Carcinoid tumour of ampulla of Vater associated with viscero-cutaneous neurofibromatosis

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Summary
A case of carcinoid tumour of the ampulla of Vater with viscero-cutaneous neurofibromatosis is reported together with a review of the literature.

KEY WORDS: periampullary carcinoid, neurofibromatosis.

Introduction
Carcinoid tumours arising in the ampulla of Vater are very uncommon (Weldner and Ziter, 1981; Godwin, 1975; Johnson and Weaver, 1981; Barber, 1976). They have been occasionally found to be associated with cutaneous neurofibromatosis. Viscero-cutaneous neurofibromatosis with carcinoid of ampulla of Vater is however, being reported here for the first time. Our patient presented with features suggestive of periampullary carcinoma and was successfully treated with modified Whipple's procedure.

Case report
A 32-year-old male was admitted in July 1981 with an 8-month history of 3 episodes of fever without chills and rigors. From February 1981, he had a low grade fever and fluctuating obstructive jaundice. The patient had noticed multiple nodules on his body for about 20 years. No other member of his family had similar nodules.

On examination, he was a pale, moderately jaundiced young man. There were multiple broad sessile neurofibromata all over his body. The liver was enlarged 3 cm below the costal margin and a non-tender gall bladder was palpable.

Investigations confirmed anaemia and a biochemical pattern of obstructive jaundice. A percutaneous transhepatic-cholangiogram showed dilatation of the intrahepatic ducts and common bile duct (CBD). There was irregularity of the lower end of the common bile duct, though the dye flowed freely into the duodenum. Duodenoscopy showed a mass bulging from the ampullary area.

At laparotomy, a nodule 2–3 cm in size was felt in the region of the ampulla. Duodenotomy showed it to be a pedunculated growth with intact mucosa. There were multiple nodules measuring 3–5 cm in diameter on the ante-mesenteric border of the proximal jejunum. Radical pancreatico-duodenectomy was done.

The postoperative period was uneventful and the patient was discharged on the 8th postoperative day.

Microscopically, the ampullary tumour consisted of tumour cells in trabecular and acinar arrangements separated by their fibrous septae in the region of the submucosa. The tumour cells did not show pleomorphism and only occasional mitotic figures were made out. Special stains revealed these to be argentaffin negative (Mason Fontana stain) and argyrophilic positive (Bodian-stain). Microscopic focus of identical tumour was seen in the sections from the head of the pancreas. The swellings on the serosal surface of the duodenum and jejunum were well encapsulated and involved serosal and muscle coats of the gut with mucosa and submucosa stretched over them. They were made up of spindle shaped cells, present in bundles and running in various directions. The background was fibrillar. No pleomorphism or mitosis was seen. Special stains for muscle and collagen were negative. These were interpreted as neurofibromas.

Discussion
Barber, while reviewing the literature (inclusive of his own) reported 13 cases of ampullary carcinoid up to 1976 and subsequently Johnson et al. (1981) described another case.
The occasional association of cutaneous neurofibromatosis with visceral tumours, such as neurofibromas, meningiomas, gliomas, phaeochromocytomas is well documented (Lee and Garber, 1970). However, its association with carcinoïd of the gastrointestinal tract has been reported in only 3 earlier cases (Johnson and Weaver, 1981; Barber, 1976; Lee and Garber, 1970).

Our patient, in addition, is probably the only reported case of ampullary carcinoïd with cutaneous as well as visceral neurofibromatosis. Lee and Garber in 1970 report an incidental duodenal carcinoïd with viscero-cutaneous neurofibromatosis although found at autopsy.

The origin of carcinoïd tumours is still not established (Foley and Davis, 1965; Beaton et al., 1981). Recently Pearse and Welbourn (1973) have mentioned carcinoïds to arise from cells belonging to the APUD (amine precursor uptake and decarboxylation) cell system. These cells possibly originate from primitive neural crest (Jager and Polk, 1977). The association between neurofibromatosis and ampullary carcinoïd, as reported here, may therefore be because of the same genetic abnormality involving these neural crest cells. The associated visceral neurofibromatosis may also be because of the same basic abnormality.

Depending on their site of origin, carcinoïd cells exhibit differently staining neurosecretory granules. Foregut carcinoïds are either argentaffin positive or non-reactive, hindgut are argentaffin positive and hindgut are either mixed type or non-reactive (Soga and Tazawa, 1971). The present case was argentaffin positive. The case reported by Barber (1976), was argentaffin negative; whether it was argentophile positive or not is not known. In the recently reported case by Johnson and Weaver (1981), details of the staining reaction are not available. Argyrophilic tumours secrete 5-HTP. However, in our patient, no systemic manifestations to suggest hormone-based malignant carcinoïd syndrome were seen. Ampullary carcinoïd with elevated hydroxy-indole secretion and carcinoïd syndrome has not been reported.

Ampullary carcinoïds present early owing to their strategic location. Except in 3, features of obstructive jaundice were noticed in all the reported cases (Johnson and Weaver, 1981; Barber, 1976; Lee and Garber, 1970). Symptoms of pancreatic duct obstruction are usually absent. No case has been diagnosed pre-operatively and the diagnosis of periampullary carcinoma is usually made.

Although the number of cases is limited, these tumours in general run a fairly benign course. More than 75% remained well for periods varying from 1-7 years. Local resection with or without pancreateo-duodenectomy has been the treatment of choice. Our patient is doing well after radical pancreateo-duodenectomy and has gained weight. The efficacy of chemotherapy and radiotherapy in carcinoïd tumours is not yet proved (Conti et al., 1981).

References


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