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Solitary Fibrous Tumor Associated With Papillary Thyroid Carcinoma and Pituitary Macroadenoma

Tumor fibroso solitario asociado a carcinoma papilar de tiroides y macroadenoma hipofisario

The patient is a 31-year-old male from Bolivia with a prior history of a tumor of the sacrum with pelvic extension diagnosed as a result of hypoglycemia. In his native country in 1998, he underwent partial tumor resection. Radiotherapy was proposed at that time, but the patient did not accept this treatment.

He came to our Emergency Department in December 2009 due to hypoglycemia. The patient reported progressive enlargement of the nose, the folds of the frontal region and a previously detected cervical node.

On physical examination, a mass was palpated in the left flank and iliac fossa, and another rock-hard mass was detected on digital rectal examination in association with the sacrum.

In February 2010, total thyroidectomy was performed, showing pathology consistent with papillary carcinoma with capsular invasion.

In May 2010, exploratory laparotomy discovered multiple large hypervascular tumors in the left flank that encompassed the descending colon, as well as tumors measuring 2–3 cm in diameter in the appendix, lateral wall of the cecum, antimesenteric edge of the terminal ileum and several smaller tumors implanted in the ileal loops. We carried out resection of the descending colon, colorectal anastomosis, appendectomy and resection of tumors in the cecum, terminal ileum and ileum. The pelvic mass in the sacrum was considered untreatable.

The pathology study reported that the mass was a moderate-grade (G2) malignant solitary fibrous tumor with moderate tumor differentiation, mitotic index 14/10 HPF, vessels with hemangiopericytomatous pattern and intratumoral necrosis of 5%.

The patient has presented no further episodes of hypoglycemia and has even had progressive improvement of the acromegaloid features. He started chemotherapy and is being studied by the Genetic Counseling Unit for suspected Cowden syndrome.

Solitary fibrous tumors are mesenchymal neoplasms that are usually located in the pleura; they were initially considered tumors of mesothelial origin and given various names. They were first described by Klemperer and Rabin (1931)[1] as a variant of a fibrous mesothelioma. In recent years, there have been numerous reports of fibroid tumors in other body locations.

Clinically, an extrathoracic solitary fibrous tumor causes symptoms associated with its large volume (i.e. compression). However, this same tumor is often found in the pleura, and chest radiographs are frequently performed. These tumors are known for having a benign clinical course, although relapses and metastases may be seen.

Less than 5% of cases present with symptoms of hypoglycemia or Doege–Potter syndrome due to the overexpression of insulin growth factor (IGF2) caused by the tumors themselves, which are generally large tumors with an elevated mitotic index. Nonetheless, it has been observed that in the case of retroperitoneal solitary fibrous tumors, this percentage could reach 11.5%.[2]

Although IGF production is usually implicated as a cause of hypoglycemia, elevated IGF2 levels are observed in a minority of cases.

The diagnosis of solitary fibrous tumors quite often depends on immunohistochemistry as well as the pathology study. Chan et al.[1] concluded that CD34+ positivity was the main diagnostic criteria for this entity, which was positive in 90%–95% of cases. Nonetheless,[3] this marker is not specific

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Table 1 – General Work-up.

GH<0.05 ng/ml (low)
IGF-1<25 ng/ml (low)
Insulin<2.00 mU/l (normal)
TSH 0.97 mU/l (normal)
T4L 0.45 ng/dl (normal).
IGF II (mature form): 2744 (normal up to 3000)
IGF II/IGF I>100 (suggestive of high levels of *big-IGF II*)

Complementary Tests		
Thyroid	Abdomen	Brain
Cervical ultrasound: goiter of the right thyroid lobe, with a large nodule measuring 35×33 mm Thyroid scintigraphy: dominating nodule in the right lobe, with increased uptake, inhibiting the rest of the gland FNA: pattern compatible with papillary carcinoma	Abdominal CT: tumor affecting the sacrum associated with nodules in the right pelvic and hypogastric region adjacent to the abdominal wall, as well as multiple intraabdominal lesions with solid characteristics, the largest measuring 9×11 cm in the left flank and others measuring 5 cm affecting the right iliac vessels	MRI (January 2010): pituitary macroadenoma with signs of intralesional hemorrhage, with no infiltration of the cavernous sinus or the optic chiasm

Octreotide scan: right thyroid lobe and multiple intraabdominal masses with octreotide uptake (expression of somatostatin receptors).

because there are other mesenchymal tumors, such as hemangiopericytoma, that are also CD34-positive.

The presence of this marker makes the diagnosis highly probable when accompanied by other histopathological factors, such as the presence of spindle cells within a collagen-rich stroma, with alternating hypocellular and highly populated areas (''pattern-less pattern'')[4] and surrounded by hemangiopericytoma-like vessels. They also tend to be immunoreactive to bcl-2.

Solitary fibrous tumors are considered malignant if there are atypical nuclei, focal areas with high cellularity, and areas with >4 mitoses per 10 HPF, either with or without necrosis.[3]

In general, the prognosis is favorable, but complete excision is recommended of the entire capsule with wide margins, as well as follow-up for some time due to the probability of recurrence years after surgery.

Despite having searched the literature, we have found no published cases associating solitary fibrous tumors with other types of cancers, as in our case report.

A higher prevalence of malignant disease has been described in cases with acromegaly, especially colon neoplasms. In our patient, however, the acromegaloid traits did not develop until years after the first resection of the abdominal tumor.

In addition, there is a subgroup of growth hormone-producing pituitary adenomas that may originate from a mutation of the alpha chain of the G protein, which is a mutation that can be shared by thyroid and adrenal tumors.[5]

In our patient, the undetectable levels of GH and IGF1 as well as the chronology of his disease caused us to consider whether there may be a correlation between the 3 neoplasms as part of a hereditary/genetic syndrome causing complete deregulation of the growth hormone axis (Table 1).

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