Doege-Potter Syndrome

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Abstract

Background: Amongst pleural tumors, solitary fibrous tumors (SFT) comprise a histologic spectrum of rarely metastasizing fibroblastic mesenchymal neoplasms that are not common. Doege Potter syndrome (DPS) is characterized by large solitary pleural fibrous tumors associated with recurrent episodes of hypoglycemia due to increased serum levels of IGF-2. If untreated, it can cause fatal hypoglycemia with grave consequences. It is a case of Doege Potter syndrome (DPS) that presented to institute of chest medicine, Mayo Hospital Lahore on October 30, 2017.

Key Words: Doege Potter Syndrome(DPS); Non-islet Cell Tumor Hypoglycemia(NICTH); Solitary Fibrous Tumor (SFT); Insulin like Growth Factor(IGF-II)

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Introduction

Solitary fibrous tumor (SFT) that is associated with NICTH is referred as the Doege-Potter syndrome. It is a rare Para neoplastic syndrome presenting as a hyperinsulinemic hypoglycemia from the ectopic secretion of a pro-hormone of insulin-like growth factor II (IGF-II) from a solitary fibrous tumor.[1] Surgical resection is curative in the majority of cases. They are large encapsulated well defined tumors with high rates of mitosis and a malignancy rate of 12-15%.[2] It may cause pain and symptoms resulting from adjacent structures compression.[3] If the diagnosis is not suspected and treatment is delayed, it can lead to hypoxic cerebral injury or death.[1] The underlying tumor can be a benign or malignant pleural tumor but may be present in extra pleural sites. This refractory hypoglycemia is directly proportional to increased cellular replication of tumor.[4] Less than 100 cases are reported so far. Surgical resection of tumor usually leads to symptom resolution

We are reporting a case of Doege-Potter syndrome which presented to us with worsening of shortness of breath and recurrent episodes hypoglycemia and significant weight gain.

Case Report:

A 45 years old female of low socioeconomic status, with no smoking history presented to Pulmonology outpatient department (OPD Mayo Hospital Lahore) with worsening of cough and shortness of breath for last 1 month. These Symptoms were present for last 4 years with multiple hospital admissions. These complaints were also associated with significant weight gain of 8-10 kgs in last 1.5 years. There were multiple episodes of altered state of consciousness and drowsiness which later on came out to be recurrent attacks of hypoglycemia. She didn't give any history of Diabetes Mellitus. On examination she was dyspneic with respiratory rate of 26/min and O2 saturation of 92% at supplemental O2 of 2 L/min. There were decreased chest movements, decreased vocal fremitus, percussion note was dull, decreased vocal resonance and decreased intensity of breath sounds on the left side of the chest. Bilateral pedal edema was also present.

X-ray chest PA view revealed homogenous opacification occupying lower half of the left hemi-thorax with well defined upper margins. While, CT chest with IV contrast showed large mildly enhancing soft tissue density mass in the left hemi-thorax sparing upper lobe, causing mediastinal shift and

abutting great vessels. Her metabolic profile was normal but her C-peptide levels were low (0.02ng/ml). CT guided True cut biopsy demonstrated spindle cell lesion and on immunohistochemical analysis tumor expressed STAT6 and BCL-2 tumor markers. Immunohistochemical assay was done from Agha Khan University Hospital, Karachi. Thus, a diagnosis of a benign pleural SFT with Doege Potter syndrome was made. The patient was optimized for her medical issues. Later on, she underwent surgical resection of tumor, leading to symptom resolution.

Discussion

The relationship between fibrous pleural tumors and low blood sugar level was first described by Doege and Potter in 1930.4, 5Doege Potter syndrome (DPS) is a Para neoplastic condition of non-islet cell tumors. Only two to four percent of patients with SFT present with hypoglycemia.[6] Male to female ratio is 3:1.[7] SFTs may arise at any age, but are most common in the fifth to seventh decades. Dysregulatory function of IGF-1 and IGF-2 molecules by over expression have a major contribution in disruption of normal cellular function. It eventually leads to tumor formation.[8] Being a high molecular weight particle, IGF-2 protein has a lower tendency to couple with plasma proteins.[9] As high molecular weight IGF-2 assays are not accessible in the market, so ratio of IGF-1 and IGF-2 is used in the diagnosis of DPS. The clinical presentation of both benign and malignant SFT is usually comparable and local recurrences can occur in both. Surgical removal of the tumor is a gold standard treatment for both original and recurrent benign and malignant tumors,[10] which improves symptoms of hypoglycemia. Chemotherapy is another available option. Treatment for hypoglycemic attacks includes scheduled snacks, nocturnal or continuous dextrose infusion or enteral tube feeding, corticosteroids and continuous glucagon infusion.[11] Treatment with glucocorticoids reduces the frequency of severity of hypoglycemic attacks. As far as the progress is concerned there is 10 years disease specific survival rate of 73-100% with 10-25%recurrence rate in 10 years depending upon the type of tumor.

Conclusion

Doege Potter syndrome is an uncommon paraneoplastic phenomenon. On balance, very patient with pleural tumors and repeated attacks of low blood sugar levels along with low C-peptide levels, DPS should be considered. Early detection of disease is very important for prompt treatment.

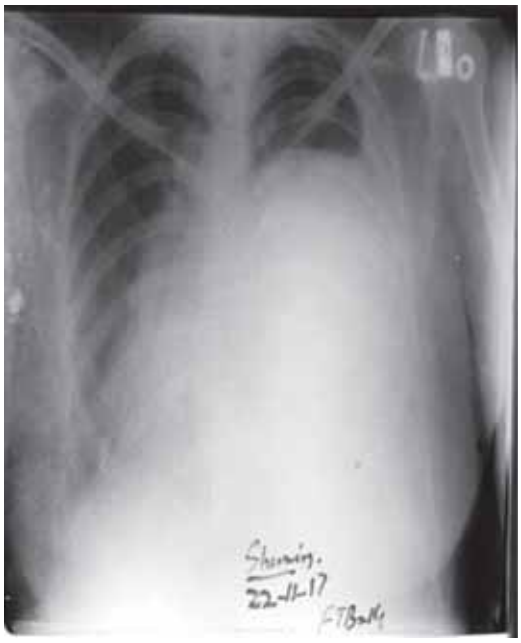

Figure A. X-ray chest PA view showing homogenous opacity in left middle and lower radiological zone

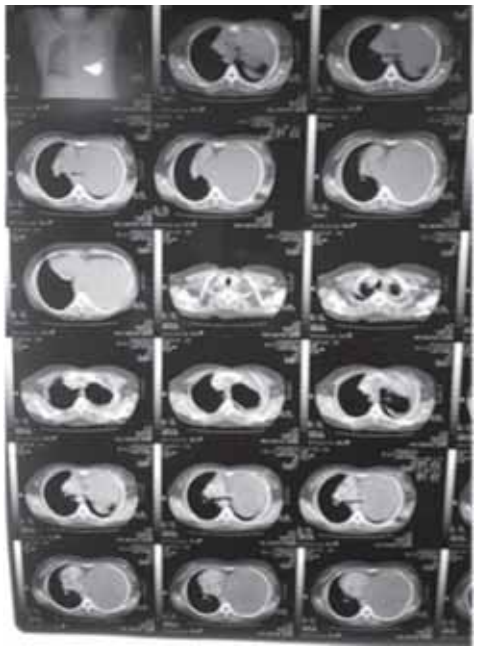

Figure B. CT chest with iv contrast showing soft tissue density mildly enhancing mass lesion taking most of the left hemi thorax.

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