

# Adalimumab in a Relapsed Vascular Behcet's Disease

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## INTRODUCTION

Behcet's disease is a rare chronic relapsing multisystem inflammatory disease with variable size vessel vasculitis. The first line treatment for vascular Behcet's disease is high dose corticosteroid and cyclophosphamide. Anti-Tumor Necrosis Factor alfa (TNF- $\alpha$ ) therapy is recommended for refractory/relapsed vascular Behcet's disease. However, there is limited evidence of adalimumab use as compared to Infliximab.

Here we report a case of vascular Behcet's disease with a relapsed after a course of pulsed cyclophosphamide which was then effectively controlled with Adalimumab.

## REPORT

A previously well 20-year-old male, presented with 2 months history of prolonged fever, recurrent orogenital ulcers, constitutional symptoms, reduced effort tolerance accompanied with a day of hemoptysis. Diagnosis of Behcet's disease was made with constellation findings of venous thrombosis (bilateral pulmonary embolism, a large right ventricular thrombus (Figure A), arteritis (bilateral pulmonary artery saccular aneurysms), orogenital ulcers, positive pathergy test, high inflammatory markers, with exclusion of other autoimmune diseases and infection.

Hence, pulsed Methylprednisolone and intravenous (IV) Cyclophosphamide (CYC) were commenced immediately. He was then discharged with prednisolone 1mg/kg/day with treatment dose of Enoxaparin. Repeated imaging after 4 doses of IV CYC revealed resolved right ventricular thrombus with residual pulmonary embolism and smaller pulmonary aneurysm (Figure B).

He was later presented with hemoptysis after 5<sup>th</sup> dose of IV CYC in which coiling of left descending pulmonary artery aneurysm was done. Hence, pulsed IV CYC course were extended to total of 8 doses (Total cumulative dose: 6.76g). He was clinically improved in which then switched to azathioprine as maintenance therapy.

However, a relapsed occurred 6 weeks after the last dose of IV CYC. He had thrombosis of left popliteal vein and Positron Emission Tomography scan revealed active vasculitis of left popliteal artery extending distally to left peroneal and left posterior tibial arteries. He was given pulsed methylprednisolone and initiated on subcutaneous Adalimumab 40mg biweekly. Since then, the disease was well controlled. He was able to taper down prednisolone to 5mg daily after 4 months of Adalimumab without any relapse.

## DISCUSSION

Behcet's disease is characterized by neutrophilic inflammation with T-cell dysregulation resulting in autoinflammatory process. TNF- $\alpha$  plays a central pro-inflammatory role in the pathogenesis of Behcet's disease by driving the excessive immune response seen in this autoinflammatory vasculitis.

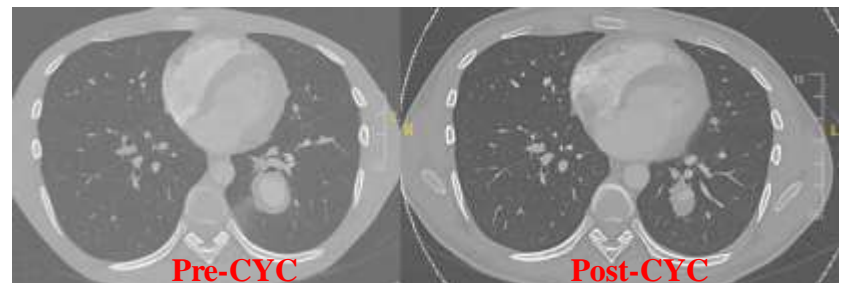
Emerging therapy like anti-TNF- $\alpha$  (Infliximab and Adalimumab) therapy offers new promises in effectively treating especially severe and refractory Behcet's cases. While most current data involves the use of Infliximab in refractory cases, evidence on Adalimumab is rather limited (mainly derived from case series or observational studies).

Adalimumab is a fully human monoclonal antibody targeting TNF- $\alpha$ , a key cytokine involved in the inflammation of Behcet's Disease. Advantage of Adalimumab over Infliximab includes offering patient greater convenience of self-administering subcutaneous injection at home and Adalimumab may have a lower risk of antibody development due to its less immunogenicity.

Our patient is a case of vascular Behcet's disease who had relapsed post IV CYC. His disease was responded well to Adalimumab treatment and able to taper prednisolone to low dose without any relapse.



**Figure A:**  
ECHO showed large right ventricular thrombus



**Figure B:**  
CTPA showed left descending pulmonary artery aneurysm  
(Pre CYC size 2.9cm x 2.5cm x 2.8cm and Post 4 doses CYC size 1.0cm x 1.2cm x 1.2cm)

## CONCLUSION

Adalimumab is effective in treatment of relapsed vascular Behcet's disease and can be used as maintenance therapy. However, further studies will be needed to confirm the efficacy of Adalimumab in relapse prevention.

## REFERENCES

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