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Intrathyroidal Schwannoma

Benign nonepithelial tumors of the thyroid gland are very rare lesions in children and adults. They include lesions such as vascular tumors, smooth muscle tumors and tumors of nerve origin. Primary Schwannoma, also known as neurilemmoma, of the thyroid gland was first reported in 1964 with most cases seen in the adult population. Schwannomas are peripheral nerve tumors originating from neuronal sheath cells (Schwann cells). They mostly occur in the 40 to 60 years old age groups without sex predilection. They are benign tumors that can be found anywhere in the body with half of them originating in the head and neck region. Neurilemmomas of the neck region arise from the cranial nerves with the vagus nerve or its branches being the most frequently affected followed by the cervical sympathetic chain. In the thyroid gland they arise from the sensory nerves or from autonomic innervation of the gland. Half of all reported cases of intrathyroidal Schwannomas come from Asia. The clinical presentation of a intrathyroidal schwannoma is a typical palpable non-tender thyroid nodule. Thyroid function tests are within normal limits. They have a slow but progressive growth causing compression of vital structures of the neck. Thyroid scintigraphy demonstrates a cold area within the affected lobe. Ultrasound describes a well-delineated, solid or predominantly solid tumor of low echogenicity with variable cystic degeneration. Fine needle aspiration biopsy of the tumor is diagnostic of the histologic nature of the mass. Two growth patterns are seen within the lesion: a predominantly cellular area composed of spindle-shaped Schwann cells with little stromal matrix (Antoni A type tumor), and a less cellular myxoid area with microcyst formation (Antoni type B tumor). On immunohistochemistry Schwannomas are positive for S100 and Vimentin, and negative for Desmin and SMA. Management of intrathyroidal Schwannomas is either enucleation or total thyroid lobectomy. Intrathyroidal Schwannomas are associated with an excellent prognosis once completely removed.

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Glucagonoma

Glucagonoma is a rare neoplasm of the pancreatic neuroendocrine islet alpha-cells where they secrete abundant glucagon occurring in one of every 20 million individuals. Glucagonoma tumors excessive secretion of proglucagon-derived peptides is clinically characterized by a necrolytic migratory erythema (NME), diabetes mellitus, weight loss, anemia, painful glossitis, stomatitis, thromboembolic complications, dilated cardiomyopathy and neuropsychiatric disturbances. Vast majority of glucagonomas are sporadic and occurs in adults. Children with MEN type 1 can harbor this tumor. Median time between onset of symptoms and diagnosis is 3-4 years. Glucagon hypersecretion increase hepatic glucose output antagonizing the effect of insulin and causing Diabetes. Also it exerts a catabolic role attenuating protein synthesis. Glucagonoma syndrome is the triad of glucagon-secreting tumor, diabetes and NME. The NME is the most specific manifestation of glucagonoma with early recognition leading to a rapid diagnosis of the presence of a glucagon-producing tumor. NME distributed in the groin, perineum and distal extremity is characterized by an annular pattern of erythema and centrally formed fragile vesicles, bullae and crusts present in 70% of patient with glucagonoma. Glucagonoma is a slow growing and low malignancy tumor. Metastasis represent the main prognostic factor for glucagonoma with 100% survival in cases without metastasis. Metastasis occur to the liver and peripancreatic lymph nodes. Somatostatin analog therapy may be useful in relieving glucagonoma syndrome by inhibiting glucagon secretion and counteracting its effect. CT-Scan is the diagnostic modality to diagnosed a glucagon producing tumor of the pancreas. Glucagonoma typically occurs in the distal pancreas. Fasting glucagon levels are elevated. Complete resection of the primary pancreatic tumor and limiting metastasis, including liver transplantation, is the only chance of cure.

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Vipoma

Vipoma is a very rare malignant neuroendocrine tumor. Most Vipomas arise in the pancreas with 10% of them arising from other tissues of neural crest origin of the body. In children a ganglioneuroblastoma can behave as a Vipoma. Most neurogenic tumors associated with the Vipoma syndrome have been found in children. Vipomas secrete vasoactive intestinal peptide (VIP), a hormone which stimulates adenosines 3',5'-cyclic phosphate (cAMP) production by the intestinal tract causing watery diarrhea, hypokalemia, hypophosphatemia, hypomagnesemia, hyperchloremic metabolic acidosis from severe intestinal loss of bicarbonate and achlorydia syndrome due to inhibition of gastric acid production. Occasionally hypercalcemia due to release of PTH by the tumor, glucose intolerance and hypotension can occur. With the diarrhea the patient can develop flushing similarly to the carcinoid syndrome. Majority of Vipomas are sporadic cases and 50-60% have metastasized by the time the diagnosis is made. VIP is elevated in all cases of Vipomas and can be measured in blood. Diagnosis can be confirmed using imaging such as US, CT-Scan (hyperattenuating lesion in the arterial phase that becomes inconspicuous in the venous phase), MRI, Somatostatin-receptor scintigraphy or PET-Scan. Vipomas appear as well-defined homogenous mass with central necrosis and hypervascularized. Most Vipomas tumors in the pancreas occur in the tail. Surgical extirpation is the mainstay of treatment of Vipomas. If a tumor has been identified, complete surgical excision is the primary form of treatment. If the tumor cannot be removed completely surgical debulking may have a palliative effect. Medical therapy with somatostatin analogue can be used for symptomatic relieve in cases of inability to remove the tumor completely. Others alternatives include peptide receptor radionuclide therapy, streptotozin chemotherapy, ablation, hepatic artery embolization or liver transplant.

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