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Research Article

A rare overlap syndrome: Behcet's disease and Granulomatosis with polyangiitis

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Running title: Overlap of Behcet's disease and Granulomatosis with polyangiitis

ABSTRACT:

Behcet's disease (BD) and granulomatosis with polyangiitis (GPA) are clinicopathologic process characterized by blood vessels' inflammation. BD usually presents with recurrent oral and genitalia aphthous lesions, ophthalmic and skin involvement and it can damage to any size of blood vessels, while GPA is ANCA associated vasculitis of mainly small arteries and veins with upper and lower respiratory tracts involvement, together with glomerulonephritis. To our knowledge their overlap has not been mentioned in previous literatures till now. Here we reported a patient with clinical presentations, which perfectly fulfilled both BD and GPA criterias.

Keywords: Behcet 'disease, Granulomatosis with polyangiitis, Overlap.

INTRODUCTION:

Behcet's disease (BD) is a type of vasculitis which can involve small, medium and large vessels and usually presents with recurrent oral and genitalia aphthous lesions, ophthalmic and skin involvement. Clinical judgment and physician suspicious has the main role in the diagnosis of the BD. there is not any associating specific laboratory indexes, but HLA-B51 may be positive in a subgroup of patients [1-3].

Granulomatosis with polyangiitis (GPA) formerly known as Wegener disease, Is Fibrinoid necrotizing vasculitis of mainly small-sized arteries with granulomatosis inflammation in various organs mostly ears, nose and throat(ENT), lungs, kidneys. Large arteries are not normally involved in this disease. It is believed that the main pathogenesis of the disease is due to ANCAs specifically anti- PR3 antibodies [4, 5] .

Although some cases of ANCA-associated large-vessel involvement has been found in literature but Behcet's disease overlap with GPA has never been reported. The aim of this study is reporting

a rare overlap syndrome, and its manifestations and the way we approach the patient [2, 6].

Case report:

A 25-year-old male referred to the Rheumatology clinic because of pyoderma gangrenosum, petechiae and purpura on his left shin beside the history of constitutional symptoms, recurrent oral, genitalia and skin aphthous lesions for one year, therefore he was admitted for further evaluation of Behcet's disease. The oral lesions were consist of recurrent, multiple and major aphthae, on his gingiva, tongue and buccal areas which healed spontaneously.

The place of genital aphthae was on his scrotum beside the scars of past lesions. Skin aphthae and pseudofolliculitis were detected in perineal and perianal areas. The patient was also suffering from morbid obesity with a body weight=170 kg and BMI=39.5. He had ESR of 100, and CRP of 95Although HLA-B5 was negative. Biopsy of the skin lesions showed small vessel leukocytoclastic vasculitis.

During the BD workups specific history clarifies the presence of a severe headache, recurrent bloody nasal discharge, hemoptysis, cough and resistant sinusitis since one year before admission, and dyspnea FC4. Further evaluation revealed hematuria, proteinuria and casts in urine analysis and positive anti PR3ANCA with the titer of 25 with the upper limitation of 0.5. CXR and lung spiral HRCT showed multiple cavities and nodules in both lungs.

Pansinusitis also detected in PNS spiral CT scan (Fig1).

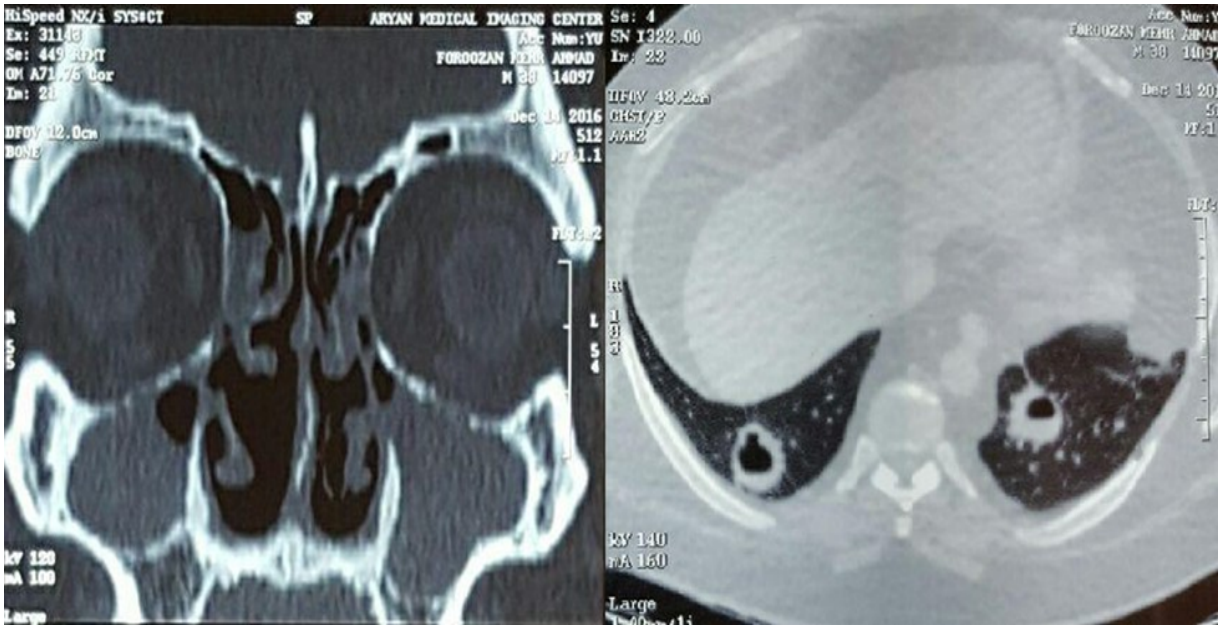

Fig. 1: Lung spiral HRCT showed multiple cavities and nodules in both lungs. Pansinusitis also detected in PNS spiral CT scan.

The patient perfectly fulfilled ICBD/revised ICBD/Persian gulf criteria for Behcet's disease besides ACR/ Emma and revised ACR/ Emma criteria for GPA.

According to multi-organ involvements he was candidate for Induction therapy with high dose of steroid and monthly cyclophosphamide but Due to morbid obesity combined with significantly rose in AST and ALT level (3 times upper than normal limit) and Sonography compatible with fatty liver grade 4, we started maintenance therapy with moderate steroid dose and azathioprine.

Then he went under laparoscopic sleeve gastrectomy which was drastically effective and lowered his weight to 130kg and AST and ALT level to 35(40), thereafter we started the induction therapy which led to symptom relief, decreasing of acute phase reactants and remission of cavities in CXR (Fig2).

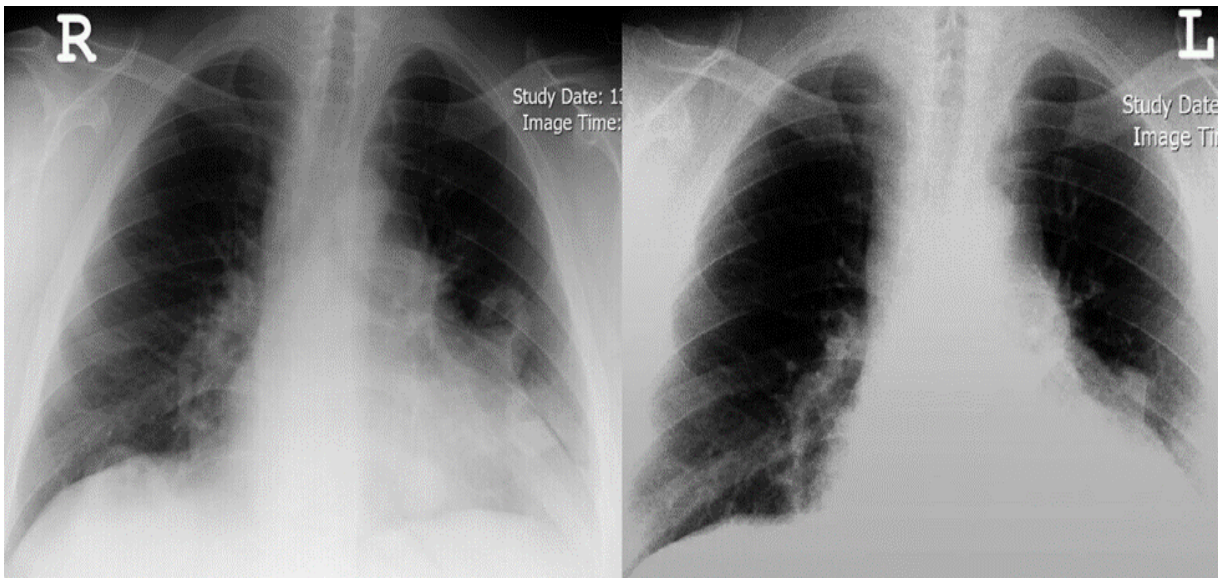

Fig. 2: Chest X Ray before and after the treatment

DISCUSSION:

To our knowledge this patient is the first case of GPA/BD overlap. Both BD and GPA are vasculitides with multi-organ involvements. BD can damage all vessels of any size, whereas GPA is mostly vasculitis of small sized vessels. BD mostly presents with recurrent oral and genitalia aphthous lesions, eye inflammation and skin involvements, hence rarely reported sinusitis and lung cavities or nodules, in contrast GPA is the anti-PR3 associated vasculitis and mostly present with ENT, lung and kidney involvement [7-9]. Here we reported a patient with recurrent, multiple oral, genital and skin aphthous lesions and pseudofolliculitis which perfectly fulfilled ICBD/revised ICBD/Persian gulf criteria for Behcet's disease besides ACR/EMA and revised ACR/EMA criteria for GPA by pansinusitis, nasal ulcer, bloody nasal discharge, cavities and nodules in CXR and lung HRCT scan, hematuria, proteinuria and positive anti PR3-ANCA [3, 5, 7-11]. Although in this patient oral aphthae and pyoderma gangrenosum can be explained by GPA, but the concurrence of oral, genital and skin aphthous lesions and pseudofolliculitis are purely suggestive of Behcet's disease[9] . In conclusion, our patient is the case of GPA/BD overlap, which has not been reported in previous articles. although while approaching to patients' signs and symptoms, we commonly expect to have one kind of disease for each patient, but sometimes there may be overlap syndromes, in which patients fulfill criteria for two or even more diseases at the same time. Therefore, in the diagnostic field, various etiologies should be kept in the mind, in this way overlap syndromes will be diagnosed, which may influence the treatment approach and effect on patients' morbidity and survival.

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