

Title:

Carcinoid syndrome and somatostatin analogues

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Carcinoid syndrome and somatostatin analogues

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Dear Editor,

Carcinoid syndrome (CS) is a paraneoplastic syndrome associated with serotonin secretion, which undergoes renal excretion as 5-hydroxyindoleacetic acid in the urine (u5-HIAA). The goal of this retrospective, observational study was to review a personal series of CS cases by performing a search of the clinical and ultrasound records of patients with metastatic liver disease.

Fourteen patients with CS were identified over a period of 28 years (Table 1). The mean age was 58 years (range, 40-80 years), 64 % were female and 36 % were male, all of them Caucasians. Given the search method used, all patients had liver metastases and the primary neuroendocrine tumor was located in the bowel (50 %), in the gastroduodenal area, in an unknown site (14 %) and in the mesentery, pancreas and lung in 7 % of cases. The tumors were G1-G3 and their clinical manifestation was CS, with flushing and chronic diarrhea. Flushing developed in 86 % of cases, diarrhea in 78 % and heart disease in 21 % of cases. One case included steatorrhea and one case included bronchoconstriction (7 %).

Of all 14 patients, ten received treatment with somatostatin analogues (octreotide and lanreotide) in standard doses via the subcutaneous or intramuscular route (1), which reduced symptoms in 70 % and u5-HIAA in 50 % of cases. Liver tumors were reduced in one case (7 %) and primary tumors in the ileum were reduced in 33 % of cases. The main side effect of treatment was development of bladder lithiasis.

The incidence of CS is 0.27/100,000 in the United States (2), occurring in 10-25 % of neuroendocrine tumors (2-4), primarily in the digestive system (70 %) followed by the respiratory system (25 %). Age at presentation was from 55 to 60 years (3). In our series, the mean age was 58 years.

Flushing develops in 85 % (3), diarrhea in 80 % (3,4), heart disease in 60 % (other reviews find 20-40 %) (4), and bronchospasm in 10-20 % of cases. Liver metastases are present in 87-100 % of cases (4). Treatment with somatostatin analogues reduces symptoms in 50-80 %, and u5-HIAA in 45-50 % of cases (3,5), which is similar to our series. Other treatments include chemotherapy, interferon, everolimus and loperamide for diarrhea.

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Table 1. Casuistry

<i>Age/sex, size</i>	<i>Location</i>	<i>Procedure</i>		<i>LM</i>	<i>CS/Treatment</i>	<i>Symptoms</i>		<i>Response</i>
53F 20 mm	Duodenum	Biopsy	NET G3	LM	CS	Flash	Diarrhea	
65F 20 mm	Intestine	Surgery	NET	LM and PM?	CS: octreotide	Flash	Diarrhea	Yes
64F	Unknown		"	LM	CS	Flash	Diarrhea	
80F	Appendix?	Surgery	"	LM	CS: octreotide	Flash		Yes
70F 30 mm	Intestine	Surgery	"	LM	"	Flash	Diarrhea	No
45F 22 mm	Intestine	Surgery	"	LM	"		Diarrhea	Yes
56M 25 mm	Pancreas	Surgery	"	LM	CS: multiple	Flash?	Steatorrhea	Yes
47M	Mesentery	"	NET G1	LM	CS: lanreotide	Flash	Diarrhea	Yes
73M 40 mm	Ileum	Surgery	"	LM	CS	Flash	Diarrhea	
40M 90 mm	Ileum	Bypass	NET G2	LM	CS: octreotide	Flash	Diarrhea	Yes
45M 50 mm	Retroperitoneum	Biopsies	NET G2	LM	CS: multiple	Flash	Diarrhea	No
63F	Lung	Surgery	NET G2	LM	CS	Flash	Diarrhea	
65F	Unknown		"	LM	CS: octreotide	Flash		No
50F 20 mm	Ileum	Surgery	NET G1	LM	CS: octreotide	Flash	Diarrhea	Yes

NET: neuroendocrine tumor; LM: liver metastasis; PM: pulmonary metastasis; CS: carcinoid syndrome.