Role of Surgery in Linitis Plastica

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Gastric linitis plastica: which role for surgical resection?

Managing Gastric Linitis Plastica
Keep the scalpel sheathed

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Overview

- Classification
- Challenges in Diagnosis
- Management
• Classification – Borrmann/Lauren
• Current Staging
• Resection Margins
1926 – Borrmann – based on how tumor looks like

1965 – Lauren – based on how it looks like under a microscope

1990 – WHO etc...
### Gastric Cancer

#### Table 1 - Continued

American Joint Committee on Cancer (AJCC)

TNM Staging Classification for Carcinoma of the Stomach

(7th ed., 2010)

<table>
<thead>
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<th>Anatomic Stage/Prognostic Groups</th>
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- T1 N2
- T2 N2
“R” Status

- R0 – No Macroscopic or Microscopic Margin
- R1 – Microscopic residual cancer (+ margin)
- R2 – Gross residual cancer
What is Linitis Plastica?
What is Linitis Plastica?

Progressive decrease in the incidence of the intestinal type of gastric cancer and an increase in the diffuse type of gastric carcinoma.

It was defined as a distinct entity in 1859 by Dr. William Brinton, who described it as a benign disease.

Variant of gastric adenocarcinoma found in 7–14% of cases. It represents a diffusely infiltrative and desmoplastic process usually involving the entire stomach giving it stiffness described as “leather bottle stomach.”
“Linitis” was due to the presence of irregular bands of filamentous tissue in the hypertrophic submucosa, resembling fibers of linen.

Malignant cells, when detected, were often described as few and scattered. As a consequence, for many years, it was controversial if the condition was benign or malignant.
Case

- 64 y/o F who presented last year in December with Sx. of Dull epigastric pain and post prandial discomfort.
- EGD - Revealed Gastritis
- Sent home
She came back to ER 2 months later with similar symptoms

CT – thickened wall of stomach and some perigastric lymph nodes

She was sent over to GI for a repeat EGD that showed a **Borrmann 3** lesion along the anterior wall and greater curvature. Biopsy suspicious for Adenocarcinoma.

Diagnostic Laparoscopy – Lymph nodes – positive for adenocarcinoma
So is Linitis Plastica a Borrmann 4 or Lauren diffuse or Scirrhous?
Linitis Plastica doesn’t fit into any particular classification

LP is often improperly defined as a Borrmann IV tumor

While LP type GCs involve diffuse spreading into all layers, Bormann type 4 AGCs are typically confined to a specific area (e.g., the antrum) and do not contain meandering hypertrophic mucosal folds
- Poor prognosis - difficulty of early detection and the unique developmental process of this cancer.

- Endoscopy typically does not detect abnormalities until several months after disease onset - Late Dx
The most characteristic feature of LP is the macroscopic thickening of the stomach wall, diffusely involving the entire stomach.

Cancer cells trigger a stromal reaction involving fibrosis – like Scirrhous tumors.

LP does not always present as complete involvement of the stomach. It may appear in plaques which gives the appearance of a segmental lack of distensibility.
The concept of linitis plastica is unique - it links diverse aspects of specific subtypes of tumors (macroscopic, microscopic, and environmental).

Cannot be classified as a distinct entity.
Diagnostic Challenge

- Typical symptoms are dyspepsia, nausea, vomiting, and anorexia.

- Unfortunately, those symptoms are not reliable for establishing a timely diagnosis, as they usually present insidiously, and manifest only in an advanced stage.
• **Endoscopy** is considered the gold standard for the diagnosis of GC in general.

• LP tumors involves primarily the submucosa and muscularis propria of the stomach. Negative in 30% of cases.

• Due to their poorly cohesive nature, cancer cells are often scattered between the tumor stroma
**EUS** can show submucosal and muscular thickening, and EUS fine-needle aspiration allows reaching of the submucosal layer. Even with this strategy, however, negative biopsies have been reported.
CT allows for comprehensive staging of the tumor, and could give rise to reasonable suspicion when identifying a stomach with thickened walls.

Two studies have described a specific enhancement pattern in LP patients.
• **PET**, has poor diagnostic significance, as poorly differentiated, diffuse, mucinous carcinomas have all been reported to have a low uptake.

• **MRI** - Alternative to CT, due to its advantages in characterizing tissue nature and obtaining soft tissue contrast, but the topic is still controversial.
• Suspicion of linitis plastica should include **laparoscopy** for staging.

• In consideration of the well-known peritoneal tropism of the disease, a diagnostic laparoscopy with peritoneal washings should be **mandatory** to **complete** the staging.
A gold-standard diagnostic instrument for LP has yet to be defined.

In the absence of clear definition for LP, the development of a diagnostic strategy is difficult.

Future diagnostic advancements may be obtained by the use of blood-based biomarkers.
Prognosis

- Difficult to classify.
- Difficult to diagnose.
- Presents at an Advanced Stage
- Poor prognosis with a five year survival of 3-10% in various studies.
Controversy in Management

- The question is whether surgical resection is suitable for these patients or to begin with a nonoperative treatment.

- Symptoms at the time of presentation are nonspecific leading to delayed diagnosis.

- Lymph node involvement - always present at the time of diagnosis, and due to its diffuse nature, microscopic disease is often found at the resection margins.

- Peritoneal dissemination is frequently encountered at the time of surgery, reason for recurrence.
Curative resection is possible in less than half of patients and early recurrence is common, leading to a median survival, ranging from 6 to 12.

In series of select resectable gastric LP, more than half of the patients had T4 tumors and 83% had involved perigastric lymph nodes.
Outcomes in these earlier studies were discouraging.

The overall poor prognosis of LP gastric cancer has led some authors to conclude that LP is not a surgical disease, and many oncology providers remain biased against surgical resection for gastric LP.


**Gastric linitis plastica is not a surgical disease.**

Aranha GV¹, Georgen R.
Complete surgical resection (R0) has consistently been associated with improved survival in LP patients.


1114 total gastrectomies in the surgical treatment of primary gastric adenocarcinoma--a 30-year single institution experience.

Jähne J¹, Piso P, Meyer HJ.
Treatment

• Resection margins (>5 cm) have been advocated to avoid R1 resection, and they are considered the current standard for patients with Borrmann III and IV tumors, in accordance with the Japanese Guidelines.

• However, studies focusing on LP tumors are lacking.

• So let do a frozen tissue biopsy
But ...

It should also be considered that in scirrhous gastric cancers and LP phenotypes a frozen tissue could be less reliable due to the lack of tumor cellularity.
Chemo?

- As these tumors present in an advanced stage, Neoadjuvant therapy may work great.
- Studies were done to assess chemo- and radiosensitivity in LP tumors.
Currently, targeted therapy is considered only for certain Gastric cancer subtypes, and almost all the chemotherapeutic regimens are directed solely against the cancerous cells.
But ...

The desmoplastic stroma may represent both an enhancer of tumor cell growth and invasiveness and a shield against the host’s immune response and against standard chemotherapy.

Which means – it may or may not work.

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Intrinsic subtypes of gastric cancer, based on gene expression pattern, predict survival and respond differently to chemotherapy.

Currently, appropriate management strategies include an accurate diagnostic multi-instrumental assessment and diagnostic laparoscopy in all cases.

Aim – Curative surgery, when feasible, should be performed.

Further advancements are needed in regards to the development of targeted therapies, which will address both cancer cells and their stroma.