A Polypoid Tumour in the Duodenum

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Clinical History

In this case, endoscopic biopsies from the duodenum were submitted in 2012 from a 74 year-old patient with abdominal discomfort.

Upon endoscopy, several small polypoid protrusions in the second part of the duodenum were seen. No other remarkable findings where reported during endoscopy in the gastric mucosa or upper jejunum.

After histological examination, the referring clinician was contacted. He reported that the patient was otherwise healthy with no clinical history of malignant disease, and no other manifestation of a neoplastic process. The patient was followed for 7 years, and regularly biopsied once a year until 2019. In all these biopsies, similar findings were reported.
Follicular Lymphomas in all its Sites and Shapes
Variants and Subtypes – WHO 2017

- FL grades 1/2
- FL grade 3A
- FL grade 3B

- BCL-2 expression negative FL 1/2 (10%), 3A (30%)
- t(14;18) negative FL

- Predominantly diffuse nodal follicular lymphomas

- Pediatric-type follicular lymphomas

- Primary extranodal follicular lymphomas
  - Primary cutaneous follicle centre lymphoma
  - Duodenal-type follicular lymphoma
  - Other extranodal follicular lymphomas
    - Ocular adnexae, breast, testis, thyroid gland

*With particular biological and/or clinical characteristics*
Duodenal-Type Follicular Lymphoma

A  Endoscopy. White nodules in the second portion of the duodenum.
B  Lymphoma cells in neoplastic follicles and villi.
C  Uniform lymphoma cells without tingible-body macrophages.
D  CD21-positive FDCs locate at periphery of follicles.
E  CD20-positive lymphoma cells in the villi as well as follicles.
F  Very low Ki67 index.
G  Lymphoma cells positive for CD10.
H  Lymphoma cells strongly positive for BCL2.
Primary Intestinal Follicular Lymphoma
Follicular Lymphoma of the Duodenum

Genetic data

PCR: t(14;18)+ in 12/18
FISH: t(14;18)+ in 7/8
Karyotyping: 4/4 t(14;18) as sole aberration on the level of the

*BCL2* Fusion

Vienna data: Courtesy of Dr. Andreas Chott
Duodenal-type and nodal follicular lymphomas differ by their immune microenvironment rather than their mutation profiles

BCL2 Pseudonegative FL (Using Dako Clone)

- 10-15% of FL 1/2 reported to be BCL2 expression negative
- Two groups of Bcl2 Clone-124 negative FL
  - 50% also negative in IHC using Bcl2 clones E17 and SP66
    - No t(14;18), no Bcl2 mutation,
  - 50% of cases: Bcl2 IHC+ using clones E17 and SP66
    - t(14;18)+, missense Bcl2 mutation

Burkhart et al, Hematol Oncol 2014
Adam et al, Hum Pathol 2013
Hoeller et al, Hum Pathol 2012
> DNA of sufficient quality

> No amplification: Mutation of the primer binding site?
Table 5. Tumor distribution among cases of CD10⁺ follicular lymphoma (FL), CD10 downexpressed FL, and MALT lymphoma

<table>
<thead>
<tr>
<th></th>
<th>Esophagus</th>
<th>Stomach</th>
<th>Small intestine</th>
<th>Large intestine</th>
</tr>
</thead>
<tbody>
<tr>
<td>FL</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CD10⁺ (n = 137)</td>
<td>0</td>
<td>5</td>
<td>133</td>
<td>4</td>
</tr>
<tr>
<td>CD10 downexpressed (n = 35)</td>
<td>0</td>
<td>7</td>
<td>35</td>
<td>3</td>
</tr>
<tr>
<td>MALT lymphoma (n = 1024)</td>
<td>2</td>
<td>815</td>
<td>32</td>
<td>176</td>
</tr>
</tbody>
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