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Pancreatic NET is accompanying with extra-adrenal retroperitoneal paragangliomas

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Introduction

Pancreatic neuroendocrine tumors (PNETs) are rare, accounting for less than 3 percent of all pancreatic tumors. They may be benign or malignant and they tend to grow slower than exocrine tumors. They develop from the abnormal growth of endocrine (hormone-producing) cells in the pancreas called islet cells. This is why these tumors are sometimes referred to as “islet cell tumors.” Pancreatic NETs are either functional (produce hormones) or nonfunctional (produce no hormones). Functional NETs cause the pancreas to overproduce hormones consequently causing hormone-related symptoms. The majority of PNETs are nonfunctional tumors. As a result, these tumors are typically diagnosed once the tumor is advanced and is causing symptoms such as pain or jaundice.

Extra-adrenal retroperitoneal paragangliomas are extremely rare neuroendocrine neoplasms with an incidence of 2-8 per million. They are essentially of two types : chromaffin or sympathetic paraganglia made of chromaffin cells and nonchromaffin or parasympathetic ganglia made of glomus cells. They are neuroendocrine cells, the former with primary endocrine functions and the latter with primary chemoreceptor functions

Method & result

A 53-year-old woman with a history of hyperlipidemia and fatty liver has a suspected metastatic pancreatic NET due to multiple masses found incidentally on a CT examination. A lab finding was within normal range of tumor markers (CEA, CA19-9). No specific findings were also seen in the physical examination. An additional test, MRI, has come up with CT findings but a PET-CT showed no distant metastasis. Based on the diagnosis, we proceeded a laparoscopic distal pancreatectomy with splenectomy & Lt. adrenalectomy and excision of retroperitoneal mass. The patient was discharged without specific complication at postoperative 5 days. Postoperative histopathologic examination revealed a mass in the pancreas tail as pancreatic NET, multiple masses on the left adrenal gland and descending colon as paraganglioma, not metastatic pancreatic NET. No recurrence and non-specific findings were seen on follow up CT 1 month later

Conclusion

CT and MRI, in combination with tumor markers, are the preferred modalities for surveillance and monitoring progression. And the surgery is the gold standard of the treatment and we need continuous observation to confirm the recurrence and progress of the disease.