Malignant Acanthosis Nigricans as A Para-Endocrine Syndrome: A Rare Case Presentation

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Background

We report a case of malignant acanthosis nigricans with two unusual aspects: an atypical presentation and a remarkable response to medical treatment.

Case presentation

A 52-year-old man with no significant past medical or surgical history presented with the chief complaint of sudden-onset acanthosis nigricans versus hyperpigmentation over the neck and, unusually, in the trunk which was associated with mild lower limb oedema. His general examination revealed generalised obesity only.

Pelvi-abdominal ultrasound revealed multiple hepatic focal lesions, suggestive of either metastatic or multicentric hepatocellular carcinoma for further evaluation.

Then, a step to investigate potential malignancy was taken, performing a contrast-enhanced magnetic resonance imaging (MRI) of the abdomen and pelvis. This revealed multiple hepatic masses, mostly metastatic and an ill-defined pancreatic lesion.

Subsequent Positron Emission Tomography-Computed Tomography (PET-CT) imaging confirmed the hepatic focal lesions, and a true-cut biopsy of the hepatic lesion was performed.

Two specimens were sent for histopathological analysis. While the first specimen showed an undifferentiated carcinoma, the second specimen demonstrated conclusive evidence of metastatic neuroendocrine carcinoma grade III.

During hospitalization, the patient developed resistant hypokalemia and hypertension. 24-hour urinary free cortisol and overnight dexamethasone suppression test confirmed hypercortisolism.

Moreover, a high-dose dexamethasone suppression test confirmed an ectopic source in addition to a non-suppressed ACTH level.

Management and outcomes

The patient was started on adrenal blockade with metyrapone (1gm/day) and chemotherapy in the form of cisplatin and etoposide.

In addition, metabolic and supportive care, including basal-bolus insulin for hyperglycaemia, spironolactone and angiotensin convertase enzyme inhibitors (ACEi) for hypokalemia and hypertension and calcium/vitamin D supplementation for calcium homeostasis were given in their recommended doses.

Follow-up revealed a marked reduction of his 24-hour urinary free cortisol from 3880 ug/d at the time of diagnosis to 584 ug/day. Also, improvement of the metabolic profile and regression of the acanthosis nigricans were noticeably observed.

Discussions

Despite the beneficial role of ketoconazole in such cases, it has not been used because of sepsis-induced transaminitis and fear of further hepatotoxicity. Moreover, surgical intervention was not done because of the poor performance status.

In addition, an endoscopic ultrasound (EUS) guided biopsy from the pancreatic lesion was not performed because of significant thrombocytopenia.

Having a poorly differentiated neuroendocrine carcinoma that lacks classic carcinoid symptoms like flushing and diarrhea made somatostatin analogue an inappropriate treatment regimen.

Conclusion

Acanthosis nigricans is not always a marker of insulin resistance. Whenever rapid onset and involvement of unusual sites, malignancy should be highly considered**.**

Keywords

Malignant acanthosis nigricans, neuroendocrine carcinoma, ectopic Cushing, adrenal blockade, metyrapone.