Dilated cardiomyopathy, a rare and reversible manifestation of the glucagonoma syndrome

V. Antonopoulou*[a], A. Diamantopoulos[a], A. Beka[a], T. Stratigou[a], G. Kyriakopoulos[b], I. Landrou[c], P. Prigouris[c], S. Tsagarakis[a], DA. Vassiliadi[a]

[a]Department of Endocrinology, Diabetes and Metabolism, National Expertise Center for Rare Endocrine Disorders and member of the Endo-ERN, "Evangelismos" General Hospital of Athens, Athens, Greece, [b] Pathology Department, "Evangelismos" General Hospital of Athens, Athens, Greece,
[c] 4th Surgical Department, "Evangelismos" General Hospital of Athens, Athens, Greece

Introduction

Glucagonoma is a rare functional pancreatic neuroendocrine tumor occasionally associated with a characteristic syndrome due to glucagon hypersecretion.

Glucagonoma-associated dilated cardiomyopathy has been reported in only a few case reports.

Observations

A 69-years-old female presented with glucagonoma syndrome: weight loss, diarrhea, nausea, depression, stomatitis, normocytic anemia, recent onset diabetes and necrolytic migratory erythema *(Figure 1A)*

She also had heart failure due to dilated cardiomyopathy *(Table 1)* without evidence of ischemia on coronary angiography.

Biochemically, glucagon levels (2888 pg/ml), chromogranin A (56nmol/l) and pancreatic polypeptide (PP) (474pmol/l) were markedly increased *(Table 2)*.

Imaging revealed a 4.1 cm pancreatic mass with significant uptake on ^{68}Ga-Dotatoc-PET/CT (SUVmax:62) and in a local lymph node (SUVmax:36) *(Figure 2).*

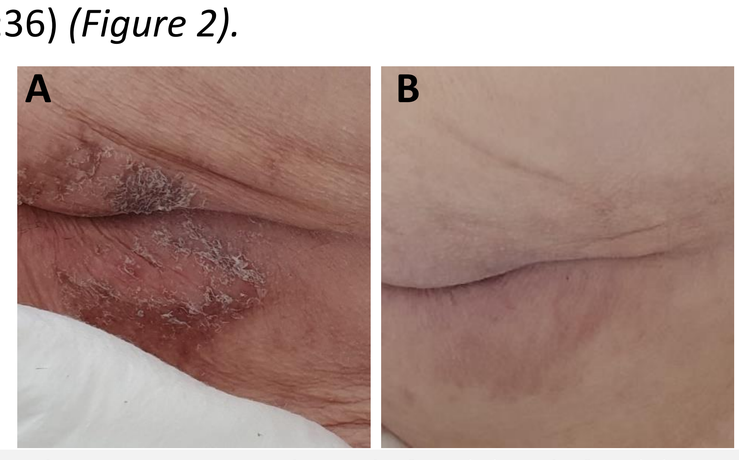

Figure 1 . Necrolytic migratory erythema before (A) and after (B) sc octreotide

She was initially started on sc octreotide resulting in disappearance of the necrolytic migratory erythema *(Figure 1B)* but only partial clinical and biochemical response.

Subsequently she underwent distal partial pancreatectomy, splenectomy, and cholecystectomy.

Histopathology demonstrated PanNET G2, ki67:12%, Glucagon(+), ChromograninA (+), Synaptophysin(+), PP(+), SSTR2a(+++) and metastasis in 3/22 local lymph nodes, pT3N1.

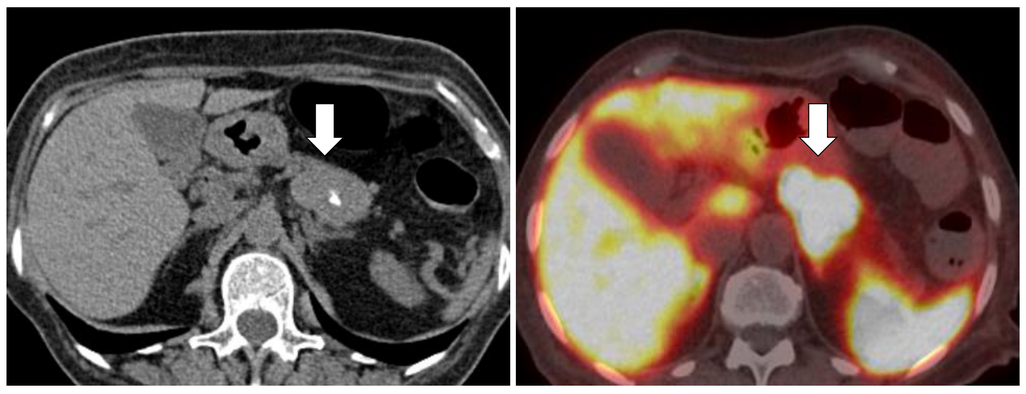

Figure 2 . CT scan (A) and 68Ga-Dotatoc-PET/CT (B) showing (arrow) a 4.1cm pancreatic mass with high SUV

Parameters	Baseline	14-days P.O.	2-months P.O.	Reference Range
LVEF, %	**25**	**30**	**45**	55-70
LVEDD (cm)	**5.9**	**5.8**	4.9	3.6-5.6
NT-proBNP (pg/ml)	**16646**	**9444**	**1448**	<900

Table 1. Cardiac function parameters at baseline and postoperatively

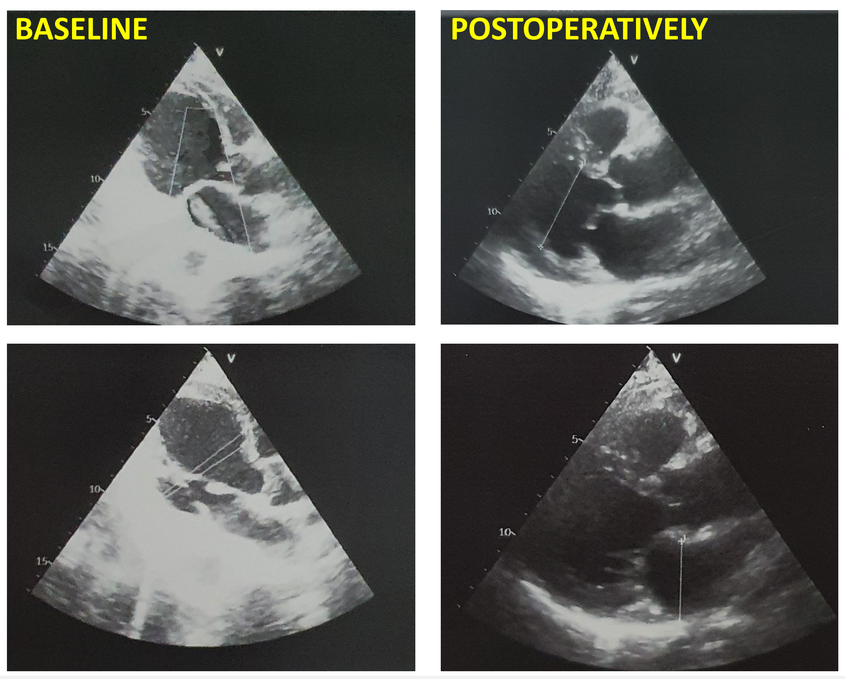

Fig1. Echocardiographic features of dilated cardiomyopathy before and 2 months after surgery

Fifteen days after surgery, glucagon levels returned to normal, LVEF elevated to 30% and NT-proBNP reduced.

Two months post-surgery, further increase of LVEF was observed, left ventricular end diastolic dimension decreased and NT-proBNP further dropped.

Her general condition improved, she gained weight and improved glycemic control whereas nausea and diarrhea resolved. Glucagon, chromogranin A and PP levels returned to normal.

Parameters	Diagnosis	sc octreotide	postoperatively 14-days	postoperatively 2-months	*Normal values*
Glucagon pg/ml	**2888**	**733**	216	321	*<250*
Pancreatic Polypeptide (pmol/l)	**474**	93	21	18	*<100*
Chromogranin A (nmol/l)	**56**	**23.8**	**49.2**	5.7	*<3*
Glucose (mg/dl)	104	119	75	175	

Table 2. Biochemical parameters at baseline, during SSA treatment and postoperatively

Discussion

Dilated cardiomyopathy and cardiac failure without other risk factors has rarely been associated with glucagonoma.

Surgical resection may result in reversal of cardiomyopathy.

Our case provides additional evidence that dilated cardiomyopathy is a rare clinical manifestation of the glucagonoma syndrome and these patients obtain substantial benefit following surgical tumor removal.

References

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Conflicts of Interest

No conflict of interest