with acute pancreatitis related fully or in part to extreme hypertriglyceridaemia. They have differing aetiologies for a similar lipid phenotype. Long-term management of each patient is different, as are their respective prognoses. **CT**

COMPETING INTERESTS: None. The views expressed are purely those of the author.

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