

carcinoma or its necrotic products (Chapter 4). On a sad note, Armand Trousseau (1801-1867, physician at Hotel Dieu, Paris), for whom this sign is named, correctly suspected that he had carcinoma when he developed spontaneously appearing and disappearing (migratory) thromboses.

The course of pancreatic carcinoma is typically brief and progressive. Despite the tendency of lesions of the head of the pancreas to obstruct the biliary system, fewer than 20% of pancreatic cancers overall are resectable at the time of diagnosis. Most have invaded vessels and other structures that cannot be removed surgically, or have metastasized to distant organs. There has long been a search for tests that could be useful in the early detection of pancreatic cancer. Serum levels of several antigens (e.g., carcinoembryonic antigen and CA19-9 antigen) are often elevated in individuals with pancreatic cancer. These markers, while useful in following an individual patient's response to treatment, are relatively nonspecific and also lack the sensitivity needed to be used as tests to screen the wider population. Several imaging techniques, such as endoscopic ultrasonography and computed tomography, have proved of great value in establishing the diagnosis once it is suspected, but are also not useful as screening tests.

## KEY CONCEPTS

- Cigarette smoking is the leading preventable cause of pancreatic cancer.
- Pancreatic cancer is one of the most aggressive of the solid malignancies.
- Many invasive pancreatic cancer arises from histologically well-defined precursor lesions called *pancreatic intraepithelial neoplasia* (PanIN).
- Ductal adenocarcinomas elicit an intense desmoplastic response.
- The genes most frequently mutated or otherwise altered in pancreatic cancer include *KRAS*, *p16/CDKN2A*, *TP53*, and *SMAD4*
- Clinically, most patients present with abdominal pain and weight loss, sometimes accompanied by jaundice and deep vein thrombosis, and succumb to the disease within 1 to 2 years.

## Acinar Cell Carcinoma

Like normal acinar cells, acinar cell carcinomas form zymogen granules and produce exocrine enzymes such as trypsin and lipase. Fifteen percent of individuals with acinar cell carcinoma develop the syndrome of metastatic fat necrosis caused by the release of lipase into the circulation.

## Pancreatoblastoma

Pancreatoblastomas are rare neoplasms that occur primarily in children aged 1 to 15 years. They have a distinct microscopic appearance consisting of squamous islands admixed with acinar cells. They are malignant neoplasms, but survival is better with these tumors than it is for pancreatic ductal adenocarcinomas.

## SUGGESTED READINGS

### Congenital Anomalies

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