CASE 7

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• 33-year-old woman three weeks after the delivery presented with raised inflammatory parameters and abdominal pain

• Diagnostic laparoscopy was performed and appendix removed
No signs of acute inflammation
serosa was slightly hyperemic
• Discontiguous small clumps of cells arranged circumferentially around the tip of the appendix

• Proliferation of large polygonal cells with copious eosinophilic cytoplasm and small inconspicuous nuclei
Granular cell degeneration of appendiceal smooth muscle
<table>
<thead>
<tr>
<th>Immunohistochemistry</th>
<th>Result</th>
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<tbody>
<tr>
<td>α-SMA</td>
<td>+</td>
</tr>
<tr>
<td>Desmin</td>
<td>+</td>
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<tr>
<td>NKI-C3</td>
<td>+</td>
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<tr>
<td><strong>HMB 45</strong></td>
<td>+</td>
</tr>
<tr>
<td>S100</td>
<td>-</td>
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<tr>
<td>Melan A</td>
<td>-</td>
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<tr>
<td>CD117</td>
<td>-</td>
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<td>CD68</td>
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</tbody>
</table>
• HMB-45 may cross-react with some other proteins probably in lysosomes in these cells

• HMB-45 specificity!
Ultrastructure: sparcity of cellular organelles
Heterogeneity of granules
Small and large electron optic dense vesicles with coarse material
Artefacts due to deparaffinisation
Ultrastructural findings

- Transition between smooth muscle and granular cells were demonstrated.

- The ultrastructural findings were similar in both types of cells.

- Actin-like filaments and condensation noted at the periphery of granular cells were identical to those of the smooth muscle cells.

Sobel et al. Arch Path 1971
Granular cell degeneration of appendiceal smooth muscle

Described in 1971, no further cases in the literature

Incidence: about 5% of appendices in multiple sections

Neither palpable nor grossly apparent

In the inner muscular layer

Response to the injury (luminal stasis or inflammation)

Sobel et al. Arch Path 1971
Granular cell degeneration of appendiceal smooth muscle

Isolated nodules of granular cells in the submucosa represent granular cell transformation of muscularis mucosa
Other parts of the GIT?

- In the pylorus
- In the distal oesophagus
- Ascending colon, cecum and rectum
- In the distal portion of the common bile duct

Stout, 1953, 1955, 1957
Azzopardi, 1956
Other parts of the body?

- In the wall of the bronchial tree
- In the uterus
- In the neurohypophysis or its stalk

Churg and Work, 1959
Conclusion

Granular cell degeneration is defined by granular morphology and distinctive circumferential growth pattern.

Differential diagnosis – tumors (PEComa, granular cell tumor)

HMB-45 positivity!
Conclusion

- The granular cells closely resemble smooth muscle cells
- Response to the injury
- No clinical significance

- Modified smooth muscle cells (Churg and Work, 1959)