Thyroid storm & bulbar thyrotoxic myopathy at presentation of Graves Disease in a 22-month-old

Summary of Clinical Case: Presentation

- 22-month female, Thai ethnicity
- Family history of autoimmune thyroid disease & diabetes

Presenting features (history):
- Tachycardia, hypertension
- 3 days cough & fever, 2 weeks rhinorrhoea & throat clearing
- 6 months diaphoresis & growth spurt

Clinical assessment:
- Diaphoretic & flushed
- HR 200 bpm (NR<140 bpm), BP 145/90 mmHg (95th% 108/66)
- Raised JVP, bounding pulse
- RR 36 bpm (<30), work of breathing, right sided crepitations
- Length 98th% (MPH 10-25th%)

Initial Investigations

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
<th>Normal Range</th>
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</thead>
<tbody>
<tr>
<td>TSH</td>
<td>&lt;0.005 pmol/L</td>
<td>0.70-5.97</td>
</tr>
<tr>
<td>fT4</td>
<td>&gt;100 pmol/L</td>
<td>12.3-22.8</td>
</tr>
<tr>
<td>fT3</td>
<td>45.5 pmol/L</td>
<td>3.69-8.46</td>
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<tr>
<td>TSH receptor Ab</td>
<td>&gt;20 IU/L</td>
<td>&lt;1.8</td>
</tr>
<tr>
<td>TPO Ab</td>
<td>343 IU/mL</td>
<td>0-34</td>
</tr>
<tr>
<td>Thyroid US</td>
<td>Diffuse enlargement, heterogeneous. No nodules.</td>
<td></td>
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<tr>
<td>Chest X-ray</td>
<td>Mild cardiomegaly</td>
<td></td>
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<tr>
<td>ECG</td>
<td>Left ventricular hypertrophy, sinus tachycardia</td>
<td></td>
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<tr>
<td>Echocardiogram</td>
<td>Mild left ventricular hypertrophy</td>
<td></td>
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Thyrotoxic Myopathy & Dysphagia

- Severe proximal myopathy (+ve Gowers sign) with myopathic dysphagia & aspiration confirmed on modified barium swallow (right). Myasthenia gravis investigations negative (pyridostigmine trial, MuSK/AchR Ab). Managed with NG feeds, succioning secretions, & thyrotoxicosis treatment.
- Clinical impression: Graves thyrotoxic proximal & bulbar myopathy with secondary aspiration pneumonia triggering thyroid storm.
- Resolution of proximal myopathy by 6 weeks & dysphagia by 3.5 months with removal of NG

Management of Thyroid Storm

Block & Replace therapy with thyroxine commenced at 7 weeks in addition to Carbimazole just prior to discharge home

What is known?

- Thyroid storm is rare in children with ~ 30 cases reported (youngest 33 months) & there is limited evidence guiding management
- Diagnosis of thyroid storm in children is complicated by the lack of paediatric-specific diagnostic criteria.
- Thyrotoxic myopathy presenting with dysphagia is rare but has been reported in adults, lasting up to 14 weeks
- Myasthenia gravis may be associated with autoimmune disorders such as Graves disease & thus is an important differential to consider in dysphagia

What does this case add?

- Thyrotoxicosis/Graves disease should be considered as a differential in young children with tachycardia
- This is the youngest reported case of thyroid storm & only reported paediatric case of thyrotoxic myopathy presenting with dysphagia
- Aspiration pneumonia secondary to thyrotoxic bulbar myopathy/dysphagia may precipitate thyroid storm
- Dysphagia secondary to bulbar thyrotoxic myopathy can be prolonged