A patient with ankylosing spondylitis and hemoptysis: is there a hidden disease?

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Background An overlap syndrome of Behçet's disease and ankylosing spondylitis is a rare autoimmune disease of connective tissue in which a patient presents with symptoms of two diseases.

Case presentation We report a case of non-smoking 25 years old known to have ankylosing spondylitis presented by haemoptysis due to a cause not usually associated with his primary disease leading to the search for another cause.

Conclusion The coexistence of Behçet's disease and ankylosing spondylitis may be encountered in clinical practice, although it is rare. If there is a resistance to the conventional treatments, TN-alpha blocking agents may be an alternative therapeutic option in these diseases.

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Case presentation

A 25-year-old male, nonsmoker, was referred to the rheumatology department 2 years ago complaining of neck pain and decreasing range of movement of the cervical spine. The pain was more prominent in the early morning and was relieved by exercise. He was diagnosed as having ankylosing spondylitis, confirmed by positive HLA-B27. His father was previously diagnosed clinically and serologically to have the same disease.

Cytotoxic therapy in the form of corticosteroid, cyclophosphamide, and colchicines was started and continued for 3 months with no much notable improvement; hence, he discontinued the medication by himself. After 3 months, the patient started to show symptoms such as fever, fatigue, weight loss, and arthralgias along with recurrent painful oral and genital ulcers, papulopustular skin lesions on his trunk and limbs for which nonspecific topical analgesics and steroid were applied. After 1 month, the patient developed frank hemoptysis for which he sought medical advice at our department.

On admission, he denied any other chest complaints.

On physical examination, he was found to be vitally stable with five oral aphthous ulcers (Fig. 1) and two genital ulcerative lesions. A spine examination revealed marked limitation of both the cervical and low back. Ophthalmologic examination was considered to be normal. Local chest examination was unremarkable.

Laboratory results showed a high sedimentation rate of 110 mm/h and C-reactive protein level of 16.75 mg/l; other routine laboratory tests such as complete blood count and urinalysis were all within normal limits. The D-dimer test was negative. Sputum examination using Ziehl–Neelsen stain showed negative result for acid–fast bacilli.

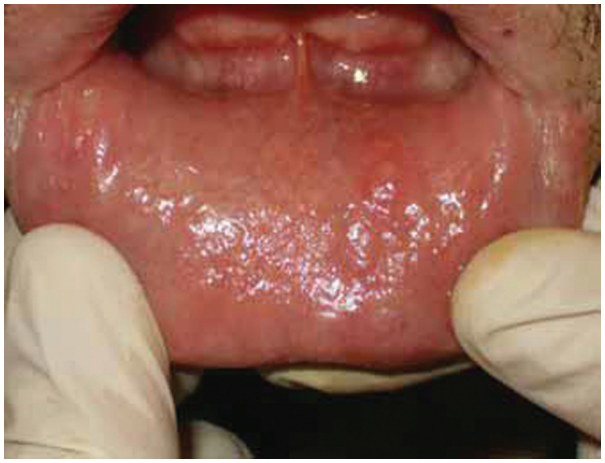
Chest radiograph showed right hilar shadow. Multislice computed tomography of the chest with intravenous contrast was performed (Fig. 2) revealing right pulmonary artery aneurysm with pulmonary artery wall thickening, which may be related to either vasculitis or thrombosis. Transthoracic echocardiography revealed estimated pulmonary artery pressure of 55 mmHg.

He was diagnosed as having coexisting ankylosing spondylitis and Behçet's disease (overlap syndrome). We began treatment with pulse steroid and pulse cyclophosphamide and colchicines and continued for 3 months with improvement in oral and genital ulcerations; hemoptysis stopped but there was no improvement in pain. Hence, he was transferred to the physical medicine and rehabilitation department for physiotherapy to alleviate stiffness and pain.

Comment

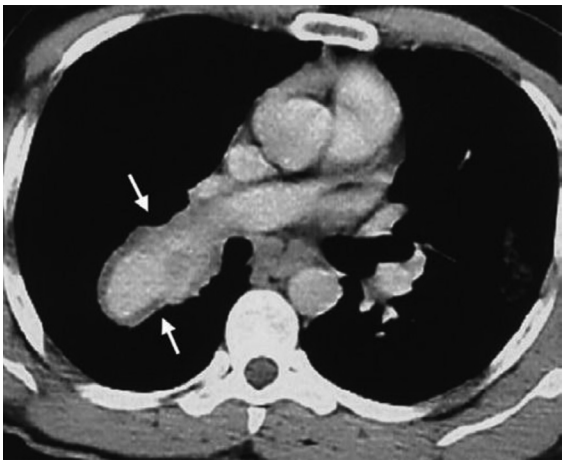
An overlap syndrome is an autoimmune disease of connective tissue in which a patient presents with symptoms of two or more diseases. Diagnosis depends on the diseases for which the patient shows symptoms and for which the patient has positive antibodies in laboratory serology. Although the prevalence of coexisting Behçet's disease and ankylosing spondylitis is much debated, it has been reported in few studies.

Fig. 1



Typical oral aphthous ulcers in a common site.

Fig. 2



Contrast-enhanced computed tomography scan of the chest showing pulmonary artery aneurysm.

Ankylosing spondylitis is a connective tissue disease of unknown pathogenesis [1]. Young male individuals aged 20–40 years are the most commonly affected; 90% of them are HLA-B27-positive, with male : female ratio of 10 : 1 [2]. The reported prevalence of pulmonary disease ranges from 0 to 30%, including apical interstitial lung disease with or without apical fibrobullous disease, pleural diseases (such as pleuritis, exudative pleural effusion, apical pleural thickening, and pleural calcifications), thoracic cage immobility, and hemoptysis, which may result from apical cavitary disease or rarely capillaritis. Other clinical manifestations include peripheral arthritis, enthesitis (inflammation of tendons and ligaments), and extra-articular organ involvement [1].

Behçet's disease is a multisystem disease characterized by a triple-symptom complex consisting of genital ulcer, aphthous stomatitis, and iritis, frequently accompanied by vasculitis [1]. Behçet's disease was reported to have a strong association with HLA–B51 rather than with HLA–B27; however, the number of reported patients with coexisting Behçet's disease and ankylosing spondylitis (overlap syndrome) has increased. The role of proinflammatory cytokines such as tumor necrosis factor–α (TNF–α) and genetic factors is important in the pathogenesis of both diseases [3].

Reports concerning the coexistence of these two diseases are found rarely in the studies. Dubost *et al*. [4] reported three patients with coexisting Behçet's disease and ankylosing spondylitis among 11 patients with Behçet's disease. We found increased erythrocyte sedimentation rate and C–reactive protein levels, suggesting that the patient was in the active phase in terms of both diseases. Similar to our patient, ankylosing spondylitis may coexist with Behçet's disease [5,6]. Therefore, this coexistence should be kept in mind. TNF–α blocking agents are commonly used in the treatment regimens for ankylosing spondylitis [7], which was not given to the patient in our study because of unavailability.

Conclusion

The coexistence of Behçet's disease and ankylosing spondylitis may be encountered in clinical practice, although it is rare, and this condition should be kept in mind. If there is a resistance to the conservative treatments, TNF-α blocking agents may be an alternative therapeutic option in patients with these diseases.

Acknowledgements

Conflicts of interest

None declared.

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