

Thymic Carcinoma Presenting as Sjögren's Syndrome

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ABSTRACT

Sjögren's syndrome (SjS) is a chronic and progressive autoimmune inflammatory disorder characterized by infiltration of T cell lymphocytes into the exocrine glands (parotid and lacrimal) resulting in oral and ocular dryness. Approximately 40% of patients with Sjogren's syndrome will develop systemic (extraglandular) features. Thymic tumors are rare neoplasms that arise in the anterior mediastinum. While the association between autoimmune disorders and thymomas has been well documented, the relationship between thymic carcinomas and autoimmune disorders is less defined. We herein report a case of a 59-year-old female with symptoms of SjS and though it is a rare complication, we think investigation for possible malignancy should be a part of routine work up after the diagnosis is established

Introduction

Thymic carcinoma, an epithelial tumor, is a rare, invasive mediastinal neoplasm and accounts for 0.06% of all thymic tumors [1]. Thymic carcinoma constitutes a heterogeneous group of tumors in terms of clinical and pathological characteristics. The prognosis is often poor with a tendency to metastasize and have an aggressive course. The five-year survival rate is approximately between 30-50% [2].

Primary Sjogren's syndrome manifests as dry eyes and dry mouth with or without associated systemic features. On the other hand, secondary Sjogren's syndrome is characterized by dry eyes and dry mouth that occurs in the presence of another autoimmune disorder. Secondary Sjogren's is commonly seen with rheumatoid arthritis, systemic lupus erythematosus (SLE), or scleroderma [3]. As the disease progresses, it can also produce extraglandular manifestations that can affect multiple organ systems including but not limited to the kidneys, lungs, joints, and peripheral nerves. Sjogren's syndrome is diagnosed after clinical features of sicca syndrome are present with either serologic evidence or glandular evidence of autoimmunity as the cause [3].

There are several autoantibodies that are commonly seen in Sjogren's syndrome. SSA and SSB antibodies (also called anti-Ro and anti-La respectively) are created against several autoantigens [4]. SSA antibodies can be seen in a number of autoimmune disorders including Sjogren's syndrome, SLE, inflammatory myositis, primary biliary cholangitis and interstitial lung disease. SSB antibodies are specific for SLE and Sjogren's syndrome [4].

The SS-A autoantigen is an intracellular RNA-protein complex that is the target of autoantibodies present in patients with SjS and SLE [5,6]. SS-A52 amino acid sequence of Ro52 autoantigen was reported in 1991, it is expressed in the cytoplasm of ductal cells of patients with SjS. It may have a role in mRNA

stability and an important role in the regulation of inflammation [7]. SS-A60 amino acid sequence of Ro60 autoantigen was reported in 1980, it is found in the cytoplasm and nucleus of acinar cells and ductal cells of patients with SjS. It also might play a role in the regulation of inflammation [7].

We present a rare case of thymic neuroendocrine tumor in the setting of primary

Sjogren's syndrome with SS-A and SS-B antigen expression in a 59-year-old female who presented for evaluation of sicca complaints with corresponding antibodies to SSA and SSB. It was the evaluation of her new onset Sjogren's syndrome that led to the diagnosis of a thymic carcinoma.

Case Presentation

A 59-year-old-female with a past medical history of Lyme disease was referred to rheumatology by her primary care physician for positive rheumatologic lab work. The patient admitted to malaise, dry eyes, dry mouth, significant fatigue, occasional joint discomfort and generalized aches and pains for many years. She denied any muscle pain, hair loss, Raynaud's phenomenon, significant morning stiffness, rashes, sun sensitivity, chest pain, dizziness, and urinary symptoms.

Past surgical history included a hysterectomy. Family history was significant for heart disease in her parents and rheumatoid arthritis in her father. She denied smoking cigarettes, alcohol consumption or illicit drug use. Home medications included: progesterone 100 mg capsule daily for menopause. She was allergic to Bacitracin and penicillin. Her vitals during the office visits: Blood Pressure (BP) 122/77, Pulse 60 bpm, Temp 97.8 °F (36.6 °C), Height 1.702 m (5' 7"), Weight 63.5 kg (140 lb), Body mass index (BMI) 21.93 kg/m². On the physical exam she appeared well-developed. She had dry eyes, dry mouth, and no scleral icterus. Cardiac exam was significant for a regular rate and rhythm with no friction rub or gallops. She had normal breath sounds with no stridor or wheezes. Her abdomen was soft,

nontender and nondistended. The musculoskeletal exam showed mild hypertrophic bony changes to both knees and the carpometacarpal (CMC) joints. There was also bony hypertrophy noted in the Proximal Interphalangeal (PIP) and distal interphalangeal (DIP) joints of the hand.

Metacarpophalangeal (MCP) joints were spared. There was crepitus in both knees. Cervical spine had a normal range of motion and the neck was supple. Skin was warm and dry with no erythema or rash noted. No abnormal muscle tones.

Serum labs are shown in tables 1 and 2 and imaging results are shown in figures 1-3.

Table 1: Lab Work on Initial Presentation: Complete Metabolic Panel and Complete Blood Count

Tests	Results	Normal Ranges	Units
Sodium	138	135-147	mmol/L
Potassium	4.1	3.5-5.5	mmol/L
Chloride	101	96-108	mmol/L
HCO ₃ , bicarbonate	23	19-29	mmol/L
BUN, blood urea nitrogen	14	6-20	mg/dL
Creatinine	0.85	0.49-1.02	mg/dL
Glucose	98	70-99	mg/dL
Calcium	9.5	8.6-10.4	mg/dL
AST, aspartate aminotransferase	18	<32	U/L
ALT, alanine transaminase	14	<33	U/L
ALP, alkaline phosphatase	83	40-156	U/L
WBC, white blood cell	5.73	4-10	x10 ³ /uL
RBC red blood cell count	4.13	3.58-5.19	x10 ³ /uL
Hb, hemoglobin	12.8	11-15.5	g/dL
MCV, mean corpuscular volume	90.3	78-98	fL
RDW, red cell distribution width	12.7	12-15.5	%

Table 2: Lab work on presentation: Rheumatologic Labs

Tests/titers	Results	Description	Reference	Units
Antinuclear antibody (ANA)	1:320	ANA pattern of nuclear and discrete nuclear		
ANA	1:1280	ANA pattern of nuclear and speckled		
Double stranded deoxyribonucleic acid (DsDNA)	12.4	N/A	<27	IU/ml
Rheumatoid Factor	13	N/A	<14	IU/ml
Anti-smith Antibody	6.1	N/A	<20	{CU}

Erythrocyte sedimentation rate (ESR)	23	N/A	<31	mm/ h
Sjogren's syndrome antibody A (SSA) 52 RO (ENA) Antibody (Ab) IgG	87.3	N/A	<20	{CU}
SSA 60 (RO) (ENA) Ab IgG	>1374.8	N/A	<20	{CU}
SSB Ab	250.4	N/A	<20	{CU}
Ribonuclear protein antibody RNP Ab	<3.5	N/A	<20	{CU}
Lyme IgM Ab	0.47	N/A	<0.91	

Lyme disease investigations were as follows- Lyme Western Blot (WB) IgG band 41 positive, Lyme WB IgG band 18, 23, 28, 30, 39, 45, 58, 66,93 were all negative, Lyme WB IgM band 23 and 41 positive, Lyme WB IgM band 39 negative. She underwent chest x-ray (CXR) as of her initial workup which released a 7cm, well-circumscribed sharply margined lesion likely within the anterior mediastinum, with no associated interstitial infiltrate. She subsequently had a Computed Tomography (CT) Chest without contrast was reported a large mass suspicious for thymoma and

ground glass changes in the left upper lobe. There are multiple bilobar hepatic hypodense lesions, possible metastasis and nodular left adrenal thickening. Positron Emission Spectrum (PET) scan revealed a 72 x 42mm well circumscribed anterior mediastinal mass at the level of the main pulmonary artery that was hypermetabolic with increase uptake of F18 fluorodeoxyglucose (Figure 1, 2 & 3). There was an additional well- circumscribed anterior mediastinal soft tissue nodule located immediately deep to the manubrium at the level of the aortic arch, measuring 17 x 13 mm.



Figure 1: Full body F18 Fluorodeoxyglucose Positron Emission showing increased uptake in the anterior mediastinum, a coronal view.

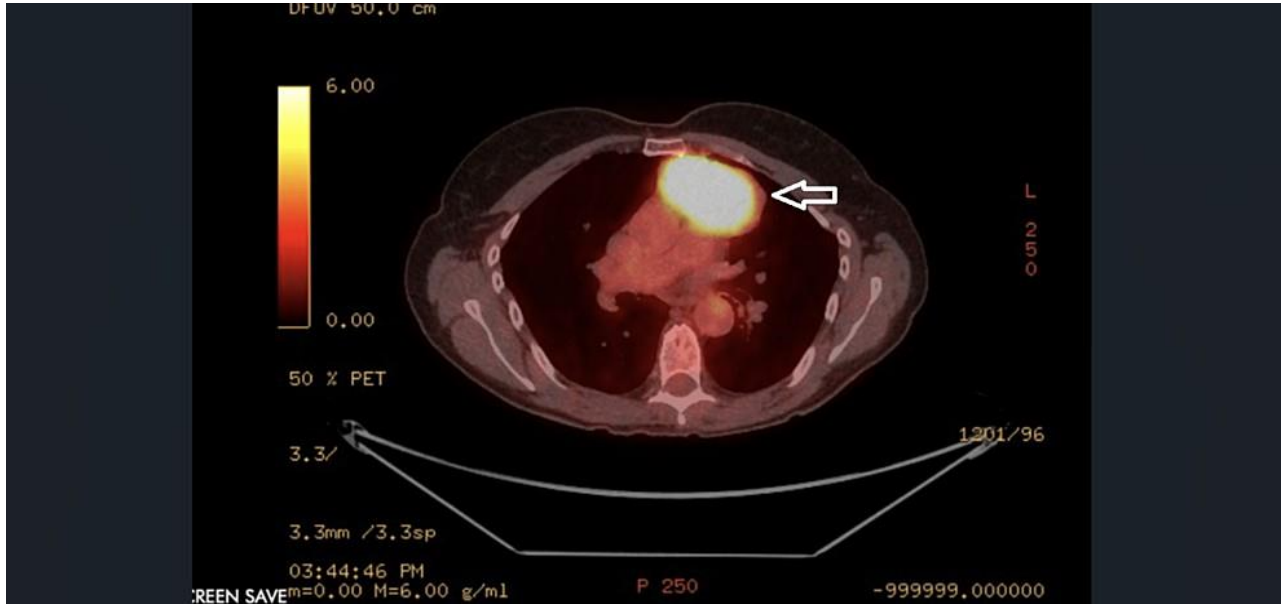


Figure 2: 72 x 42 mm well circumscribed anterior mediastinal mass is demonstrated centered at the level of the main pulmonary artery. The lesion is markedly hypermetabolic. There is an additional well- circumscribed anterior mediastinal soft tissue nodule located immediately deep to the manubrium at the level of the aortic arch, measuring 17 x 13 mm.

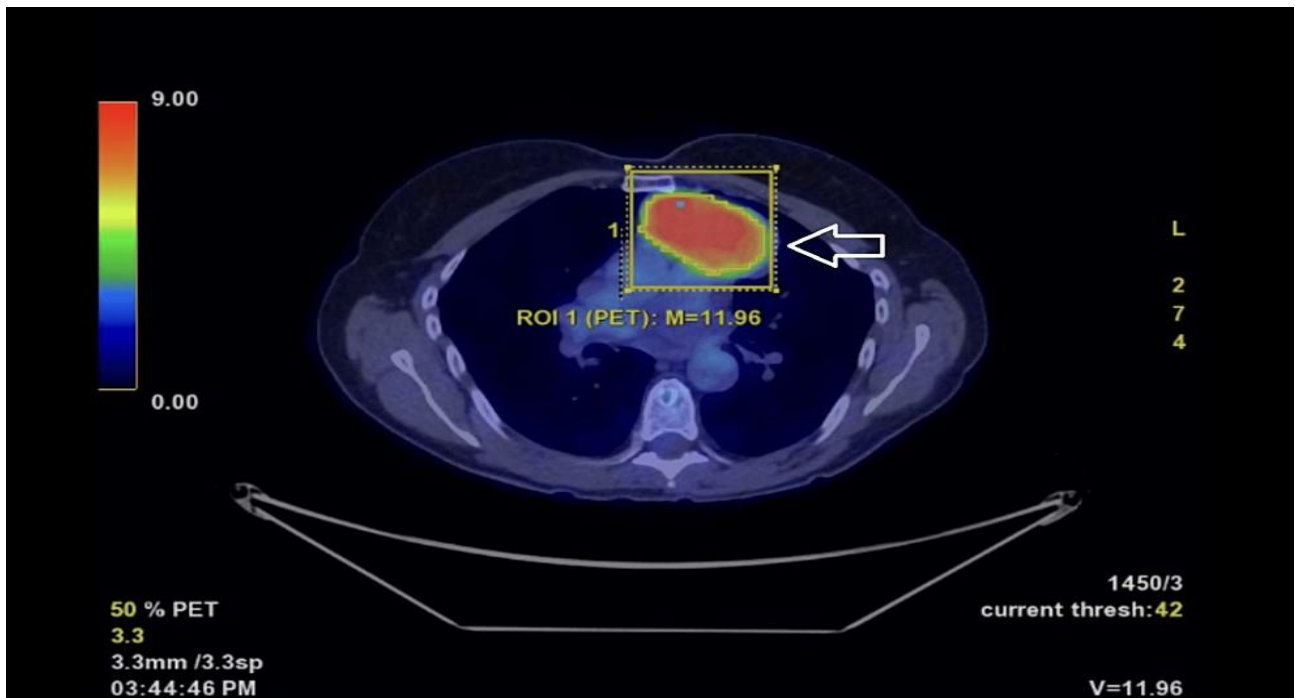


Figure 3: 72 x 42 mm well circumscribed anterior mediastinal mass is demonstrated centered at the level of the main pulmonary artery. The lesion is markedly hypermetabolic. There is an additional well- circumscribed anterior mediastinal soft tissue nodule located immediately deep to the manubrium at the level of the aortic arch, measuring 17 x 13 mm.

The patient had been treated for a positive Lyme test prior to her visiting our clinic. Based on her findings, she subsequently underwent a thymectomy. The left upper lobe ground glass opacities and the nodular left adrenal thickening, probably adenomatous, remained stable on repeat imaging. Postoperative radiation therapy was given. The patient continues to follow up in the clinic. As of one year after the resection, she has had no signs of recurrence.

Discussion

Sjögren's syndrome is a chronic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands resulting in xerostomia and xerophthalmia. The association with thymoma is well documented. It's thymic carcinoma that is less common, thymic carcinomas account for 12% of all thymic tumors [8-9]. A retrospective analysis was carried out on 112 patients from 1979 to 2003 who had attended the out-patients department at University College Hospital, London. Patients were followed from the time of primary SjS diagnosis until the endpoint of development of cancer or death. In this study, 25 patients were found to have cancer and 11 of those cases were lymphoma. The study confirmed that there is an increased incidence of lymphoma in patients with primary SjS [10]. About 5% of patients with Sjögren's syndrome eventually develop lymphomas, however thymic tumors associated with SjS are less common.

The Incidence Rate (IR) of primary SjS is 6.92 per 100,000 person-years. IR of thymic carcinoma is relatively rare, it is often accompanied by autoimmune diseases such as systemic lupus erythematosus, allergic vasculitis, and Hashimoto thyroiditis. Our case was associated with Sjogren's syndrome [11-12].

This case highlights the importance of performing a thorough evaluation of patients presenting with autoimmune disorders and Sjogren's syndrome in particular. Knowing that SjS often overlaps with other connective tissue diseases, a pretreatment evaluation is

essential. The Anti Ro52 antibodies are associated with mixed connective tissue disease-interstitial lung disease (MCTD-ILD) and the antibody titers correlate with ILD severity. The prevalence of ILD secondary to various CTD varies, in SjS it is 9-20% [13].

The confirmation of the diagnosis and the assessment of disease severity are the first steps in the workup. The diagnosis of SjS is made based on the presence of clinical features with evidence (either serologic or glandular) of autoimmunity. The Schirmer test, the presence of serologic markers (Anti SS-A with or without anti SS-B positivity) and suggestive histologic evidence (lymphocytic sialadenitis of the labial and salivary gland) can be utilized to establish the diagnosis. Assessment of disease activity and severity is gauged by the stability of the disease overtime as well as the development of new clinical findings such as fatigue, arthritis, skin manifestations, weight loss or anemia.

In this case, through taking an extensive history and a detailed physical exam we were able to orient our investigations toward an autoimmune disease and more specifically SjS. For any autoimmune disease, part of the workup will include blood tests such as complete blood count, C-reactive protein, a chemistry panel, urinalysis, antibody panel (ANA, RF, dsDNA, anti SS-A, anti SS-B, anti-citrullinated protein, complement levels). Because of the new diagnosis of Sjogren's syndrome, chest imaging was performed.

This resulted in identification of a mediastinal mass. Thymomas are associated with paraneoplastic disorders. Thymomas are believed to trigger the onset of an autoimmune disorder. One study suggests those who present with an autoimmune disorder before the diagnosis of a thymoma are more likely to have myasthenia than other disorders [14]. As the majority of data is based on nonmalignant thymic tumors, the association with thymic carcinomas is less well defined.

Currently, the treatment for Sjogren's syndrome is mainly focused on symptomatic therapies, such as artificial tears, artificial

saliva, moisturizing skin lotions. In addition to this, a systemic treatment with corticosteroids and immunosuppressants such as hydroxychloroquine, and methotrexate may be utilized based on extraglandular manifestations. The use of biologics such as rituximab may be needed, but they are not applied widely because of their significant side effects. Thymectomy may be a useful treatment in SjS with associated thymic lesions [12]. Our patient's symptoms improved after the thymectomy.

Conclusions

The workup of a suspected autoimmune disorder should include evaluation of potentially affected organs. Patients with

Sjogren's syndrome are at increased risk for extra glandular involvement including the lung, renal and neurologic systems.

Thymomas have long been associated with the development of a co-existent autoimmune disorder. Our evaluation revealed an anterior mediastinal mass and thymoma or lymphoma were suspected. Further evaluation revealed a thymic carcinoma. This case represents an uncommon presentation of thymic carcinoma and Sjogren's Syndrome. CXR should be considered in patient who are diagnosed with Sjogren's syndrome since it is a cheap and easy test that can be used to screen for thymic lesions, interstitial lung diseases as part of investigation for extra-glandular manifestations of the disease.

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