CASE 2

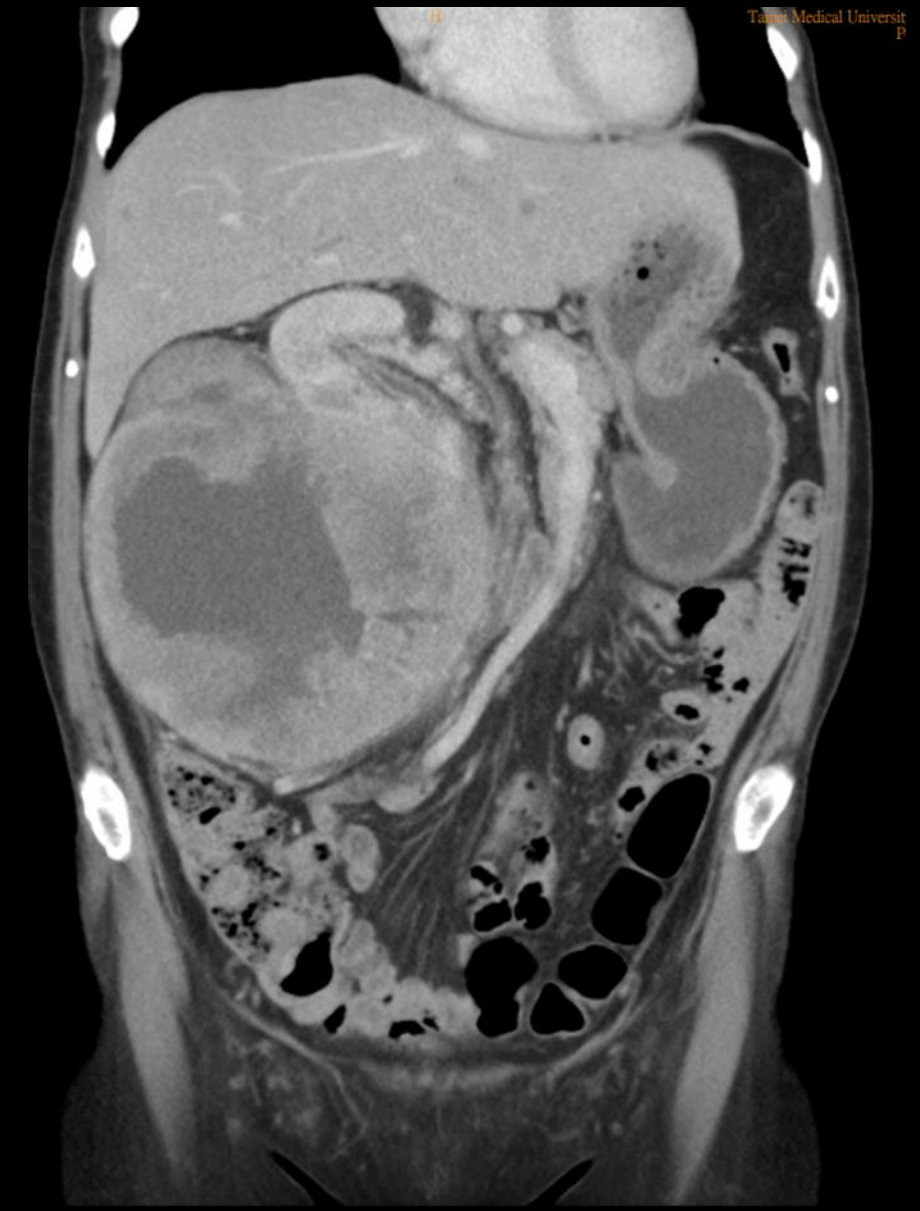
65 Y/O F

Brief history

- **Past history**
- HTN
- major depressive disorder
- breast cancer s/p partial mastectomy with radio therapy and chemotherapy
- **Chief complain**
  abdominal fullness with a palpable mass which got larger and larger recently.

Imaging studies

- Abd CT 0828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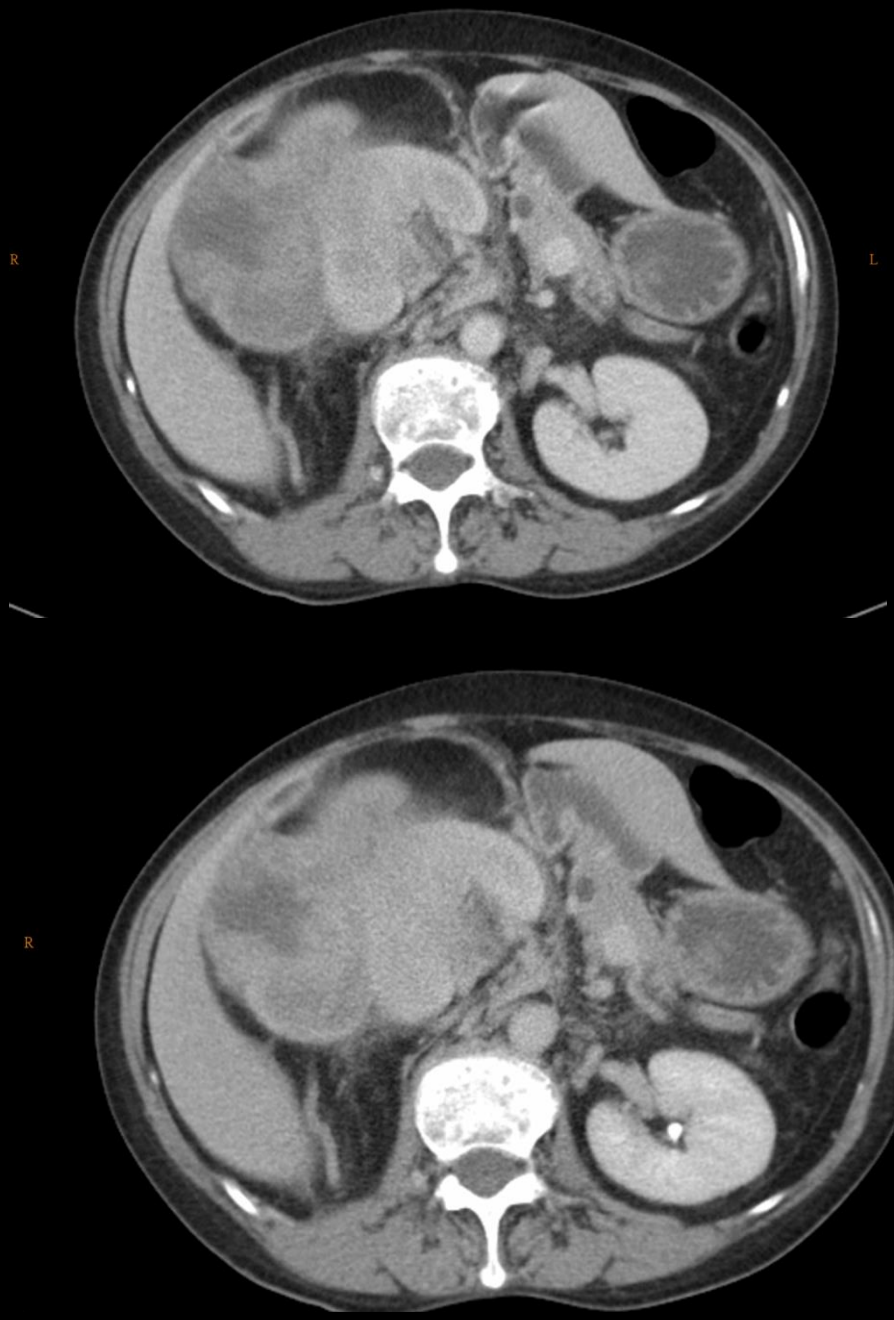
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(2024/09/09) Soft tissue, retroperitoneum, CT-guided biopsy, spindle cell tumor (see descriptions)

The specimen submitted contains 4 tissue fragments, measuring up to 1.8 x 0.1 x 0.1 cm in size, fixed in formalin
Grossly, they are white and soft.
All for sections.
Microscopically, section shows spindle cells tumor composed of moderately pleomorphic spindle cells arranged in fascicles patterns. Some ectatic vessels are seen. No evidence of tumor necrosis is seen. The mitosis feature is less than 1 per 10 high power fields. Immunohistochemistry studies reveal these tumor cells are negative for CD117 and equivocally positive for CD34. The gastrointestinal stromal tumor is not likely. Further immunohistochemistry studies reveal these tumor cells are positive for Cyclin D1, focally positive for Desmin, weakly positive for CD10 and focally weak positive for progesterone receptors (5%, weak), but negative for S-100, smooth muscle specific actin, h-caldesmon and estrogen receptors. Together with above pictures, the spindle cells tumor with focal smooth muscle and endometrial stromal differentiation should be considered. Please correlate the clinical pictures!

報告登打： 鄭建睿醫師 (病解專醫字第210號) 2024/09/19 00:00

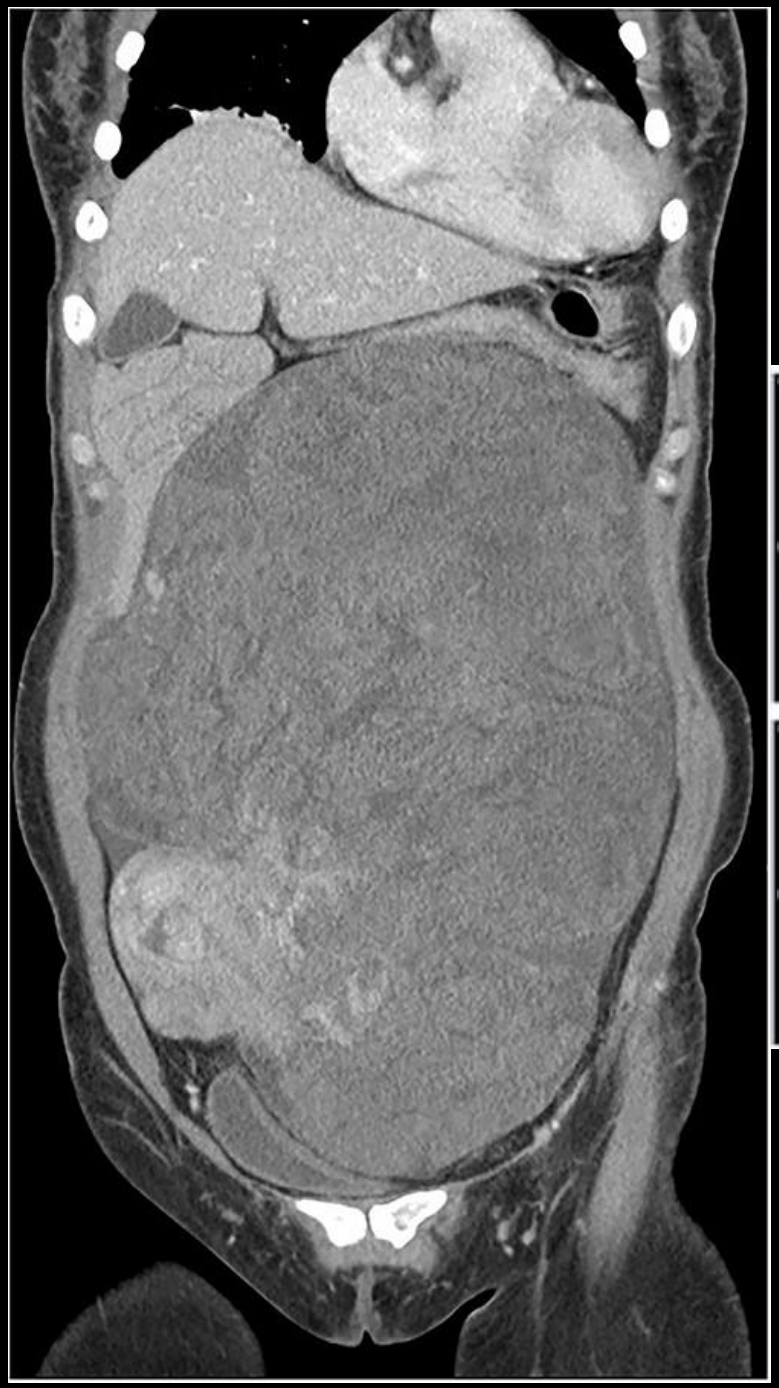
病理號碼 TH2422225　報告日期 2024-09-19

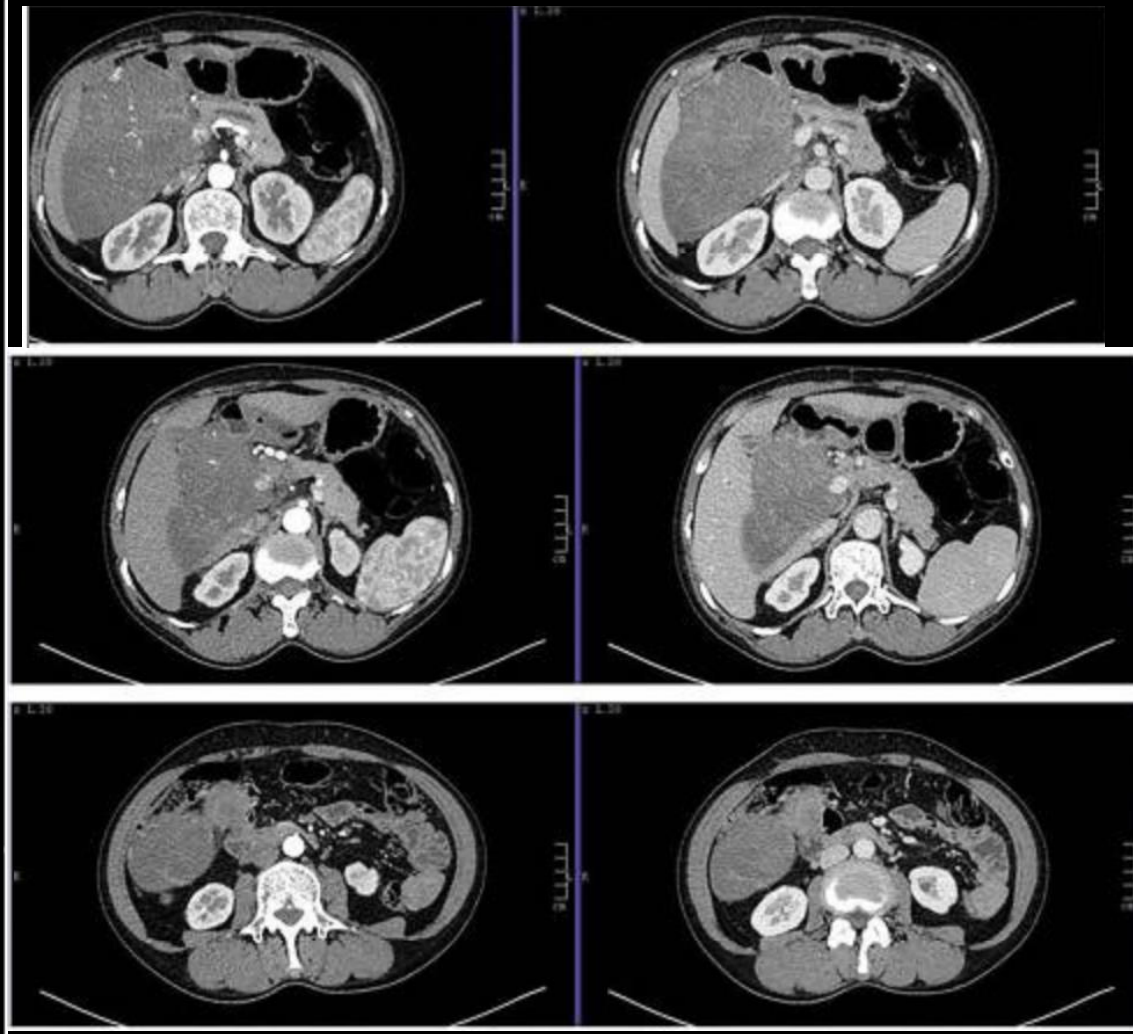
Discussion

- retroperitoneal spindle cell tumor
  - characterized histologically by a mixture of fat cells and fibroblast-like spindle cells in a matrix of collagen and mucoid material
  - rare, can occur in human soft tissue, bone, or in any part of the human body
  - Previous article reported inert biological behavior, associated with distant metastasis, and well prognosis.

Discussion

- retroperitoneal spindle cell tumor
  - Clinical examination, imaging, and histology are non-specific in the evaluation of the uncommon tumor and distinguish it from other solid masses
  - pathological and immunohistochemical tests are critical
    - β-Catenin
    - CD34 and CD117 antibodies





Hua, H., He, Z., Lei, L., Xie, H., Deng, Z., Cheng, Z., Zuo, S., Sun, C., & Yu, C. (2021). Retroperitoneal Spindle Cell Tumor: A Case Report. *Frontiers in surgery*, *8*, 764901. https://doi.org/10.3389/fsurg.2021.764901

Il D Kim, Eun J Ahn, Jung-won Yoon, Anna Choi, Sung H Joo, Retroperitoneal spindle cell tumor: a case report, *Journal of Surgical Case Reports*, Volume 2023, Issue 10, October 2023, rjad601, https://doi.org/10.1093/jscr/rjad601

Type of Mass, Origin, and Cell of Origin	Benign	Malignant
Neoplastic		
Mesodermal origin		
Adipose tissue	Lipoma	Liposarcoma
Smooth muscle	Leiomyoma	Leiomyosarcoma
Connective tissue	Fibroma	Malignant fibrous histiocytoma, fibrosarcoma, chondrosarcoma, synovial cell sarcoma
Striated muscle	Rhabdomyoma	Rhabdomyosarcoma
Blood vessels	Hemangioma, hemangiopericytoma	Angiosarcoma
Perivascular epithelioid cells	Perivascular epithelioid cell tumor (PEComa) group: angiomyolipoma, lymphangioleiomyomatosis, clear cell "sugar" tumor, clear cell myomelanocytic tumor, pigmented melanotic tumor	Sarcoma of perivascular cells
Interstitial cells of Cajal	Gastrointestinal stromal tumor (GIST)	. . .
Primitive mesenchyme	Myxoma	Myxosarcoma
Notochordal remnant	Chordoma	. . .
Miscellaneous	Myelolipoma, fibromatosis, angiomyofibroblastoma	. . .
Uncertain	Xanthogranuloma	. . .
Neurogenic origin		
Nerve sheath	Schwannoma, neurofibroma	Malignant schwannoma, neurogenic sarcoma, neurofibrosarcoma
Sympathetic nerves	Ganglioneuroma, ganglioneuroblastoma	Neuroblastoma
Chromaffin tissue	Paraganglioma, pheochromocytoma	Malignant paraganglioma or pheochromocytoma
Germ cell, sex cord, and stromal cell origin		
Germ cell	Mature teratoma, immature teratoma	Seminoma, malignant teratoma, embryonal carcinoma, yolk sac tumor, choriocarcinoma, mixed germ cell tumor
Sex cord stromal	Granulosa cell tumor, thecoma, Sertoli-Leydig cell tumor	. . .
Lymphoid or hematologic origin	. . .	Lymphoma, posttransplant lymphoproliferative disease, extramedullary plasmacytoma
Nonneoplastic*		

Key Imaging, Epidemiologic, Clinical, and Histologic Features of Primary Retroperitoneal Neoplasms

Category of Tumor	Key Radiologic Features	Key Epidemiologic, Clinical, and Histologic Features
Adipocytic		
Lipoma	Fat-containing mass with no/trace internal complexity	Rare in the retroperitoneum Even with no internal complexity, the mass should be considered a well-differentiated liposarcoma
Well-differentiated liposarcoma	Predominantly fat containing, with thick septa and soft-tissue nodularity (usually ≥1 cm)	Most common primary retroperitoneal sarcoma No potential to metastasize, but poor long-term survival due to high risk of local recurrence and potential for degeneration to a dedifferentiated liposarcoma
Dedifferentiated liposarcoma	Fat containing, with increasing internal complexity	Potential to metastasize Can arise in a preexisting or current well-differentiated liposarcoma
Myxoid liposarcoma	Paucity of fat, often present in septa or as small nodules Variable myxoid tissue (T2-hyperintense with variable enhancement)	Propensity to metastasize to unusual soft-tissue and bone locations
Pleomorphic liposarcoma	Trace of fat to no visible fat Heterogeneous appearance with necrosis and/or hemorrhage	Aggressive tumor May metastasize early to the lungs in 75% of patients
Smooth muscle		
Leiomyoma	Similar appearance to that of smooth muscle Hypointense at T2-weighted MRI, with variable enhancement	Uncommon Most often diagnosed in women of reproductive age, usually hormonally driven
Leiomyosarcoma	Heterogeneous mass with involvement of a contiguous vessel and variable necrosis	Second most common primary retroperitoneal sarcoma Classified as extravascular, intravascular, or a combination of both
Fibroblastic		
Solitary fibrous tumor	Heterogeneous appearance Often highly vascular with prominent collateral vessels at CT Flow voids at MRI	Most common location in the abdomen is the retroperitoneum Malignancy rate, 20%–40% Small percentage are associated with paraneoplastic syndromes, most commonly Doege-Potter syndrome (hypoglycemia due to excessive production of insulin-like growth)
Myxofibrosarcoma	Imaging appearance is dependent on histologic grade: low-grade tumors show abundant myxoid matrix, while high-grade lesions are more heterogeneous; may show "tail" sign at contrast material–enhanced T1-weighted MRI	Common extremity sarcoma in elderly patients, but retroperitoneal location is rare High rate of local recurrence regardless of grade; however, high-grade tumors are more likely to metastasize

Skeletal muscle:		
rhabdomyosarcoma	Heterogeneous, nonspecific infiltrative appearance, with regions of necrosis	Most common soft-tissue sarcoma in children aged 15 years and younger Most commonly seen in the head or neck Retroperitoneal location seen in 7%–19% of patients
Miscellaneous		
Undifferentiated pleomorphic sarcoma	Heterogeneous nonspecific appearance; calcifications seen in 5%–20% of cases	Formerly known as malignant fibrous histiocytoma Diagnosis of exclusion
Alveolar soft-part sarcoma	Heterogeneous hypervascular mass; necrosis and hemorrhage may be present	Manifests as a painless slowly growing tumor but frequently metastasizes
Cystic		
Lymphatic malformation	Involves more than one retroperitoneal compartment Insinuates between structures Fluid-fluid levels may be seen Chylous content may be detectable with chemical-shift MRI	Considered a form of slowly growing vascular malformation Most common in the head and neck; only 5% in the abdomen Treatment is percutaneous sclerotherapy and/or surgery
Tailgut cyst/retrorectal cystic hamartoma	Uni- or multilocular cystic masses in the presacral space, with variably thick septa and occasional calcifications	Congenital lesions arising from the embryonic hindgut Treatment is surgical excision owing to risk of infection and malignant degeneration
Cystic teratoma	Presence of fat-fluid levels and calcifications (coarse or toothlike)	Germ cell neoplasms Retroperitoneal location is rare in adults

Neurogenic

Neurofibroma	Target sign: peripheral signal hyperintensity with central signal hypointensity	Subtypes: localized, diffuse, plexiform Neurofibromatosis 1 is associated with the presence of multiple neurofibromas or a plexiform neurofibroma
Schwannoma	Difficult to distinguish from a neurofibroma May show fascicular sign (numerous internal ringlike structures at T2-weighted MRI)	Mostly sporadic Small subset associated with neurofibromatosis Rate of successful surgical excision generally higher than that of neurofibromas Malignant degeneration rare
Malignant peripheral nerve sheath tumor	Difficult to distinguish from neurofibroma and schwannoma Clues include larger size, ill-defined borders, internal heterogeneity with necrosis and/or hemorrhage, and new or worsening neurologic deficit	Approximately 50% arise de novo The rest are seen in neurofibromatosis 1, with a small subset occurring in patients with a remote history of radiation therapy
Ganglioneuroma	Paravertebral mass Heterogeneous with a variable amount of myxoid stroma "Whorled" appearance at T2-weighted MRI	Benign tumors arise from paravertebral sympathetic ganglia Manifest most commonly in children, adolescents, and young adults Rarely secrete sufficient amounts of catecholamines to cause sympathetic symptoms (eg, flushing)
Ganglioneuroblastoma	Heterogeneous Calcifications, necrosis, and hemorrhage more common than ganglioneuroma Avid radiotracer uptake at iodine 123 (^{123}I) metaiodobenzylguanidine (MIBG) MRI in approximately 70% of cases	Malignant tumor Commonly arises in adrenal medulla Primarily affects children aged 2–4 years
Paraganglioma	Classically hyperintense at T2-weighted MRI, with hypervascularity More complex appearance is frequently observed because of hemorrhage and/or necrosis Gallium 68 (^{68}Ga) tetraazacyclododecane tetraacetic acid-octreotate (DOTATATE) PET/CT is superior for localizing and staging	Most common location in abdomen is the origin of the inferior mesenteric artery near the aortic bifurcation (organ of Zuckerkandl) Associated with multiple endocrine neoplasia syndromes and Von Hippel–Lindau syndrome