specific differentiation.^{1,10} Therefore in 2012, Stockman *et al.* first designated the distinct entity as ‘malignant gastrointestinal neuroectodermal tumour’.¹

According to available current data, the biological behaviour of malignant GNET is highly malignant and almost all of the reported cases followed an aggressive clinical course with local recurrence in the short term, and metastasis to lymph nodes or haematogenous spread early or even at initial diagnosis. However, our case presented an indolent clinical course. The patient had evidence of the tumour from the age of 37, which progressed slowly for 10 years without any medical intervention. After excision of the tumour, the patient was alive without evidence of disease for 8 months. Interestingly, Prado *et al.* have reported one case of indolent GNET of the colon which also had a 10 year long clinical course.¹¹ Our case still needs further follow-up observation to evaluate long-term outcome.

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Parietal cell/oncocytic/mitochondrion-rich gastric carcinoma: a need for clarity in terminology

Sir,

In 1984 Capella *et al.* introduced three cases of a newly observed variant of gastric adenocarcinoma, which they named parietal cell gastric carcinoma (PCGC).¹ A small number of case reports followed this first description of the entity until 2002.² PCGCs are primary adenocarcinomas of the stomach that display a glandular or solid pattern of growth occasionally mimicking lymphomas or mesenchymal tumours of smooth muscle origin.² Neoplastic cells are usually polygonal with round nuclei and oncocytic features with abundant eosinophilic, granular cytoplasm reactive with phosphotungstic acid haematoxylin (PTAH) and Luxol fast blue.^{2–4} Commonly, immunohistochemical stains for antimitochondrial antibodies are strongly positive.^{2,3} The ultrastructural features of tumour cells correspond to an ‘oncocytic cytoplasm’ with abundant mitochondria, tubulovesicles, intercellular lumina filled with undulating microvilli and intracellular canaliculi.^{2–5}

In 2002 Takubo *et al.* presented 10 cases of gastric adenocarcinoma with morphological features similar to those of parietal cell carcinoma, however immunohistochemical staining of the tumours with four different antiparietal cell antibodies (anti-H⁺-K⁺-adenosine triphosphatase antibodies) was negative in all cases.⁶ They claimed that, if the H⁺/K⁺-ATPase proton pump is considered an important property of normal parietal cells, then the 10 carcinomas they encountered could not be said to have shown parietal cell differentiation. Therefore, they used the term oncocytic carcinoma, or carcinoma with oncocytic differentiation, to describe the presented neoplasms.⁶ They also suggested that their cases may represent a different histological subtype from parietal cell carcinoma, however none of the previously reported cases of parietal cell carcinoma had been tested with antiparietal cell antibodies.⁶ Subsequently, in 2005, the same group reported another four cases of the same type of gastric adenocarcinoma and, again, negative staining with antiparietal cell antibodies.⁷ Additionally, they emphasised similarities of these cases with malignant oncocytomas to suggest the term oncocytic carcinomas or carcinomas with oncocytic differentiation.⁷

In 2003 Yang *et al.* presented the case of a gastric tumour with both parietal cell and incomplete intestinal type differentiation, and found that the tumour cells were strongly and extensively positive for parietal cell biomarkers H⁺/K⁺-ATPase and HMFG2 (human milk fat globule-2).⁵ They agreed with the necessity of immunostaining for parietal cell markers to confirm a morphological suspicion of parietal cell differentiation.⁵ HMFG-2/platelet derived growth factor receptor- α (PDGFR α) is currently considered a typical biomarker of parietal cell differentiation.⁸

Following the publication by Takubo in 2002, the terms ‘parietal cell’ and ‘oncocytic gastric carcinoma’ have been alternately used in the literature, however it still remains unclear whether the terms represent different entities.² Additionally, it has been suggested recently that there may be a partial overlap of such neoplasms with ‘gastric adenocarcinoma of the fundic gland type with predominant parietal cell differentiation’.⁸

Both tumours reported so far either as parietal cell or oncocyctic carcinomas appear to have received similar forms of treatment and have a similar favourable prognosis.^{2,6} Given the extensive morphological similarities of these tumours, if it were to be accepted that the different names represent different entities or subtypes of the same entity, the biological significance of this distinction should be clarified, and the potential implications for treatment and prognosis.

The rarity of primary gastric adenocarcinomas with an oncocyctic morphology implies that more cases must be studied before the biological significance of novel efforts in the field of histological classification can be ascertained.²

In 2010, Caruso *et al.* reported on nine cases of gastric adenocarcinoma with eosinophilic cytoplasm predominantly in the supranuclear portion, for which they suggested the term 'mitochondrion-rich carcinomas' (MRC). They chose this term as the tumour appeared to be the gastric counterpart of mitochondrion-rich follicular carcinoma of the thyroid. They underlined that none of the lesions had densely packed mitochondria as is characteristically found in oncocyctomas. They highlighted the fact that a distinction between true oncocyctic neoplasms and MRC of the thyroid was recently available, and argued that care was taken not to confuse oncocyctic carcinomas with MRC in their observation.⁹ Their cases were characterised by small size, rare lymph node metastases, and low stage at diagnosis. All nine patients underwent curative gastric resection and were still alive without evidence of recurrence at a median follow-up of 42 months (range 1–90), suggesting a low grade malignancy with an excellent prognosis. Note was made of the fact that mitochondria can accumulate only in cells of tumours that are not actively dividing, thus indicating a low proliferative turnover and, commonly, association with benign tumours or tumours of low malignancy.⁹ However, 2 years later the same cohort of nine patients was used in a study of HER2 status in unusual histological types of gastric adenocarcinoma, while this time the term oncocyctic adenocarcinomas was used as equal to MRC, although without explaining this change of approach.¹⁰ Further studies of the same group on these tumours utilised the term MRC exclusively.¹¹

Obviously, despite differences of uncertain significance that have been emphasised upon describing the particular features of each series, it remains unclear whether parietal cell gastric carcinoma, oncocyctic gastric carcinoma and mitochondrion-rich gastric carcinoma represent three or two different entities, or one single entity, or subtypes of an entity. Especially with the understanding that they account for rare histotype(s) of gastric adenocarcinoma for which acquisition of more data will allow for extraction of solid conclusions, the remaining lack of clarity in terminology impedes the process of reviewing these tumours, as has recently been underlined.² In the current World Health Organization (WHO) classification of tumours of the stomach (5th edition), parietal cell carcinoma is included, and attention is paid to oxyntic gland adenoma with transition to gastric adenocarcinoma of the fundic gland type, which displays predominant parietal cell composition in <1% of reported cases.¹² However, little attention appears to have been paid to the aforementioned confusion in the literature between entities or possible subtypes that may exist.¹²

We think it would be prudent that, should no further clarity on the matter arise in the meantime, these facts are taken into account prior to the next update of the WHO classification of

gastric carcinoma, so that a clear and solid common language is used, that will facilitate the process of understanding better this group of uncommon tumours.

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Isolated abdominal kaposiform lymphangiomatosis: a novel presentation of a rare entity

Sir,

Kaposiform lymphangiomatosis (KLA) is a rare and novel disorder of the lymphatic vascular system which was first described by Debelenko, Perez-Atayde, and Croteau *et al.*^{1–3} at the Boston Children's Hospital. It has been described as a