

#### Title:

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DOI: 10.17235/reed.2020.7323/2020 Link: <u>PubMed (Epub ahead of print)</u>

Please cite this article as:

Ortega Lobete Olga, Diaz Ruiz Raquel, Perez Carazo Leticia. PRIMARY GASTRIC ADENOCARCINOMA IN PATIENT WITH PEUTZ-JEGHERS SYNDROME. Rev Esp Enferm Dig 2020. doi: 10.17235/reed.2020.7323/2020.



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## CC 7323

# PRIMARY GASTRIC ADENOCARCINOMA IN PATIENT WITH PEUTZ-JEGHERS SYNDROME

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Keywords: Peutz-Jeghers. Gastric adenocarcinoma. Capsule endoscopy.

Dear Editor,

We present the case of a 42-year-old woman under study for dyspepsia without response to empirical treatment. She has perioral mucocutaneous pigmentation.

Gastroscopy reveals a 5cm gastric tumor in the antrum, as well as multiple sessile gastric and duodenal polyps smaller than 1 cm (fig 1a). Obtaining large-capacity forceps biopsies, a well-differentiated adenocarcinoma with gastric origin is diagnosed, with over expression of the HER2/neu without microsatellite instability in the context of a hamartomatous polyposis. The genomic study confirms an alteration in the STK11 gene translated to a truncated protein product, compatible with a Peutz-Jeghers syndrome (PJS).

In the endoscopic ultrasound study the lesion appears as a heteroecogenic mass with anechoic areas and microcalcifications, arising from the superficial layers of the gastric wall, respecting the muscular layer. The lesion contacts without infiltrating the pancreatic parenchyma which is normal (fig 1b).

The study is completed by small-bowel capsule endoscopy (CE) in which multiple micropolyps are observed all along the duodenum, jejunum and ileum; detecting several of them larger than 10mm in the proximal jejunum and the ileum. CT scan shows lymph



node and peritoneal involvement. Given the extension, systemic therapy is started with partial response. It is decided to perform cytoreductive surgery (total gastrectomy, right hemicolectomy due to wall implants and resection of distal ileum due to larger than 10mm polyps) and large polyps mucosectomy in the proximal jejunum by anterograde enteroscopy, after which a second-line chemotherapy is started.

### **DISCUSSION**

PJS is a hamartomatous polyposis that is characterized by a high cumulative risk of cancer in different locations; although gastric malignancy (adenocarcinoma), as in the case we present, is not the most common (1-2).

In this type of polyposis, it is important to study the small bowel using CE to define number, location and size of the polyps, as well as identifying the enteroscopy access route which is indicated in symptomatic patients, polyps larger than 10mm or rapid growth (2-4).

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Figure 1. Endoscopic view of well-differentiated gastric adenocarcinoma in Peutz-Jeghers syndrome. In lower image measured with endoscopic ultrasound.