

Introduction :

Solitary fibrous tumor(SFT) is associated with multiple paraneoplastic syndromes, including refractory hypoglycemia, hypertrophic pulmonary osteoarthropathy(HPOA) and elevated B-hCG . We present a patient with SFT associated with hypoglycemia, neurologic disease, and HPOA.

Case Description:

A 60-year-old male with hypertension presented with slurred speech. As per family, the patient was in his normal state of mind 4 days prior to admission and was able to independently perform his ADLs. He was found in bed covered by urine and feces and was admitted for possible CVA. His physical examination was remarkable for reduced air entry on the left lower lobe and bilateral clubbing of the fingers. Neurological examination was notable for staccato speech, intention tremor, unsteady wide-based gait, dysdiadochokinesia with normal cognition.

On investigation, MRI of the brain was unremarkable and the chest X-ray displayed marked elevation of the left hemi-diaphragm. The CT scan of the chest showed a large (21x16.5x16.7cm) complex heterogeneous mass

which compressed and displaced the left lower lobe(Figure 1).

The biochemical workup were all within normal range except for fasting blood glucose levels which were always less than 60 mg/dl. The patient had persistent hypoglycemia despite continuous intravenous dextrose supplementation.

The patient underwent CT-guided lung biopsy and pathology report showed rounded and spindled cells arranged in a variable background containing circumferentially hyalinized vessels (Figure 2).

Immunohistochemistry staining was positive for STAT6, a specific marker for solitary fibrous tumor. The findings were consistent with the solitary fibrous tumor. Further testing showed low level of C-peptide, insulin, IGF-BP3, IGF-1 with IGF-2 level 200. The blood sugar was stabilized with octreotide subcutaneous injection and dextrose oral gel.

Paraneoplastic cerebellar degeneration was suspected and antibodies were sent; Anti-Hu, anti-Yo, anti-Ri were negative.



Figure 1. Large (21x16.5x16.7cm) complex heterogeneous mass which compressed and displaced the left lower lobe

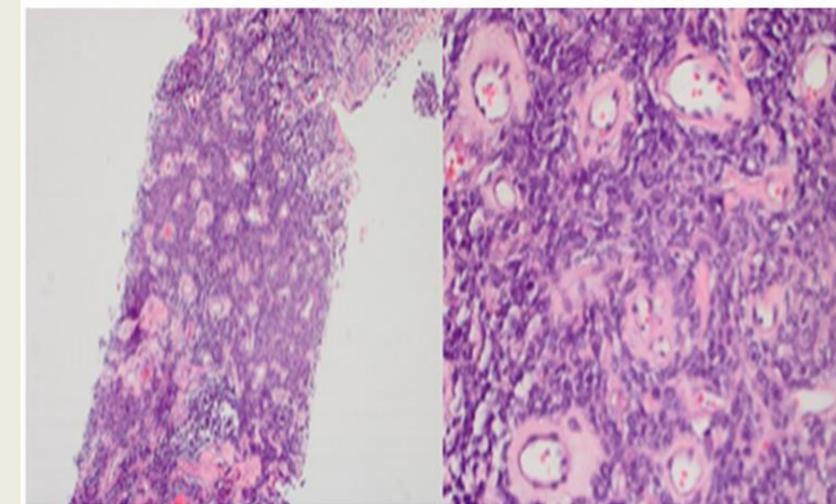


Figure 2. Rounded and spindled cells arranged in a variable background containing circumferentially hyalinized vessels

Discussion:

Solitary fibrous tumor is a rare mesenchymal tumor. The standardized incidence rate is estimated to be 1.4 per million [1,2]. It is difficult to diagnose SFT on radiological imaging, and the sensitivity of CT is low [3]. The diagnosis is confirmed by histopathology with immunohistochemistry. Several paraneoplastic syndromes have been related to SFT, including refractory hypoglycemia known as “Doege-potter syndrome”, hypertrophic pulmonary osteoarthropathy (HPO) [3,4]. Paraneoplastic syndromes are caused by autoimmune processes and several antibodies have been used to facilitate the diagnosis but some patients have no identifiable antibodies.

Ref:

1. Thorgeirsson T, Isaksson HJ, Hardardottir H, Alfredsson H, Gudbjartsson T. Solitary fibrous tumors of the pleura. An estimation of population incidence. *Chest*. 2010;137:1005–6.
2. Ardisson F: Thoracic malignant solitary fibrous tumors: Prognostic factors and long-term survival. *J Thorac Dis*, 2011; 3: 84–85
3. Robinson LA. Solitary fibrous tumor of the pleura. *Cancer Control* 2006;13:264-9
4. Ardisson F: Thoracic malignant solitary fibrous tumors: Prognostic factors and long-term survival. *J Thorac Dis*, 2011; 3: 84–85