



Case Report

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Diagnosis of Papillary Intralymphatic Angioendothelioma in the Abdominal Viscera: A Case Report

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**Citation** Salam Raju A, de Silva D, Farooque Y, Rutland T. Diagnosis of Papillary Intralymphatic Angioendothelioma in the Abdominal Viscera: A Case Report. Case Reports in Clinical Practice. 2024; 9(6): 266-271.

DOI:10.18502/crcp.v9i6.18946

Running Title Papillary Intralymphatic Angioendothelioma in the Abdominal Viscera**Article info:****Received:** October 29, 2024**Revised:** November 24, 2024**Accepted:** December 18, 2024**Keywords:**

PILA; Intralymphatic angioendothelioma; Dabska tumour

ABSTRACT

Papillary Intralymphatic Angioendothelioma (PILA), or Dabska Tumour, is an exceedingly rare lymphovascular neoplasm of intermediate malignancy, predominantly affecting cutaneous and soft tissue sites. This case report highlights an unusual presentation of PILA in the mesentery of a 38-year-old male, a site not previously documented. He initially underwent a splenectomy for symptomatic splenic hemangioma. His symptoms recurred two years later, presenting with haemoperitoneum and extensive abdominal involvement, including the colon, small bowel mesentery, and diaphragms. An en-bloc extended right hemicolectomy and partial hepatectomy achieved complete resection.

Histopathology revealed hobnail endothelial cells and characteristic papillary structures, confirming PILA. Despite its rare visceral occurrence and diagnostic challenges, the patient showed no recurrence at six months post-surgery. This case underscores the importance of histopathological analysis and awareness of atypical presentations. Wider surgical excision remains the cornerstone of treatment, with vigilant follow-up essential for detecting recurrence and guiding future management protocols for this rare entity.

Introduction

Papillary intralymphatic angioendothelioma (PILA), also known as Dabska Tumour, is a rare lymphovascular neoplasm of intermediate malignancy that typically arises in the soft tissue of the skin or extremities. It was initially characterised by Maria Dabska in 1969. Originally identified as a malignant childhood tumour, Dabska's case series revealed occurrences in six children, two of whom

had regional lymph node metastasis and one with pulmonary metastasis [1].

PILA was previously referred to as endovascular papillary angioendothelioma (EPA). Most authors classified it as a neoplastic entity that straddles the border between benign lesions, such as hemangiomas, and malignant ones, like angiosarcomas. The classification shifted to PILA in 1998 by Fanburg-Smith et al., owing to its borderline behaviour and prominent lymphatic phenotype [2]. While typically

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observed in the dermis and soft tissues, PILA has also been documented in deeper anatomical locations such as the spleen [3–5], tongue [6], and testis [7]. Remarkably, PILA has been reported as a primary bone tumour in select cases [8].

PILA cases have been reported both *de novo* and within the context of chronic lymphedema or preexisting vascular malformations, such as hemangiomas or cavernous hemangiomas [9]. Although PILA generally carries a favourable prognosis, it presents challenges due to its potential for local recurrence and low-grade metastases.

Due to its rarity, PILA represents a diagnostic challenge, and its treatment approach remains unclear owing to the limited number of reported cases. To date, approximately 50 cases of PILA have been documented in the medical literature [10]. In light of this scarcity, understanding the optimal management strategies for PILA is essential, as is the recognition of this rare tumour.

This case report aims to contribute to the limited body of knowledge on PILA by presenting a new case and detailing both the diagnostic approach and treatment strategy for this extremely rare tumour in an adult patient. As PILA tumours are predominantly documented in case reports, sharing our diagnostic and clinical experience is crucial for enhancing the understanding of this rare vascular neoplasm and refining treatment paradigms.

Case Presentation

A 38-year-old otherwise healthy man was referred to our Upper Gastrointestinal department for symptomatic splenic lesions observed on computed tomography (CT) images. Initial imaging suggested haemangiomas, for which he underwent a laparoscopic splenectomy. He recovered without any post-operative complications. Histopathology of the spleen revealed morphological features consistent with a haemangioma. The specimen was intact.

A follow-up CT scan of his abdomen two years post-splenectomy revealed an unusual mass in his omentum, along with a moderate amount of free fluid. This appeared to be ascites; however, his liver function, including the coagulation profile and blood haemoglobin level, remained within the normal reference range. A CT-guided biopsy of this mass (no peritoneal fluid) showed a vascular lesion with some unusual hobnail cells. Given the complicated recurrence and high risk of spontaneous rupture, an

urgent laparotomy and resection of this mass were arranged.

A midline laparotomy was performed, and approximately four litres of haemoperitoneum were evacuated. Grape-like growths were centered around the transverse colon and transverse mesocolon. These structures encroached on segment 4b/5 (gallbladder fossa) and completely encased the porta hepatis. There was extension into both the right and left paracolic gutters and around the greater curvature of the stomach, with patchy infiltration into the hemidiaphragms. The small bowel appeared normal, with some patchy infiltration into the mesentery. The rest of the pelvis appeared normal. An en-bloc extended right hemicolectomy with segment 4b/5 hepatectomy and partial omentectomy was performed. Postoperatively, he recovered well without any complications and was discharged home. There were no post-operative complications at the six-week follow-up, and no recurrence was seen on repeat CT scan at the six-month follow-up.

The specimens consisted of an extended right hemicolectomy along with separate fragments of tissue from the serosa. An ill-defined lesion composed of cystic spaces of various sizes was observed within the mesenteric fat, extending into the muscularis propria of the transverse colon. No solid areas were present, and the tumour was clear of the margins. This lesion measured 26 × 15 cm at its maximum dimensions. The serosal surface of the fat adjacent to the liver segment contained multiple papillary structures with a similar appearance. No lymph node metastases were identified [Figure 1].

Histologically, most of the cystic spaces were lined by a single layer of flattened endothelial cells resembling vessels. The areas with a macroscopically papillary appearance contained endothelial cells that were hobnailed, with uniform medium-sized nuclei and a small amount of eosinophilic cytoplasm [Figure 2]. The hobnail cells lined small papillary projections or exhibited tufting. Some areas were multilayered or mildly hyperplastic. There was no evidence of atypia, mitosis, or necrosis [Figures 3 and 4].

The vascular spaces were separated by moderately cellular and haemorrhagic stroma. Spindle cells within the stroma exhibited oval to spindle-shaped nuclei without atypia, consistent with reactive fibroblasts/myofibroblasts. Focal mild to moderate chronic inflammatory infiltration, including lymphocytes, plasma cells, eosinophils, iron-laden macrophages, and lymphoid follicles, was present in the stroma. None of the vascular spaces had a smooth muscle layer.

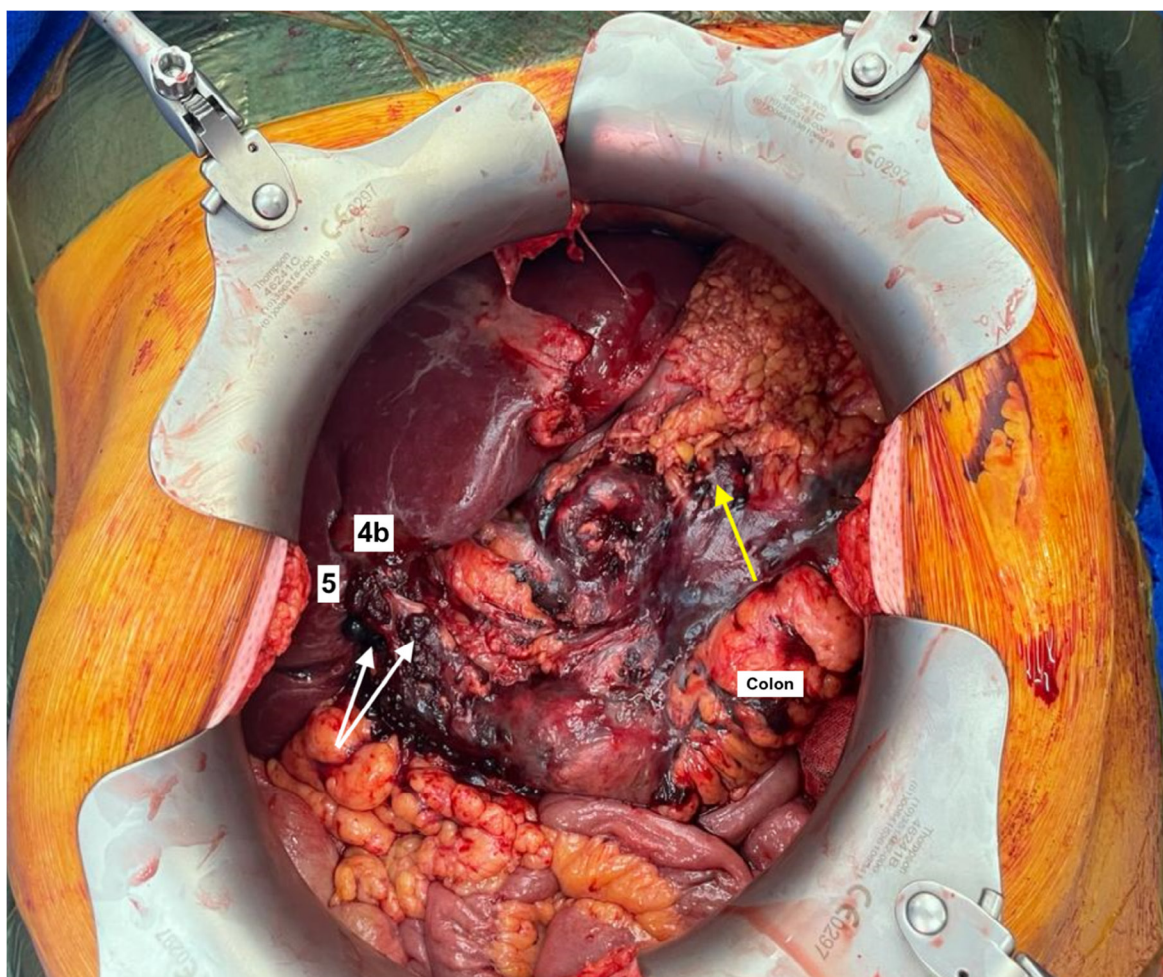


Fig. 1. Intraoperative image. Segment 4b and 5 with grapelike structures along its border (white arrows). Similar grapelike structures along the greater curvature of the stomach (yellow arrow).

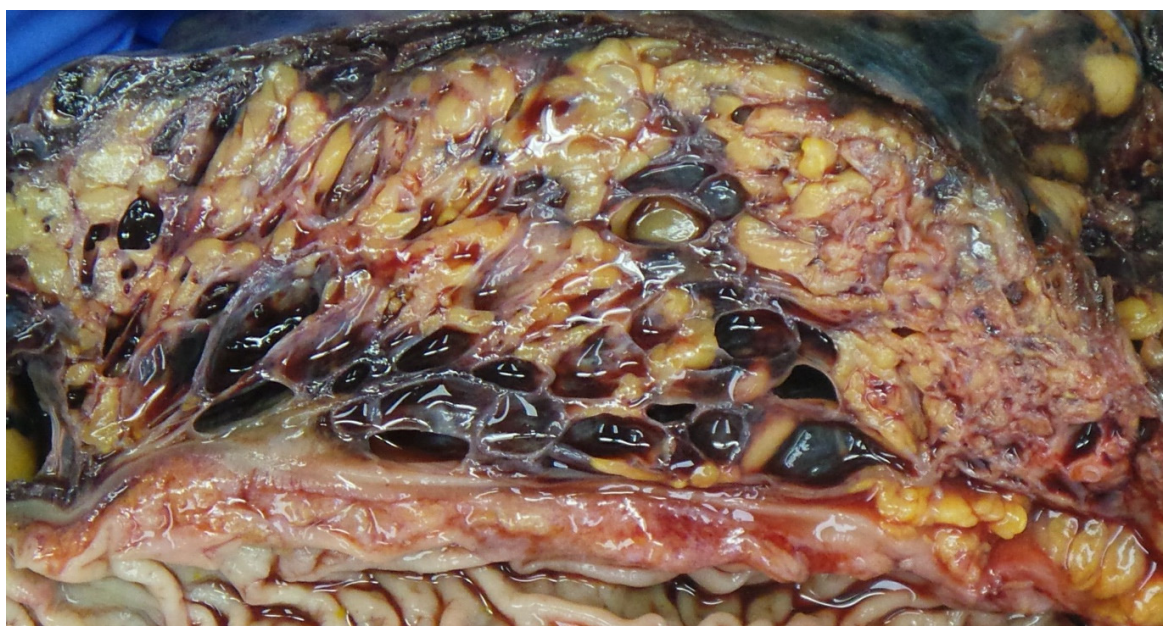


Fig. 2. Macroscopic image of the tumour involving the mesentery.

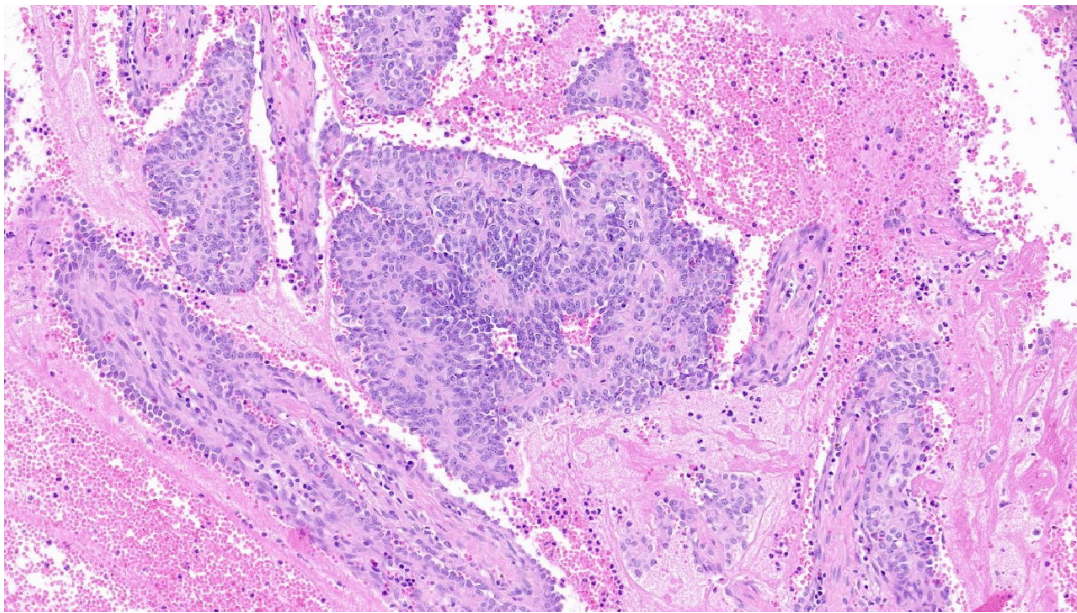


Fig. 3. 200x showing endothelial cells with papillary projections and tufting.

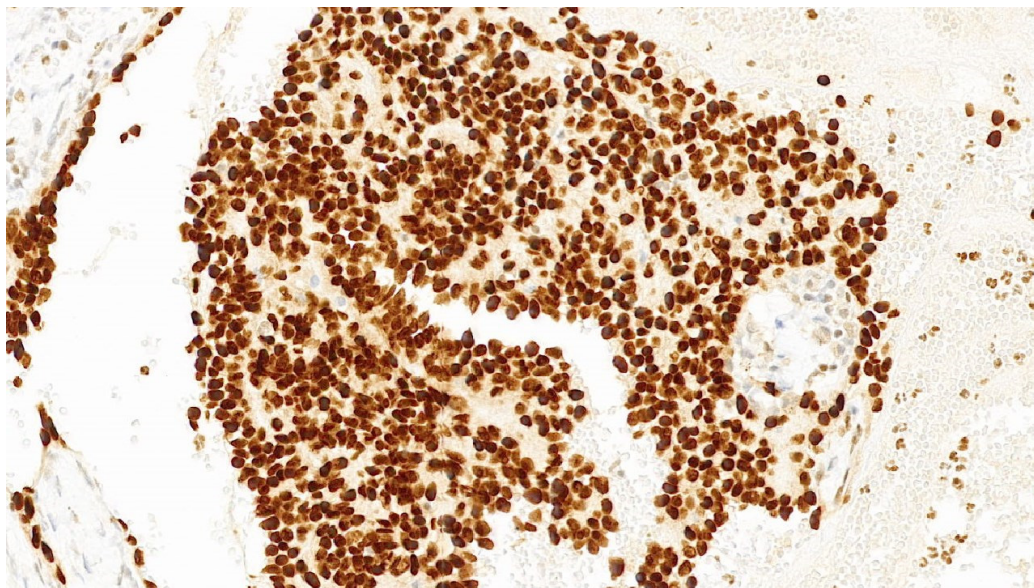


Fig. 4. 200x The endothelial cells are positive for immunohistochemical stain ERG.

Immunohistochemistry revealed that the endothelial cells lining the vascular spaces, including the hobnail cells, were positive for ERG, CD31, and focally for CD34. D2-40, a marker of lymphatic cells [11], showed focal staining of flat cells, with no staining of hobnail cells. No staining was observed for HHV8. Desmin demonstrated no muscle coating around the vascular spaces. Ki67 showed moderate staining of cells in the stroma and the lumens of the vessels, indicating lymphocytes, leukocytes, and reactive fibroblasts. The overall Ki67 proliferation index was low (1%).

Morphologically, these findings are consistent with papillary intralymphatic angioendothelioma (Dąbska tumour) arising from lymphangiomatosis.

Discussion

PILA is an extremely rare lymphovascular tumour, with fewer than 40 cases reported in the literature [12]. Most cases occur in the proximal extremities. PILA occurring in the abdominal cavity is exceedingly rare, with only three case reports documented in

the English literature. Of these, three occurred in the spleen—two in adults [13–14] and one in a five-year-old child [2]. To our knowledge, this is the first reported case of PILA in the mesentery of the colon. The tumour invaded the large bowel wall and extended into the small bowel mesentery. In this 38-year-old male, the atypical vascular tumour was also found in both hemidiaphragms and the falciform ligament.

This is a rare tumour that many histopathologists may not be familiar with, and its occurrence in visceral sites further complicates recognition. This diagnostic dilemma may result in the tumour not being identified or considered in differential diagnoses.

Histopathological features that may aid in diagnosing PILA include an associated lymphangioma or lymphangiomatosis, hobnail or matchstick-like endothelial cells lining intraluminal papillary tufts, and central hyaline cores [15].

Differential diagnosis is further complicated by location. While most PILA tumours occur in cutaneous locations—where differentials include angiosarcomas, hemangioendotheliomas, and benign lymphovascular lesions—visceral tumours can be mistaken for mesotheliomas or metastatic carcinomas, especially on small biopsies. Close attention to morphology and immunohistochemical staining helps differentiate these tumours. Low-grade angiosarcomas can occasionally show intravascular papillations; however, the lack of mitosis and atypia favours a diagnosis of PILA. The expression of lymphatic markers such as D2-40 is useful in distinguishing PILA from other similar hemangioendotheliomas, such as retiform haemangioendotheliomas.

Although regional lymph node metastases have been reported, the prognosis for long-term relapse-free survival remains favourable even with local invasion [15]. Clinically, PILA has been described as a slowly growing intradermal nodule, displaying violaceous, pink, or bluish-black hues, with lesion diameters ranging from 1 to 40 cm [1].

PILA is noted for its potential local aggressiveness, prompting many authors to advocate wide surgical excision as the primary treatment. A study by Fanburg-Smith et al. documented no recurrences, metastases, or residual tumour during follow-ups [8]. Our patient remained asymptomatic for almost two years post-splenectomy. His previous CT scans were unremarkable, and he presented with four litres of haemoperitoneum.

The diagnosis and treatment approach for PILA remain unclear due to the limited number of reported cases. While wide surgical excision offers a favourable prognosis, awareness of the tumour's metastatic potential is essential. With an increasing number of published PILA cases, a more standardised approach to diagnosis, treatment, and follow-up protocols may eventually emerge. Currently, there is no consensus on a standard follow-up protocol after surgical treatment.

Conclusions

PILA is an extremely rare lymphovascular tumour, with fewer than 40 reported cases, and its occurrence in the mesentery of the colon is documented here for the first time. Its rarity and unusual presentation in visceral sites complicate diagnosis. The use of specific markers like D2-40 may be useful. While wide surgical excision remains the primary treatment, the lack of standardised protocols highlights the need for further research and awareness.

Acknowledgement

Not applicable

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

Funding

Not applicable

Conflict of Interests

Authors declare there are no conflict of interest.

Informed consent

Written consent for publication was obtained from the patient.

Ethical statement

The report was approved by institutional review board and

Data availability

Data will be made available at a reasonable request

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