

## **Pancreatic Paraganglioma in MEN Syndrome**

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**Context** Paragangliomas are rare neuroendocrine neoplasms arising from paraganglia of sympathetic trunk. Pancreatic localization is extremely rare (only 15 cases reported worldwide). Pancreatic paraganglioma has never been described before in the context of multiple endocrine neoplasia-1 (MEN 1) syndrome. **Case report** A 40-year-old male, known for previous follicular adenoma and primitive hyperparathyroidism, was admitted to our hospital with high blood pressure, flushing of the face, sweating and dysaesthesia of the lower limbs. Abdominal imaging showed multifocal pancreatic masses. He underwent total pancreatectomy with histological diagnosis of multifocal paraganglioma with a proliferative index (Ki67) of 10%. Further clinical evaluation evidenced

pituitary microprolactinoma and adrenal hyperplasia. Even in the absence of genetic mutation, a diagnosis of MEN 1 syndrome was clinically established. During follow-up he developed liver metastatic disease that was treated with targeted biological therapy and multiple lines of chemotherapy. **Conclusion** A clinical presentation of MEN syndrome in absence of definable gene mutation is possible and becoming a clinical challenge. We discuss here a pancreatic paraganglioma presenting with rapid growth and prone to metastasis dissemination in a clinically evident MEN 1 syndrome. Further studies are needed to understand genetic causes and biology of this rare type of neuroendocrine tumor.