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RESEARCH ARTICLE

COMPLETE REMISSION OF MULTIPLE SEVERE AND REFRACTORY PULMONARY ANEURYSMS IN BEHCET'S DISEASE AFTER TREATMENT WITH INFLIXIMAB

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Abstract

Introduction: Behcet's disease is a multisystem disorder that can affect multiple organs.Vascular complications are common in Behçet's disease.Arterial complications can include pulmonary artery aneurysms. These have a high risk of rupture and can be life threatening.

Observation: This report details a case of Behçet's Disease in a 52-year-old patient of North African descent. The diagnosis was made after the patient experienced massive hemoptysis caused by multiple pulmonary artery aneurysms. The initial treatment prescribed for the aneurysm involved immunosuppressive medication, including cyclophosphamide and corticosteroids. During a 6-month follow-up period, the patient showed an unfavorable progression. Treatment with infliximab was attempted and resulted in a successful response to this treatment.

Conclusion: This is a new observation that reinforces the superiority of infliximab in severe and refractory Behcet's disease.

Introduction:-

Behçet's disease (BD) is a systemic vasculitis of an origin that is still unknown. BD can affect many organs, including vascular involvement[1]. Both veins and arteries can be damaged, with a clear predominance of venous involvement. The pulmonary artery aneurysms are the most common site of arterial involvement in the disease.[2]In fact, it is a very serious condition that can be life-threatening in as many as 50% of cases.Treatment of BD is still based on the administration of immunosuppressive drugs. Recently, thetumour necrosis factor-α (TNF-α) blockers have provided additionaltools for the treatment of resistant disease.[3]

We report the case of a North African patient with a severe, fatal and refractory form of BD treated with infliximab. The patient achieved a complete drug-induced remission.

Case Observation:

A 52-year-old man with a history of deep vein thrombosis of the left lower limb, presented to the emergency department in May 2020 with a large volume of hemoptysis, estimated at 500 ml, without any other external bleeding. The patient was initially admitted to intensive care as his hemodynamic status was compromised.

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The patient then underwent a chest angioscanning, which revealed multiple aneurysms of the two pulmonary branches, 13 in all, one of which was large and thrombosed, located in the right lower lobe.**[Figure 1, 2]**

The patient's past medical history revealed recurrent oral aphthae once a month since childhood and 3 episodes of genital aphthosis. Clinical examination demonstrated oral aphthosis on the inside of the lower lip and genital aphthosis, scarred lower lip and tongue, scrotal aphtha and painful aphtha at the urethral orifice. The rest of the examination was unremarkable. In particular, there was no signs of neurological or ocular involvement.

The diagnosis of Behçet's disease with revealing of severe vascular involvement and inaugural bipolar aphthosis was based on the 2013 international criteria.

The treatment was a conventional strategy of cyclophosphamide in combination with high doses of glucocorticoids, monthly for 6 months, based on the presence of vascular involvement, followed by Azathioprine.

In November 2020, the patient was re-admitted to the emergency department with a recurrence of a massive hemoptysis at a rate of 3 episodes, 1 month after the 6th bolus, with once again a commitment of vital prognosis, a conditioning was made with a transfusion of 4 red blood cells, after stabilisation of the hemodynamic state, a chest CT angiography was repeated, showing no changes compared to the initial one.

The surgical team was assigned to the case, but the surgical pathway proved to be challenging.

With his informed consent, we started intravenous infliximab therapy at a dose of 3 mg/kg at weeks 0, 2 and 6 and every 2 months subsequently.

After the first 3 infusions, the evolution was marked by the disappearance of hemoptysis, with the persistence of bloody cough, but minimal. Hemoglobin levels became normal.

During the 18-month follow-up scan of infliximab, all aneurysms disappeared, with the exception of the large thrombosed one, which was greatly reduced in size.**[Figure 3]**

It is considered that biological therapy is effective in vascular involvement, including arterial aneurysms.

Discussion:-

Behçet's disease is ubiquitous, but there are two main, the disease is more common in the eastern Mediterranean and Japan, with prevalences ranging from in Turkish Anatolia, the disease's prevalence is 350/100,000 inhabitants.In Japan, the disease's prevalence is 30/100,000 inhabitants.In Germany and Great Britain, the disease's prevalence ranges from 2.5 to 0.5/100.000.[4]

Behçet's disease is characterised by recurrent oral and genital aphthosis, but can also include ocular, neurological, articular and vascular involvement. Behçet's disease is frequently associated with vascular manifestation, representing around 40% of patients, mostly in a form of deep venous thrombosis, although arterial involvement is much rarer than venous involvement.[1]

The disease predominantly affects males, mostly those between their second and third decade of life. Our patient presented with

Traditional epidemiological characteristics such as gender, age and North African origin.

Arterial involvement may be present at the time of diagnosis. [5,6] But typically takes place 8-10 years of disease progression.
The dominant revealing sign is haemoptysis. Rarely, pulmonary arterial aneurysms are asymptomatic and discovered incidentally.Our patient's disease was revealed by pulmonary artery aneurysms.

The severity of the situation is illustrated by an international study, which found that the mortality rate for patients with pulmonary aneurysms was 75%. [7]

Managing pulmonary aneurysms is challenging and often inconclusive. Surgery is the most radical option, especially when there is a single or limited number of lesions.

However, our patient's case was more complicated because of the presence of a large number of aneurysms. Because of the difficulty of the surgical approach, the indication for medical treatment based on cyclophosphamide and methylprednisolone was posed.

The current protocol used by our group includes three 1 g pulses of methylprednisolone followed by prednisolone 1 mg/kg/day, tapered and if possible stopped over 6 months. Intravenous cyclophosphamide – 1 g – is given monthly for 6 or 12 months, then, if remission is maintained, switched to azathioprine, 2.5 mg/kg/day [8].

The short-term effects of the anti-TNF monoclonal antibody infliximab have been reported in several case reports and small case series, Preliminary results strongly suggest that infliximab is remarkably effective in inducing short-term remission of almost all manifestations of the disease [9] However, its use in the treatment of pulmonary vasculitis with aneurysms has not been reported so far.

The results of infliximab therapy were particularly important in our patient by minimizing the life risk and not needing to engage in major surgery. He has had longlasting remission of therapy with infliximab.

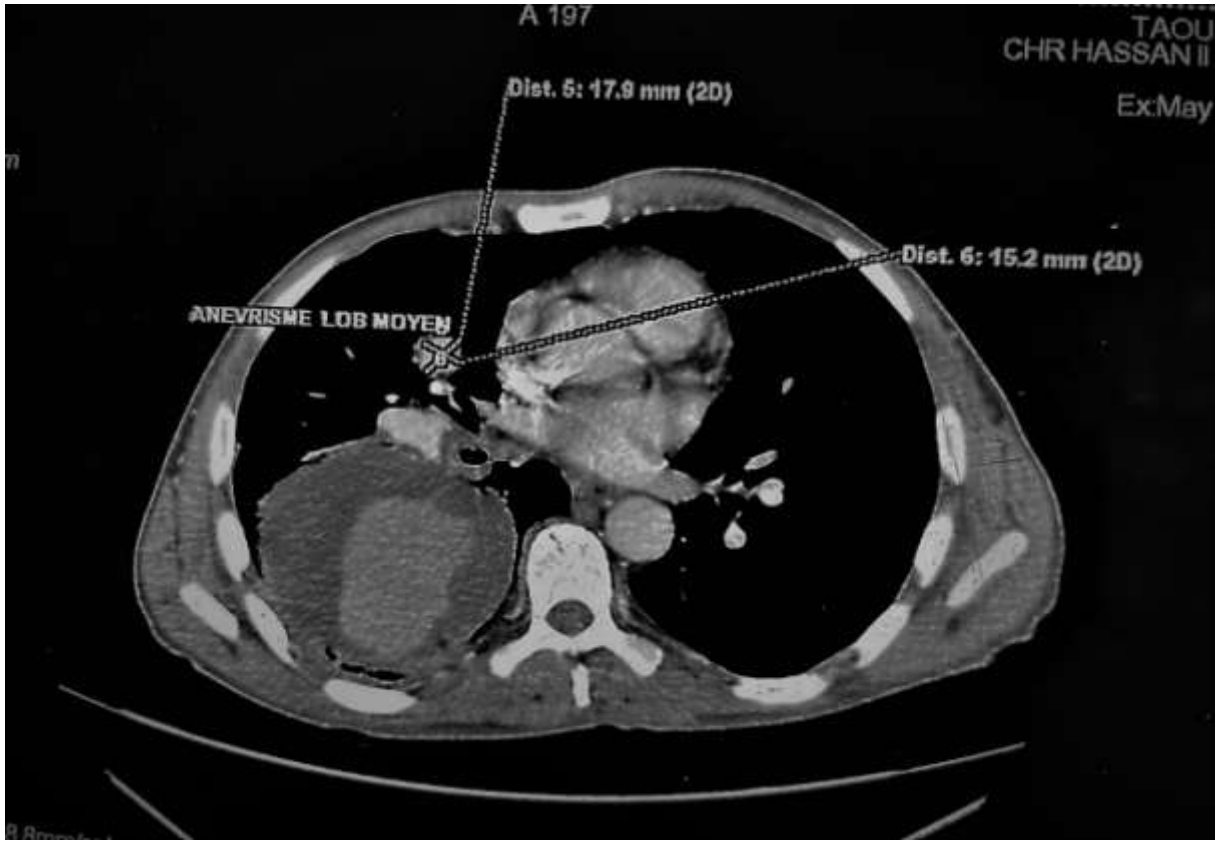

Figure1:- Aneurysm of middle lobe.

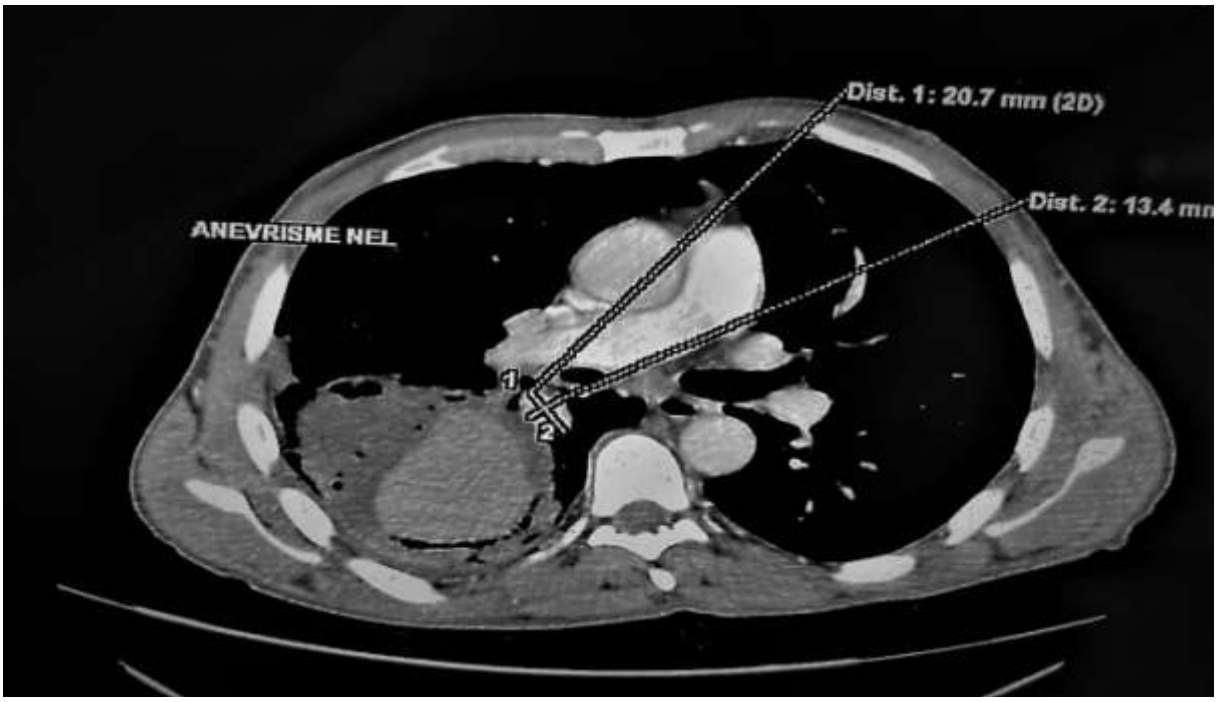

Figure 2:- Nelson aneurysm.

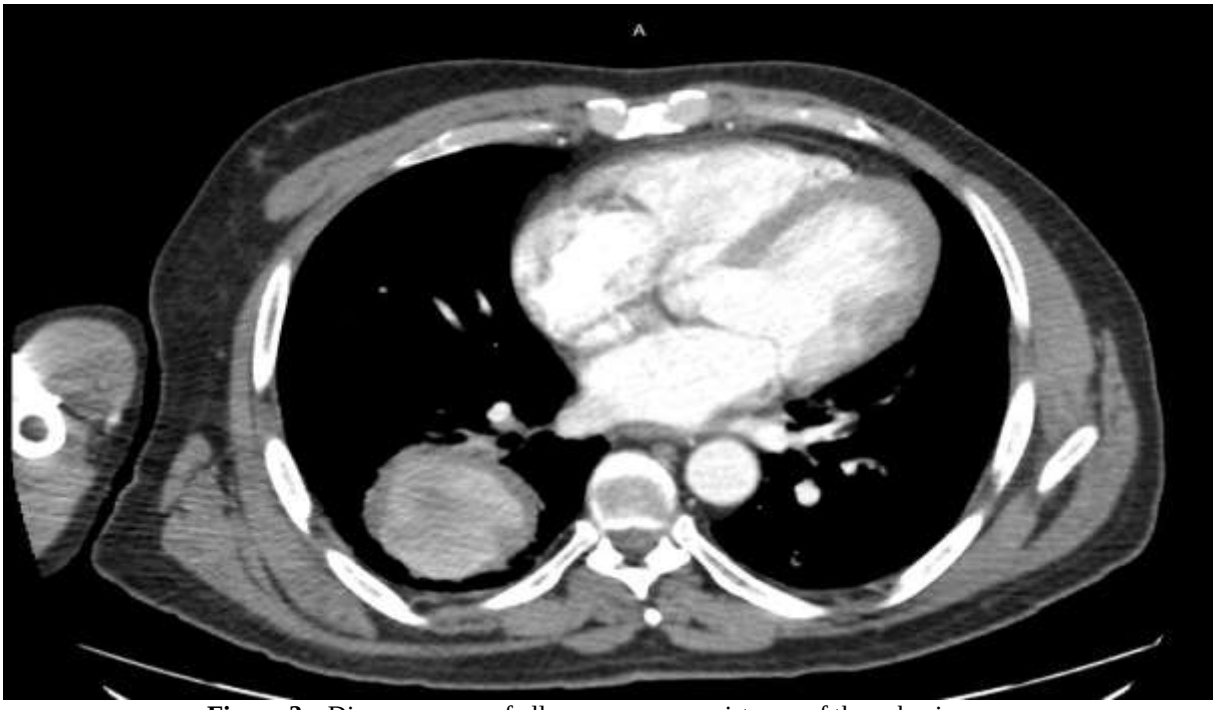

Figure 3:- Disappearance of all aneurysms, persistence of thrombosis one.

Conclusion:-

Pulmonary artery aneurysms are an important complication of Behçet's disease with a particularly poor prognosis. Knowledge of them is essential, mainly because of the seriousness of the prognosis.Our case also suggests that treatment with infliximab is very successful in such severe impairment.

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