ACP Oral Presentation:
Congenital Lipodystrophy: Can Fat Harm the Lungs?

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Background Info

- Lipodystrophy refers to abnormal lipid distribution in the body
- Congenital versus acquired
- Clinical diagnosis based on:
  - PE/Body dysmorphia
  - Lipid profiles
  - Associated Comorbidities
  - Genetic testing

Here we present a case of a patient with suspected congenital lipodystrophy who was admitted for symptomatic hypertriglyceridemia without pancreatitis.

She subsequently developed ARDS after treatment with insulin drip for rapid reduction of serum triglyceride levels.
Case Presentation

• 34 y/o Hispanic female w/ a hx of congenital lipodystrophy of unknown type
• Major complications- uncontrolled DM with severe insulin resistance, DM nephropathy, neuropathy, retinopathy, gastroparesis, hepatosteatosis significant vascular atherosclerosis, hypertriglyceridemia and nephrotic syndrome

• In the ED, severe epigastric pain radiation to the back.
• Triglyceride level of 3,184 mg/dL (her baseline of 300mg/dL). Glucose of 432 mg/dL but urine with negative ketones. Lipase of 16u/L and amylase of 29u/L. LFTs were also WNL. A RUQ ultrasound and CT abdomen showed no acute pancreatic abnormalities. Started on insulin drip admitted to medical wards.
Second day lipid profile study interpretation as evidence of Type V hyperlipoproteinemia.

Triglyceride came down to 1500mg/dL.

On the third day, tachycardia, leukocytosis, pleuritic chest pain. Intubated for ARDS.

MICU course was uncomplicated received daily IV Lasix with adequate diuresis

Triglycerides continue to come down and was at 800 before transitioning to subcu insulin.

Extubated on the 5th day and transferred to the medical floors.
<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lipoprotein, Cholesterol</td>
<td>641 * H</td>
</tr>
<tr>
<td>Lipoprotein, Triglyceride</td>
<td>3,379 * H</td>
</tr>
<tr>
<td>Lipoprotein, HDL</td>
<td>10 * L</td>
</tr>
<tr>
<td>Lipoprotein, LDL</td>
<td>68 *</td>
</tr>
<tr>
<td>APO-Lipoprotein B</td>
<td>183 * H</td>
</tr>
<tr>
<td>LDL Triglycerides</td>
<td>278 * H</td>
</tr>
<tr>
<td>VLDL Cholesterol</td>
<td>504 * H</td>
</tr>
<tr>
<td>VLDL Triglycerides</td>
<td>1,883 * H</td>
</tr>
<tr>
<td>Beta-VLDL Cholesterol</td>
<td>Not Detected *</td>
</tr>
<tr>
<td>Beta-VLDL Triglycerides</td>
<td>Not Detected *</td>
</tr>
<tr>
<td>Chylomicron Cholesterol</td>
<td>59 * H</td>
</tr>
<tr>
<td>Chylomicron Triglycerides</td>
<td>1,178 * H</td>
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<tr>
<td>Lp(a) Cholesterol</td>
<td>&lt;3 *</td>
</tr>
<tr>
<td>LpX</td>
<td>Not detected *</td>
</tr>
<tr>
<td>Lipoprotein Interpretation</td>
<td>Type V Hyperlip</td>
</tr>
</tbody>
</table>
Discussion

- In this case it was evident that the patient did not develop pancreatitis based on lipase levels and abdominal imaging.
- This is despite extremely elevated triglycerides in the blood.
- Treatment of hypertriglyceridemia with insulin drip is standard to reduce the amount of triglycerides in the blood in hypertriglyceridemic pancreatitis. Has also been reported to be safe.
- In research on pancreatitis causing ARDS, FFA found to cause oxidative damage in addition to damage from pancreatic enzymes.
• We believe she likely developed ARDS from FFA mediated pulmonary endothelial damage and surfactant disruption with rapid treatment with insulin drip
• Presented with ARDS on second day of insulin infusion with decreasing triglycerides from 3,000 to 1,500 accompanied by high free fatty acid.
• The process likely predisposed by her comorbidities
• Future consideration in such patient may indicate a slower treatment of hypertriglyceridemia with insulin or to directly treat with plasmapheresis
Questions?

Figure 2 Milky ultrafiltrate obtained after a single cycle of plasmapheresis with drop in serum triglyceride levels from > 4000 mg/dL to < 500 mg/dL.

Gupta A et al. J Gen Intern Med 2015; 31:252
Thank You!
References