Hiding in Plain Sight?  
A Case Report of Elevated Lipoprotein X

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Background

Lipoprotein X (LpX) is a pathological lipoprotein associated with cholestatic liver disease. Elevations in LpX can masquerade as elevated LDL, which may lead to a misdiagnosis of familial hypercholesterolaemia (FH).

However, unlike FH, LpX is not thought to confer increased cardiovascular risk.† We discuss a case where elevated LpX manifested as severe hypercholesterolaemia and highlight our diagnostic approach.

Case Report

A 54-year-old female, with a five-month history of nausea and progressive weight loss, was urgently admitted with suspected gastric outlet obstruction.

Liver function tests on admission showed total bilirubin 21 (RI 0-20 µmol/l), alanine transaminase 879 (RI 9-55 U/L), alkaline phosphatase 460 (RI 30-130 U/L) and gamma-glutamyl transferase 944 (RI 6-35 U/L).

Oesophagogastroduodenoscopy visualised abnormal pyloric thickening and histology confirmed gastric adenocarcinoma. Computed tomography revealed a thickened gallbladder neck secondary to infiltrative metastatic spread, providing a cause for the biochemical evidence of biliary stasis.

Lipid Investigations

A baseline lipid profile demonstrated total cholesterol 8.4 mmol/l and LDL 5.8 mmol/l, which continued to rise to 17.3 mmol/l and 15.7 mmol/l respectively.

Remnant particles were excluded by beta-quantification [VLDL 2.45 mmol/l]. Lipoprotein electrophoresis was requested on a subsequent blood specimen (Fig. 1). On the basis of these results, LpX, secondary to neoplastic cholestasis, was felt to explain the apparent increase in LDL.

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Discussion

The elevation in total cholesterol/apoB ratio was strongly suggestive of LpX, and it was thus vexing that LpX was not demonstrated on electrophoresis. One potential explanation is that the blood specimen sent for electrophoresis was drawn after the patient underwent palliative biliary stenting. This would have predictably resulted in decreased of LpX levels, as the presence of LpX is correlated with extent of biliary obstruction.

Furthermore, even if present, LpX is not always revealed on lipoprotein electrophoresis.† The electrophoresis report suggested that direct LDL measurement should be considered; however, as the patient was receiving end-of-life care, no further investigations were felt appropriate.

Learning Points

1. The presence of LpX should be considered where severe elevations in total cholesterol are noted.

2. Total cholesterol/apoB ratio has utility in the initial investigation of severe hypercholesterolaemia and can provide a means of identifying high levels of LpX, in the absence of a specific assay.