two case series by Cserni *et al.* and Singh *et al.* describing eight and four cases, respectively.^{1–9} Overall, all patients were female, the median age was 64 years and most patients presented with a palpable mass and/or mammographic abnormality.² All cases contained a mucinous and non-mucinous component with five also including focal areas of classical lobular neoplasia.² The majority were grade 2. These features are consistent with the findings in this case.² There were varying numbers of signet ring cells observed, consistent with intracellular mucin formation. Similarly to our case, E-cadherin was negative and cytoplasmic p120 was positive in nine cases, both of which are recommended as additional immunohistochemistry stains that help classify these unusual tumours,^{1,2} although the diagnosis of lobular carcinoma is primarily based on the morphology.

In all cases with available data, ER was positive, PR was positive in 10 (50%) and negative in four (20%) cases, and HER2 was over-expressed in two (10%), negative in 11 (55%), and was scored 1+ in one (5%) and 2+ in two cases (10%).² None of these cases were HER2 amplified. Ki-67 proliferation index expression ranged from 7 to 40%.²

It is rare for any involved lymph nodes to have mucinous ILC, with only one observed by Cserni *et al.*¹ This is in keeping with the current case, where the positive lymph nodes did not have mucinous morphology.

We present here the first reported case in Australia with these atypical features to further contribute to the limited available evidence on this variation of ILC. While cytology was not undertaken in this case, it is prudent to keep this rare mucinous subtype of ILC as a possible differential diagnosis for mucinous carcinoma. At this stage the clinical significance of this rare variant of ILC is uncertain but may become clear in the fullness of time.

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Giant porta hepatis schwannoma: a rare cause of a cystic hepatic neoplasm

Sir,

Porta in Latin translates to 'city gate' or 'entrance', porta hepatis, therefore the gateway to the liver. The hepatic artery, the portal vein, bile duct, lymphatics and autonomic nerves within the hepatoduodenal ligament potentially give rise to a variety of lesions. Pre-operative tissue diagnosis is difficult due to the high risk of complications including damage to adjacent structures. A 14 cm cystic porta hepatis schwannoma with 'ancient change' is presented. This represents a rare cause of a porta hepatis lesion with only 28 cases identified in the literature to date. To our knowledge it is the largest lesion so far described and first Australian case reported.

A 37-year-old woman of Asian ethnicity presented with intermittent right upper quadrant pain and fullness, and a palpable mass. She was otherwise well with no other relevant medical history. Magnetic resonance imaging showed a 140 mm heterogeneous, multiseptated cystic lesion reported to be consistent with a biliary cystadenoma or cystadenocarcinoma originating from the caudate lobe and pushing into the porta hepatis. No pre-operative tissue diagnosis was attempted. The lesion was removed successfully via an open approach with a left hemihepatectomy and hepatic artery reconstruction.

Grossly the lesion was found to be centred in the porta hepatis and adherent to but not involving the liver. It was 140×130×80 mm, thin-walled (2–5 mm) and multiloculated with thin fibrous septations and degenerate material within. There were no solid or calcified areas. The adjacent hepatic parenchyma appeared normal. A representative radiological image and macroscopic photograph of the cystic structure is shown in Fig. 1.

At microscopy, the cyst wall was predominantly fibrous with a spindle cell proliferation of variable thickness with central cystic degeneration, focal hyaline and myxoid change with mild to moderate nuclear variation favoured as 'ancient change'. No epithelial lining was identified on extensive sampling. There was no necrosis, glandular components or heterologous elements seen. There was moderate inflammation including lymphocytes and plasma cells and occasional

Fig. 1 Representative radiological (MRI) and macroscopic images of the multiloculated cystic lesion with adjacent liver.

lymphoid aggregates with germinal centre formation at the periphery and some hyalinised blood vessels. The adjacent liver parenchyma was histologically normal. Immunohistochemically, the spindle cells showed diffuse and strong staining for S100 and SOX10. The tumour showed diffuse strong staining for CD34. The spindle cells were negative for cytokeratin (CK) AE1/3, CK 7, HMB45, Melan A, oestrogen receptor (ER), progesterone receptor (PR), inhibin, ERG, smooth muscle actin, desmin, CD117 and DOG1. Representative H&E and immunohistochemistry are shown in [Fig. 2](#). A diagnosis of a porta hepatis cystic schwannoma with 'ancient change' was made.

Due to the complex anatomy in the porta hepatis, a wide variety of pathological conditions can occur. A review by Tirumani *et al.* discusses the spectrum of disease within the porta hepatis by first dividing the causes into vascular and non-vascular.¹ Non-vascular causes include those originating from the biliary tree, lymph nodes and lymphatics,

connective tissue and nerves. Neurogenic tumours include neurofibroma, schwannoma and malignant peripheral nerve sheath tumour, and may be an expression of syndromes such as neurofibromatosis type 1. It is difficult to ascertain the true prevalence of these different tumour types within the porta hepatis, as primary porta hepatis tumours are very rare with only limited case reports and small case series in the literature. Furthermore, these need to be differentiated from cystic tumours originating from the intrahepatic bile ducts, which can secondarily expand to fill this anatomical region.

A literature review using PubMed was performed to identify similar cases using search terms ('porta hepatis', 'porta hepatic', 'hilar' or 'hepatoduodenal ligament') and ('schwannoma' or 'neurilemmoma'). The reference list of each suitable paper was also reviewed for additional cases.

Twenty-eight cases were identified and summarised in [Table 1](#) including a review of 15 cases.² Cases ranged in age from 22 to 80 years (mean 49 years) with a slight female

Fig. 2 Representative H&E stained low and high power sections of the cystic structure with a variable thickness wall and spindle cell proliferation. The spindle cell component shows features of areas of inflammation, enlarged and atypical cells, and hyalinised vessels. Representative immunohistochemistry shows cells with diffuse and strong staining for S100.

Table 1 Cases identified by literature review of porta hepatitis schwannoma

Case	Publication date	Origin	Age	Sex	Largest dimension, cm	Solid/cystic	Site	Additional notes of interest
1	2020	China	58	F	4.5	Solid	HDL	Two lesions with the other 2 cm
2	2020	Japan	38	F	3	Mixed	HDL	
3	2020	China	64	F	4.9	Mixed	PH	
4	2020	China	70	M	5.5	Solid	HDL	Multiple tumours, the largest within the porta hepatitis, also 9 cm within liver and 5 cm within gallbladder
5	2019	France	67	F	6	Mixed	PH	
6	2018	Japan	80	M	7	Cystic	PH	
7	2017	Poland	22	F	3.5	NA	PH	
8	2016	China	43	M	8.5	Mixed	HDL	
9	2016	China	31	F	11	Solid	PH	
10	2016	China	50	M	4.5	Solid	HDL	Cellular schwannoma
11	2016	China	43	M	8.3	Mixed	HDL	
12	2015	Not identified	69	F	4.7	NA	HDL	
13	2015	Brazil	30	F	3.8	Solid	PH	
14	2012	China	74	F	2.5	NA	PH	
15	2012	China	62	M	3.5	NA	PH	
16	2011	China	57	F	3.5	NA	PH	
17	2011	China	45	F	7.5	NA	PH	
18	2011	Portugal	29	M	3.7	Mixed	HDL	
19	2011	United States	54	F	5.2	NA	PH	
20	2010	China	38	F	4	NA	PH	
21	2009	India	38	M	4.5	Mixed	PH	
22	2009	China	42	F	2.2	NA	HDL	
23	2006	China	45	M	7	NA	PH	
24	2006	Korea	53	F	4.5	Cystic	PH	
25	2001	United States	37	M	5	Mixed	PH	
26	1996	China	41	M	7.2	NA	HDL	
27	1995	China	33	F	4.5	NA	PH	
28	1993	Japan	62	F	6.2	NA	HDL	

HDL, hepatoduodenal ligament; NA, not available; PH porta hepatitis.

preponderance (61%). Tumours ranged from 2.2 cm to 11 cm (mean 5.2 cm). Five were solid tumours, eight mixed and three cystic. In two cases there was more than one tumour; one had two tumours within the porta hepatitis and one had multiple tumours (exact number not available) within porta hepatitis, liver and gallbladder. Sixteen cases originated in China, a further four were from other parts of Asia (Japan and Korea) and the remainder were from the United States, South America and Europe. This suggests a possible higher incidence in Asian versus Western countries, also seen within other hepatobiliary pathology.³

Pre-operative radiological diagnosis was generally guarded but included cystic hepatocellular carcinoma, metastatic melanoma, hilar cholangiocarcinoma, liver abscess, giant lymph node hyperplasia, and focal nodular hyperplasia. Pre-operative tissue diagnosis was attempted in five cases with the only one successfully completed via endoscopic ultrasound guidance. The other attempts were either non-diagnostic, incorrect (focal nodular hyperplasia or stromal tumour) or showing limited results (spindle cell neoplasia). Intraoperative frozen section was performed in one case, which suggested gastrointestinal stromal tumour.

The pathological findings were remarkably consistent where detailed, with all cases showing features of a spindle cell lesion and diffuse strong S100 positivity. All cases showed typical schwannoma except one that was a cellular schwannoma. Alternative pathology (gastrointestinal stromal tumour, mucinous cystic neoplasm, carcinoma, melanoma, angiosarcoma, leiomyoma/sarcoma, solitary fibrous tumour)

was ruled out with immunohistochemistry showing negative staining for cytokeratins, melanocytic markers (HMB45 and Melan A), smooth muscle markers (SMA and desmin), hormone receptors (ER and PR), STAT6, vascular markers (ERG and CD31) and stromal markers (CD117 and DOG1). No cases have shown recurrence.

In conclusion, the diagnosis of a porta hepatitis cystic lesion is likely to be made only once the definitive resection specimen is received in the laboratory due to the broad range of possible pathologies within the anatomic region, the difficulty obtaining a diagnostic preoperative sample at this location and of a cystic lesion, and the high chance of mimicking other clinical diagnoses. Schwannoma should be considered in the diagnosis of solid and cystic lesions of the porta hepatitis. Appropriate morphologic features and immunohistochemistry can ensure the correct diagnosis and appropriate treatment. Complete excision predicts an excellent prognosis.

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