Goblet cell carcinoid of the appendix

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Three distinct histologic patterns have been identified in appendiceal carcinoids: the argentaffin-positive carcinoid, the nonargentaffin carcinoid, and the mucinous, goblet cell carcinoid.[1] Therapy for carcinoids in the first 2 classes traditionally depends on tumour size and location with respect to the base of the appendix. The third type is rarer and behaves more aggressively,[1,2] hence the literature suggests that management should be more aggressive. We discuss, through the use of a case report, the management of goblet cell carcinoid of the appendix.

Case report

A 39-year-old woman underwent an appendectomy at a regional hospital. Pathological examination of the specimen revealed a goblet cell carcinoid of the appendix, so the patient was sent to our institution for an opinion regarding management. Our pathologist confirmed a goblet cell carcinoid and estimated its size as approximately 12 mm. There was evidence of perforation, and the appendix itself was not inflamed. The original pathology report also suggested a perforation. Some signet ring cells were also seen. After discussion of management options with the patient, a right hemicolectomy was decided on. The procedure was uncomplicated. The final pathology report indicated residual goblet cell carcinoid tumour in the appendiceal stump.

Discussion

Histologically, goblet cell carcinoids can readily be distinguished from typical carcinoids by immunostaining (Fig. 1). This is important since goblet cell carcinoids of the appendix are said to have a clinical behaviour intermediate between typical carcinoids and adenocarcinoma;[3] hence, surgical management is not straightforward.

Features said to confer a worse prognosis and for which some have advocated hemicolectomy include increased mitotic rate and cellular atypia,[4] and "predominance of carcinomatous growth." Various patterns corresponding to this carcinomatous growth have been described, including "mixed carcinoid adenocarcinoma" if there is more than 50% carcinomatous growth, and pure goblet cell carcinoid if there is less than 25% carcinomatous growth. The degree of carcinomatous growth has been shown to correlate with extension into adjacent organs and poor prognosis.[5] In our case, the amount of carcinomatous growth was estimated to be less than 10%, so this histologic finding alone was not enough to advocate surgery. The usual information required to make a decision to operate with respect to a typical carcinoid, including size of tumour and proximity to base, could not be determined from the pathological findings available to us. Even if we had this information, it is unclear whether this is relevant for goblet cell carcinoids. The decision to operate was based on the fact that this patient was an otherwise healthy 39-year-old woman with a variant of carcinoid

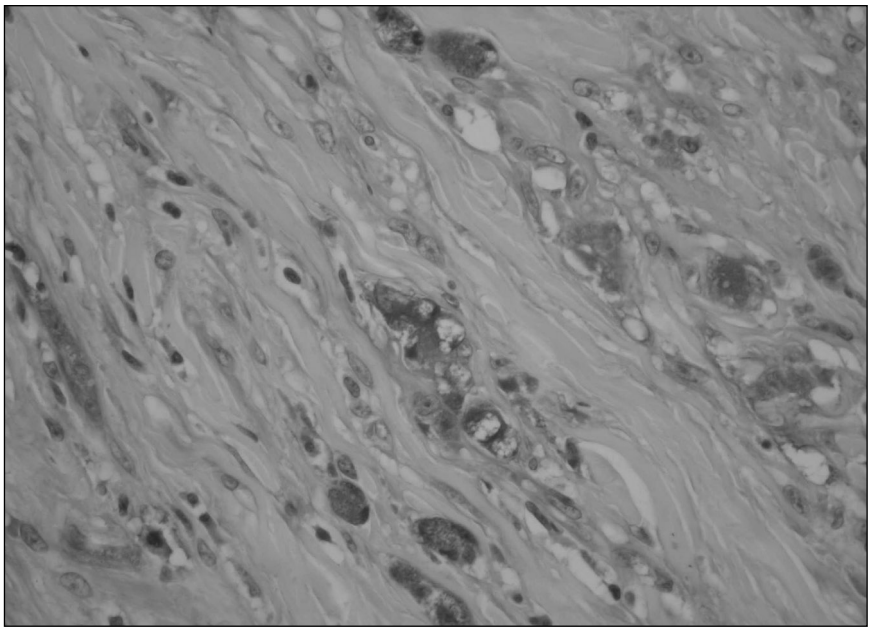

FIG. 1. Goblet cell carcinoid (mucicarmine stain; original magnification ×400).

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halfway between typical carcinoid and adenocarcinoma for which more aggressive management is advocated than for typical carcinoid.[2] Also, the pathological findings suggested that there had been a perforation but the appendix was not inflamed, so we could infer that the tumour itself had perforated.

Goblet cell carcinoid is a variant of typical carcinoid, but because its clinical behaviour is part way between typical carcinoid and adenocarcinoma the criteria normally applied to determine if hemicolectomy is necessary may not be applicable. Several pathological features have been proposed as suggesting increased aggression in these tumours and may be helpful in determining the indication for resection.

Competing interests: None declared.

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Leiomyosarcoma of the inferior vena cava

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Primary malignant tumours of vascular origin are rare. The most common is the leiomyosarcoma, a malignant neoplasm derived from smooth-muscle tissue and accounting for less than 1% of all malignant tumours.[1] Leiomyosarcomas are seen principally in adults, with the majority of patients presenting in the fifth and sixth decades. We describe a case of a leiomyosarcoma involving the inferior vena cava (IVC) with metastases to the liver.

Case report

A 56-year-old woman with long-standing hypertension and type 2 diabetes presented with vague epigastric discomfort, decreased appetite and a 12-kg weight loss over the previous 3–4 months. On examination, her abdomen was mildly tender over the epigastrium. The liver edge was palpable with minimal ascites. Enhanced CT of the abdomen demonstrated a mixed density mass arising from the IVC (Fig. 1), extending superiorly into the right atrium and inferiorly to near the upper pole of the right kidney. Transesophageal echocardiography confirmed a tumour extending into the right atrium with no obvious margin between the mass and the IVC wall. There was also a large metastatic lesion straddling the the medial and lateral segments of the left hepatic lobe (Fig. 2). The patient underwent transfemoral percutaneous biopsy of the mass, which revealed a smooth-muscle tumour. Biopsy of one of the liver lesions confirmed the diagnosis of leiomyosarcoma metastatic to the liver.

Discussion

Perl first described leiomyosarcoma of the IVC in 1871.[2] Since then, just over 200 cases have been reported in the literature. The location of the tumour in the IVC is important because it determines the symptoms and surgical resectability. Most tumours arise in the lower (44.2%) or middle (50.8%) portion, with only a small number (4.2%) arising from the upper third or suprahepatic region. IVC lesions arising below the renal veins cause pain in the right lower quadrant, back and flank, and lower leg edema. Tumours of the lower region are often amenable to surgical excision. Interruption or ligation of the IVC below the renal veins is well tolerated by most patients.[3] Tumours of the middle caval segment cause right upper quadrant pain and tenderness and sometimes renovascular hypertension. Extensive collateral venous drainage of the left kidney preserves renal function during resection of middle caval tumours. Patients with upper segment or suprahepatic tumours usually suffer from Budd–Chiari syndrome with hepatomegaly, jaundice and massive ascites. Upper caval leiomyosarcomas are the least amenable to complete removal.[4] Leiomyosarcomas of the vascular system are thought to be relatively slow growing, spreading mainly by extension into adjacent tissue planes. Metastasis has been reported in fewer

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