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Case Presentation Ectopic Cushing's Syndrome Secondary to Thymic Neuroendocrine Tumors Secreting ACTH

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Abstract : This is a case of a 36-year-old Bahraini gentleman diagnosed to have Cushing's Syndrome with a large anterior mediastinal mass. He was sent abroad to the Speciality hospital in Jordan, where he underwent diagnostic video-assisted thoracoscopy, partial thymectomy and pericardial fat excision. Histopathology of the mass was reported to be an Atypical carcinoid tumor with a low Ki67 proliferation index of 5%, the mitotic activity of 4 MF/10HPF and pathological stage classification(pTNM): pT1aN1. MRI of the pituitary gland showed an ill-defined non-enhancing focus of about 3mm on the Rt side of the pituitary on coronal images, with a similar but smaller one on the left side, which could be due to enhancing pattern rather than a real lesion as reported. The patient underwent Ga68 Dotate PET/CT scan post-operatively, which showed multiple somatostatin receptor-positive lesions seen within the tail, body and head of the pancreas and positive somatostatin receptor lymph nodes located between the pancreatic head and IVC. There was no uptake detected at the anterior mediastinum nor at the site of thymic mass resection. There was no evidence of any positive somatostatin uptake at the soft tissue or lymph nodes. The patient underwent IPSS, which proved that the source is, in fact, an ectopic source of ACTH secretion. Unfortunately, the patient's serum cortisol remained elevated after surgery and failed to be suppressed by 1 mg ODST and by 2 days LLDST with a high ACTH value. The patient was started on Osilodrostat for treatment of hypercortisolism for the time being and his future treatment plan with Lutetium-177 Dotate therapy vs. bilateral adrenalectomy is to be considered in an MDT meeting.

Keywords : cushing syndrome, neuroendocrine tumur, carcinoid tumor, Thymoma
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