The role of immunohistochemical assessment of somatostatin receptor expression in case of patients with well differentiated neuroendocrine neoplasms with symptoms of carcinoid syndrome and negative somatostatin receptor imaging.

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INTRODUCTION:

Overexpression of somatostatin receptors (SSTR) is the characteristic feature of well differentiated neuroendocrine neoplasms (NENs). We present two patients with well differentiated NENs of the intestine with negative somatostatin receptor imaging (SRI) and without SSTR expression confirmed with immunohistochemical (IHCH) examination and symptoms of carcinoid syndrome with good clinical response to the therapy with long acting somatostatin analogs.

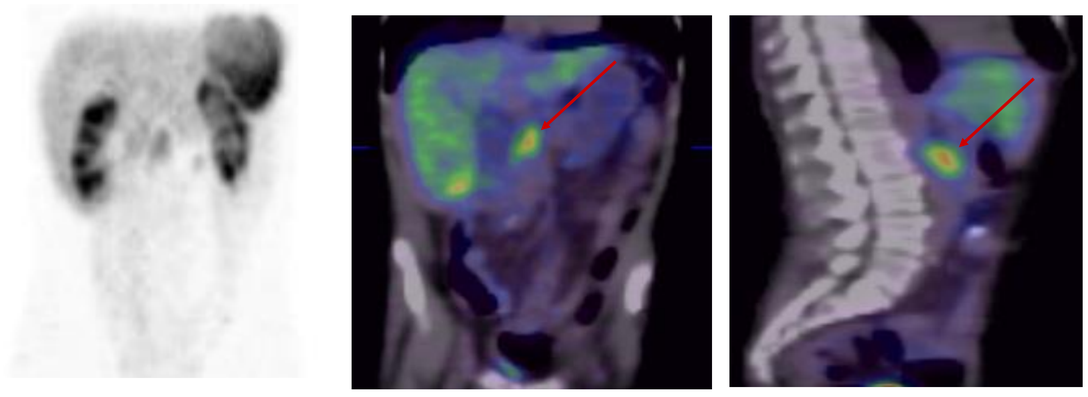
CASE REPORTS:

Patient 1

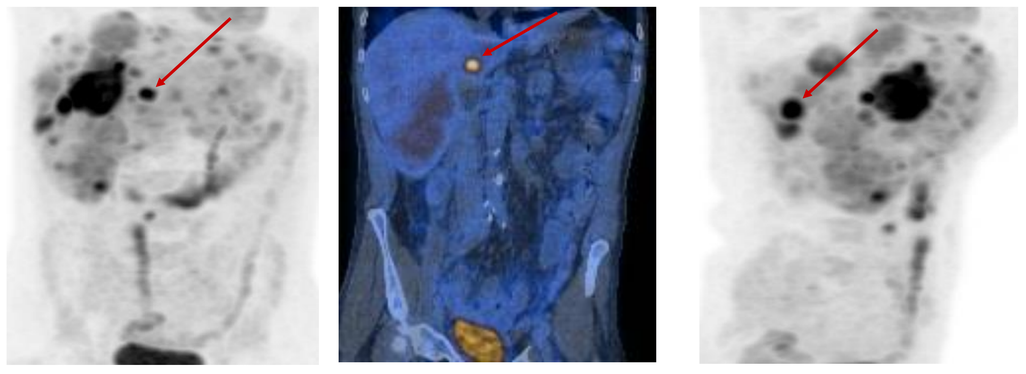
53-year old man after right hemicolectomy due to the tumor (NETG2; Ki67 5%) of the ileo-cecal valve with mesenteral lymph nodes and liver metastases. Patient presented with flushes. Therapy with long-acting somatostatin analog led to release of symptoms. Laboratory findings prior to the therapy: 5OHIAA 545.7 umol/24h (N: 10-40) and CgA 112 ng/ml (N: 0-6). CgA level measured during therapy with long-acting somatostatin analog was 90 ng/ml. While somatostatin receptor scintigraphy (SRS) revealed increased pathological uptake only in the single mesenteral lymph node immunohistochemical (IHCH) examination was performed. IHCH examination confirmed only low focal SSTR (type 1-5) expression.

Patient 2

64-year old man with inoperable tumor (NETG1, Ki67<2%) of the cecum with lymph nodes and liver metastases. Patient also presented with severe flushes and diarrhea. Therapy with long-acting somatostatin analog led to release of symptoms. Laboratory findings prior to the therapy: 5OHIAA 287 umol/24h and CgA 248 ng/ml. CgA level measured during therapy with long-acting somatostatin analog was 102.9 ng/ml. While SRS did not reveal any pathological uptake of the tracer in the lesions visualized with CT IHCH examination was performed. IHCH did not reveal any SSTR expression.



Figures 1. ^{99m}Tc-HYNIC-[D-Phe1,Tyr3-Oktreotyd] scintigraphy (SPECT/CT) in 53-year old man – increased uptake of the tracer in the single mesenteral lymph node and unhomogenous uptake in the liver with lesions of decreased uptake of the tracer.



Figures 2. ^{18}F-FDG-PET in 53-year old man – increased glucose metabolism in the single mesenteral lymph node and unhomogenous metabolism in the liver with lesions of increased glucose metabolism.

CONCLUSIONS

We would like to emphasize the observed release of symptoms of carcinoid syndrome in response to the therapy with long acting somatostatin analogs in case of patients with negative SRS and without SSTR expression in immunohistochemical examination. Therefore therapy with long actiong SSA should be considered also in case of patients with disseminated neuroendocrine neoplasms and symptoms of carcinoid syndrome without somatostatin receptor expression in the neoplastic lesions.

Presented case reports indicate the necessity of immunohistochemical assessment of the expression of SSTR in case of patients with well differentiated NENs and negative SRS results, because confirmed lack of SSTR expression leads to the change of the imaging modalities used for patients' management.

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