CASE SNIPPETS

Carcinoma of stomach in a patient with familial tylosis
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The association of tylosis with esophageal cancer has been extensively reported but association with gastric cancer is rare. We report a 55-year-old man with familial tylosis and carcinoma of the stomach for which radical gastrectomy was done. Repeat endoscopy 3 years later is normal. [Indian J Gastroenterol 2002;21:227]

Key words: Gastric cancer

Tylosis, also known as focal non-epidermalytic palmo-plantar keratoderma, is an autosomal dominant disorder of the skin that manifests as thickening of the skin of the palms and soles and is associated with oral lesions. The association of tylosis with carcinoma of the esophagus has been reported. The causative locus has been designated the tylosis esophageal cancer gene. However, tylosis associated with gastric cancer is rare.

A 55-year-old man came with two months' history of epigastric pain. He had anorexia and weight loss but there was no history of vomiting, hematemesis or melena, or any lump in the abdomen. Clinical examination was unremarkable except for hyperkeratosis of the palms (Fig) and soles, which according to the patient was present since early childhood.

Both his deceased elder brothers had tylosis and had succumbed to some abdominal disease, one following hematemesis. The patient's elder son has tylosis since he was 5 years old and the second son developed it at age 15 years.

Blood investigations were normal. Upper gastrointestinal endoscopy revealed normal esophagus and a 0.5 cm to 1 cm mucosal nodularity in the posterior wall of the antrum just proximal to the pylorus; biopsy revealed adenocarcinoma. Ultrasonography and CT scan of the abdomen were normal. Ex-}

plosatory laparotomy with radical subtotal gastrectomy was performed. The patient had an uneventful postoperative course. Histology revealed moderately differentiated adenocarcinoma of the stomach without nodal involvement.

Esophagogastroscopey of the patient and his two sons were normal three years later.

Patients with familial tylosis are prone to develop cancer of the esophagus. Tylosis has also been reported to be associated with bronchogenic carcinoma, breast and ovarian cancer, and other carcinomas such as colonic adenocarcinoma and melanomas.

Our patient had tylosis associated with carcinoma stomach. Two of his brothers had succumbed to an abdominal disease, one with hematemesis. His two sons have been placed under annual endoscopic surveillance, considering all patients with tylosis develop esophageal cancer by the age of 80 years.

References

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Nonfunctioning islet cell tumor presenting with ascites and portal hypertension

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Nonfunctioning islet cell tumors commonly cause no symptoms. A 22-year-old woman presented with lump in the left hypochondrium, refractory high-protein ascites and evidence of left-sided portal hypertension. At exploratory laparotomy, a 30 cm x 15 cm mass was seen at the splenic hilum, with large collateral vessels around. Distal pancreatectomy with splenectomy was done. Histology of the mass showed malignant

![Image: Hyperkeratosis of palms of patient and his elder son](https://via.placeholder.com/150)
islet cell tumor infiltrating the spleen. The patient died in the postoperative period. [Indian J Gastroenterol 2002;21:227-228]

Key words: Pancreas tumor, spleen

Islet cell tumors are rare, with a prevalence of 1:100,000. Nonfunctioning islet cell tumors (NFICT) constitute about 15% of these. The common presentation of NFICT is abdominal pain or jaundice; rarer presentations are with lump, steatorrhea, ascites and gastrointestinal bleeding. Preoperative diagnosis is difficult due to nonsecreting nature of these tumors.

A 22-year-old woman presented with lump in the hypochondrium and progressive distension of the abdomen over 3 years. There was no history of jaundice, alcoholism, hematemesis or melena. On examination, she was averagely built and nourished. Abdominal examination revealed massive ascites that was not tense. An irregular mass was palpated in the left hypochondrium that was extending into the umbilical and lumbar regions; it was minimally mobile in the transverse direction. Renal angle was normal.

Investigations: normal hemogram and liver function tests. Ultrasonography and Doppler study revealed ascites and a complex mass arising in and around the tail of the pancreas that could not be differentiated from the spleen. In addition, multiple venous collaterals were seen in that region; the splenic vein could not be separately visualized. Gastroduodenoscopy revealed presence of esophageal and gastric varices. CT scan showed the mass around and infiltrating into the hilum of the spleen with multiple collateral channels in the vicinity (Fig). Guided fine-needle aspiration biopsy showed features suspicious of stromal tumor. Liver biopsy was normal. Ascites was high-protein in nature and was negative for malignant cells. Bone marrow and splenic aspiration studies were normal.

Exploratory laparotomy revealed 3.5 liters of ascites. A grayish white mass, 30 cm x 15 cm x 15 cm, was seen to be situated around the hilum of the spleen and inseparable from it. Massive collateral vessels (1-2 cm in diameter) were noted around the mass and in the spleno-renal ligament; dilated vessels were also seen on the external surface of the tumor. The pancreatic tail could not be separately seen. The mass was excised with the spleen, amounting to distal pancreatoduodenectomy with splenectomy. The specimen weighed 3.5 Kg. Histology showed features of islet cell tumor infiltrating into the spleen.

The patient developed disseminated intravascular coagulopathy and died on the fourth postoperative day.

A patient is considered to have a nonfunctioning tumor when there is no clinical evidence of hormonal hypersecretion in association with a known APUDoma syndrome. Some of these tumors are thought to secrete a prohormone that is not measured by conventional methods or a hormone in quantities insufficient to produce clinical symptoms, or these patients may have symptoms with no obvious clinical complex.

Unexplained ascites has been documented in 4% of cases with NFICT. Gastrointestinal hemorrhage is usually due to variceal bleed, the cause being left-sided portal hypertension caused by splenic vein compression or thrombosis. Nonfunctioning tumors are usually single and slow growing, and those greater than 5 cm are often malignant.

Surgical excision is the recommended treatment. Prognosis is usually good after excision. Chemotherapy with a combination of streptozotocin and 5-fluorouracil may be effective.

References

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Tail gut cyst

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The tail gut is a blind extension of the hindgut into the tail fold just distal to the cloacal membrane. Rem-

Case Snippets

Fig: CT scan showing homogenous mass (A) in and around hilum of spleen (B) along with peri-splenic and anterior peritoneal collaterals and ascites