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 oncocytomas. 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

distinct histopathological entity within the subtype of generalised lymphatic anomalies (GLA) characterised by foci of 'kaposiform' spindled lymphatic endothelial cells surrounding malformed lymphatic channels.³ To date, all reported cases have included thoracic involvement, with other reported sites including bone, spleen and retroperitoneum in addition to thoracic disease.

An otherwise healthy 22-year-old Vietnamese woman presented with a 2 day history of epigastric pain radiating to her right abdomen. The pain was exacerbated by meals and was associated with nausea and vomiting. Her bowels were opening with no issues and she reported no fevers, dysuria or other infective symptoms. On examination, her abdomen was soft and she was tender in the epigastrium and right upper quadrant. Initial blood tests were unremarkable, including a normal platelet count $303 \times 10^9/L$ and fibrinogen of 3.2 g/L. Ultrasound of her abdomen showed a mass in the mid abdomen, with associated lymphadenopathy. Further evaluation with computed tomography (CT) of her abdomen and pelvis confirmed a $11 \times 4.5 \times 11$ cm mass encasing the superior mesenteric artery and vein at the root of the mesentery (Fig. 1A,B). Magnetic resonance imaging (MRI) demonstrated a lobulated, well defined mesenteric root mass which was hyperintense on T2-weighted sequences (Fig. 1C). Given the suspicion for malignancy, staging CT scans of her chest and neck were performed which showed no lung lesions or other lymphadenopathy. With the aim of obtaining tissue diagnosis, endoscopic ultrasonography was performed, but was unable to visualise the lesion. Finally, a diagnostic

laparoscopy was performed. Intra-operatively, the mass had the appearance of oedematous fatty tissue (Fig. 1D,E). As it was not possible to safely resect the mass or perform a core biopsy, an incisional biopsy was performed with ultrasonic shears.

Histopathology of the biopsy showed vascularised adipose tissue in keeping with mesentery. Within the mesentery were dilated malformed thin walled vessels, some with discontinuous mural bundles of smooth muscle, and with scattered lymphoid aggregates in the adjacent stroma, in keeping with malformed lymphatic vessels (Fig. 2A). Clusters of short spindle cells with small slit-like vascular channels were also present in areas accentuated around malformed lymphatic vessels (Fig. 2B). Occasional mast cells were present, but no plasma cells, granulomas or large or atypical lymphocytes were seen. The spindle cells were confirmed to be of lymphatic endothelial origin, staining strongly positive for markers CD34, ERG and D2-40 (Fig. 2C–F). The endothelial cells lining the smaller vascular spaces stained positive for CD34 and D2-40, and the endothelial lining of all sizes of vascular spaces stained positive for ERG, further confirming a lymphatic vascular process (Fig. 2C–F). Smooth muscle actin was positive only in the smooth muscle within walls of the larger vascular spaces, and S-100 staining was positive in adipocytes only, with both negative within the spindle cells. The spindle cells were also negative for Melan-A, HMB45, keratin AE1/AE3, SOX 10 and MDM2.

The overall histological features were consistent with KLA, with immunohistochemistry confirming the lymphatic

Fig. 1 Computed tomography (CT), magnetic resonance imaging (MRI) and intra-operative. (A) CT abdomen pelvis coronal view, (B) CT abdomen pelvis sagittal view, (C) T2-weighted sequence MRI demonstrating hyperintensity of the lesion and its relationship to major abdominal vascular structures (axial view). (D,E) Intra-operative images of the root of mesentery, demonstrating a smooth, firm soft tissue mass. Circled = lesion. A, abdominal aorta; IVC, inferior vena cava; SMA, superior mesenteric artery; SMV, superior mesenteric vein; PV, portal vein.

Fig. 2 Histopathology from biopsy. (A) H&E, large cavernous blood vessels within background adipose tissue. (B) H&E. Aggregates of small spindle cells in short fascicles with slit-like vascular spaces in association with dilated lymphatics. (C) CD34. Strongly positive staining of small spindle cells supporting endothelial origin. (D) D2-40. Strongly positive staining of small spindle cells and endothelial cells confirming lymphatic vascular process. (E) D2-40, higher power. Strongly positive staining of small spindle cells and endothelial cells confirming lymphatic vascular process. (F) ERG staining. Strongly positive staining of small spindle cells and endothelial cells lining vascular spaces. Circled = aggregates of spindle cells. A, adipose tissue; V, large vascular spaces.

endothelial nature of the malformed vessels and spindled lesional cells. Further X-rays of her skull, long bones, spine, pelvis and chest found no osseous involvement. She has since had improvements in her abdominal symptoms following commencement of treatment with sirolimus.

KLA represents a rare and relatively novel lymphatic disorder which predominantly affects children, with a median age of symptom onset and diagnosis of 6.5 and 8.5 years, respectively, with ages ranging from birth to 44 years.³ The oldest reported age of diagnosis in the literature is 50 years.⁴

Of the 20 cases described by Croteau *et al.*, 50% initially presented with respiratory symptoms such as cough or dyspnoea, and 50% presented with bleeding.³ Ultimately, 95% of patients in this series developed pericardial and/or pleural effusions with all but one requiring drainage.³

Consumptive coagulopathy was also described as an uncommon complication.³ With regards to anatomical distribution, the authors found thoracic involvement in all patients (100%) with bone (50%) and spleen (50%) being the next most common regions being involved.³ Another large case series of 20 patients by Goyal *et al.* similarly found 100% intrathoracic involvement, 35% of patients had splenic involvement and 80% had evidence of retroperitoneal disease.⁵ The retroperitoneal disease was characterised by enhancing infiltrative soft tissue, almost always involving the retrocrural region with extension into the mesentery (56% of retroperitoneal cases).⁵ On imaging with MRI, they were hyperintense on fluid weighted sequences.⁵ Consistent with this case series, other smaller studies have found invariably thoracic involvement.^{4,6–9} Therefore, this case is unique in

that involvement at diagnosis was confined to the abdominal cavity with involvement of the retroperitoneum extending into the root of the mesentery, and no evidence of intrathoracic disease.

Histopathologically, KLA is distinguished by foci of 'kaposiform' spindled lymphatic endothelial cells, often arranged in parallel fashion either as dispersed, poorly marginated clusters or anastomosing strands.³ Further immunohistochemical staining, such as with PROX-1, D2-40, ERG, CD31 or CD34 can confirm the presence of these lymphatic endothelial cells.^{7,9,10} This is distinct from kaposiform haemangioendothelioma (KHE), another disorder associated with abnormal lymphatic channels and spindled cells, however the histological architecture is of more defined solid confluent tumour, often with microthrombi.³ Clinically, KHE differs further as the lesions are typically unifocal vascular tumours presenting in infancy, which may involve skin and soft tissues, retroperitoneum or viscera,³ with larger lesions commonly associated with thrombocytopenia resulting in consumptive coagulopathy (Kasabach–Merritt phenomenon).

There is currently no standard treatment for KLA and management is directed at symptom control and management of complications such as drainage of pericardial or pleural effusions.^{3,7,9} Pharmacotherapy options such as sirolimus, an mTOR inhibitor, have been utilised in other vascular anomalies with promising results.¹¹ Croteau *et al.* reported clinical improvement in three patients following sirolimus therapy,³ and other case studies have also reported reduction in lesion size or improvement in symptoms^{6,8,9} or stable disease.⁷ One patient in a case series by Fernandes *et al.*, showed initial response to sirolimus and a 5 month period of remission before subsequent reaccumulation of pleural and pericardial effusion, and development of epidural haemorrhage resulting in fatal cardiac arrest.⁷ Limited survival and prognostic data exist for KLA; however, Croteau *et al.* reported a 5 year survival of 51% and an overall survival of 34%.³ Mean interval between diagnosis and death was reported as 2.75 years with cardiorespiratory failure being the most common cause.³ Of note, there was one patient in the series who presented as an adult, never required any interventions, and denied any issues on follow-up.³

In conclusion, KLA is a rare but emerging entity classically presenting as intrathoracic lesions in young patients. However, this diagnosis should also be considered in patients with infiltrating masses in the retroperitoneum even when intrathoracic disease is not present, as demonstrated with this case.

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Non-mammary implant associated anaplastic large cell lymphoma

Sir,

Anaplastic large cell lymphoma (ALCL) constitutes less than 3% of all non-Hodgkin lymphomas (NHL).¹ Implant associated ALCL was first reported by Keech and Creech² in 1997 in a patient with a breast implant. In the last two decades, several case reports and small case series of breast implant associated ALCL (BIA-ALCL) have been reported in the literature and this is now a provisional entity in the current World Health Organization (WHO 2017) classification of hematolymphoid neoplasms.³ The currently estimated incidence of BIA-ALCL in the United States is 33 per one million persons with textured breast implants.⁴ The textured implants are associated with a higher risk of disease development as compared to saline implants.⁵ In 2010, Palraj *et al.*⁶ reported ALCL in association with a stainless steel implant for complex right tibia fracture, and recently Mendes *et al.*⁷ documented ALCL following augmentation gluteoplasty by silicone implant. In the literature, only two cases of non-mammary implant associated extranodal ALCL have been reported. We present a case of a male with a history of oral cavity squamous cell carcinoma for which he underwent marginal mandibulectomy and device implantation (titanium; smooth surface) and developed ALCL 4 years later. To the best of our knowledge, this is the second case of non-silicone prostheses associated ALCL and the third case of implant associated ALCL at the non-mammary site.

A 63-year-old man, with well controlled diabetes and hypertension, had a non-healing ulcer over the left buccal mucosa in 2016. On oral examination an ulcero-proliferative growth measuring 3×3 cm was seen in the left buccal mucosa involving the retromolar trigone posteriorly and reaching the