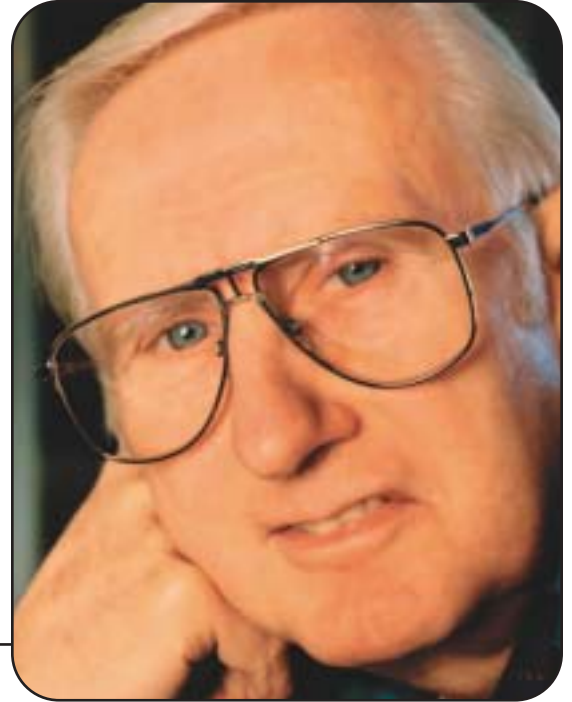


Vestigial Remnant, but Not Innocent

By Mira Parai, MD, FRCP; Nicholas G. Escott, MD, FRCP; Joseph F. Wasielewski, MD, FRCP; and Allan Kirk, MD, FRCP



A 60-year-old man presented with a large swelling of the anterior abdomen. The lesion was protruding with overlying skin. No skin ulcer was noted. Other history notes stated that the patient had smoked for a number of years, but had quit 10 years ago.

He was on levothyroxine sodium and his right thyroid lobe was enlarged. Otherwise, he was healthy. A specific investigation of the anterior abdominal tumour was done.

A computed tomography (CT) scan showed a cystic lesion, approximately 28 cm in diameter (Figure 1). The lesion was surgically removed in August 2000. The mass was an oval cystic tumour weighing 3,600 grams and measuring 27 cm by 17 cm by 18 cm. The outer surface was smooth and an edematous pedicle of soft tissue was attached, which was 3.5 cm long and had a diameter of 8 cm. The cyst was unilocular and filled



Figure 1: A CT scan showing cystic lesion (no connection with the liver).

with thick, mucoid to necrotic material. Small cystic spaces were seen in the wall. The wall thickness was 1 cm. A small portion of solid tissue, about 6 cm by 4 cm by 2 cm, was seen in between the large cyst and the pedicle. Tenacious mucus was seen adherent to the inner surface of the cyst.

In the post-operative period, the patient developed a ventral hernia that was repaired along with the removal of omental tissue in March 2001. The patient tolerated surgery well and was discharged with a return visit scheduled.

WHAT'S YOUR DIAGNOSIS?

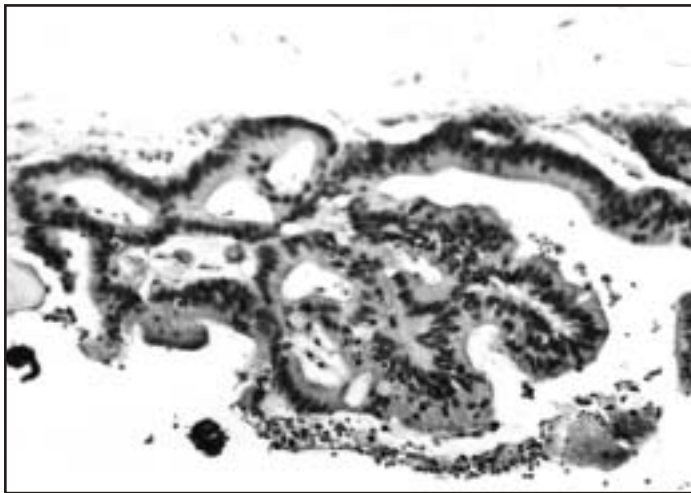


Figure 2: Low magnification, microscopic features. The lining cells are tall columnar and most areas are single-layered.

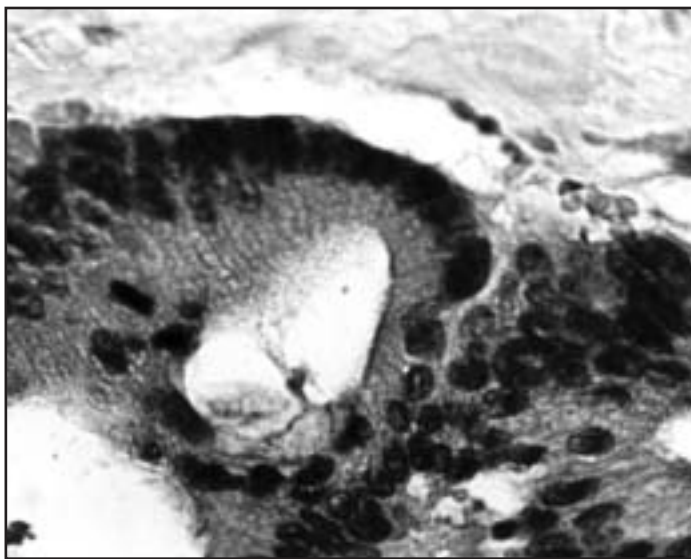


Figure 3: High magnification, showing focus of adenocarcinoma.

What's your diagnosis?

Answer

Adenocarcinoma of urachal cyst. The microscopic examination of multiple sections from the cyst, pedicle and the omental tissue show well-differentiated mucinous adenocarcinoma. The large cyst wall is lined by a tall, columnar, colonic type of epithelium, which has a benign appearance in most areas (Figure 2).

Occasional foci show malignant features, including epithelial tufting, nuclear enlargement and mitotic activity (Figure 3). Numerous small cystic spaces showed encysted mucin.

Discussion

In embryonic life, the urachus extend from the dome of the bladder to the umbilicus. The urachus is a derivative of the urogenital compartment of the cloaca.

Complete persistence of the urachus results in a vesico-umbilical fistula with urinary discharge. Urachal sinuses result from either umbilical or vesicular ends of the urachus.

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WHAT'S YOUR DIAGNOSIS?

In congenital urethral atresia, the urachus may dilate enormously, even to the extent of causing dystonia of the bladder. More common, but still rare, are urachal cysts, which result from focal persistence of the urachus. The cyst may become infected and be a suprapubic mass that depresses the dome of the bladder. Rarely, the cyst may rupture and produce peritonitis.

Carcinoma complicating a urachal cyst has been reported, but is still rare.^{1,2,3} In our case, there was no connection with the bladder wall, discarding the possibility of extension of a bladder tumour. Our case shows the major portion of the cyst wall is lined by low to tall columnar epithelium, a finding compatible with a urachal cyst (Figure 1).

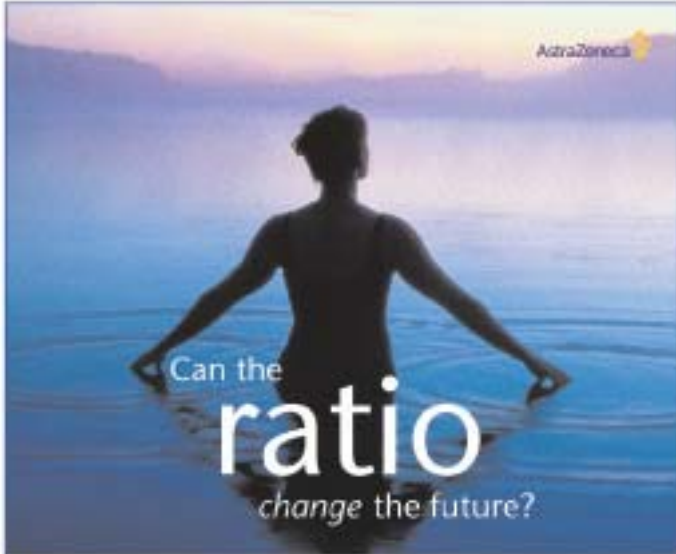

Carcinoma in a urachal cyst is rare. The reason could be that most cysts get infected and are resected when the patients are younger. Our case is interesting because of the cyst's giant size. The largest urachal cyst with adenocarcinoma reported weighed 3,800 grams and measured 22 cm by 20 cm by 20 cm.⁴

If adenocarcinoma occurs in the vesical extreme of the urachal cyst, this may present as adenocarcinoma of the urinary bladder. Total surgical removal and subsequent management assured this patient a healthy life for 2.5 years after surgery. 

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