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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/15041
DOI URL: http://dx.doi.org/10.21474/IJAR01/15041

RESEARCH ARTICLE

HYPEREOSINOPHILIC SYNDROME WITH INTRACARDIAC THROMBUS PRESENTING WITH EMBOLIC EVENTS

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Manuscript Info

Manuscript History
Received: 15 May 2022
Final Accepted: 18 June 2022
Published: July 2022

Key words:-
Embolic Events, Ventricular Thrombus, Hypereosinophilic Syndrome

Abstract

Introduction: Hypereosinophilic syndrome (HES) is a rare disorder characterised by idiopathic eosinophilia with organ system involvement. Its major tissue target is the heart, assuming diverse aspects that affect its different tunics with a varied clinical and echocardiographic expression. Cardiac involvement is frequently associated with ventricular thrombosis, portending a poor prognosis as it can be complicated by acute embolic events, along with the progressive development of restrictive cardiomyopathy, valvular dysfunction, and heart failure.
Case Report: We report the case of a 62 years old woman, who presented one month ago a pulmonary embolism, and was admitted for an ischemic stroke related to a ventricular thrombus in the context of a hypereosinophilia syndrome.
Conclusion: Embolic manifestations secondary to cardiac involvement may be a mode of revelation of HES. The mortality related to cardiac damage is high, hence the need for appropriate management.

Introduction:-

Hypereosinophilic syndrome (HES) is defined as a blood hypereosinophilia higher than 1500/ml for more than six months and for which the etiological investigation is negative.

It is therefore a diagnosis of elimination that should be considered only after having excluded the etiologies of secondary hypereosinophilia (including parasitic or viral infections, allergic diseases, drug-induced or chemical-induced eosinophilia, hypoadrenalism) and clonal eosinophilia, with evidence of organ involvement, and absence of phenotypically abnormal and/or clonal T lymphocytes (1,2).Cardiac and thromboembolic manifestations of HES are the common causes of mortality and morbidity. Among the patients with HES, the frequency of cardiac manifestations is 40%-50% (2).

We describe here a case of a patient who presented one month ago a pulmonary embolism, and was admitted for an ischemic stroke related to cardiac thrombus in a context of HES.

Case report

A 62-year-old woman, who presented one month ago with a moderate pulmonary embolism that had not been investigated. She was admitted to the department for dyspnea with alteration of the general state before developing neurological signs.

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On clinical examination, the heart rate was at 115 beats per minute without any additional murmur, arterial pressure at 100/60 mmhg, oxygen saturation at 92%, with right hemiplegia and aphasia. The rest of the clinical examination was unremarkable. The EKG showed a regular sinus rhythm, accelerated to 105bpm with a right bundle branch block aspect and diffuse repolarization abnormalities.

The biological workup showed an elevated eosinophilic polynuclear count of 27180/ml.

A bone marrow biopsy showed a eosinophilia with 47% eosinophils and no evidence of abnormal myeloid maturation.

Transthoracic echocardiography revealed the presence of a huge apical thrombus in the right ventricle and a less obvious one in the left ventricle. The ejection fraction was preserved without major valvular abnormalities at this stage.

The brain scan came back in favor of an ischemic stroke in the territory of the left sylvian artery. The thoracic CT scan, which had shown a distal pulmonary embolism, was not repeated.

The patient was put on anticoagulant therapy and bolus of corticosteroids followed by a progressive degression. The evolution was favorable with clinical improvement and regression of the PNE level from 27180 to 250/ml.

Discussion:-

Hardy and Anderson had reported three cases of hypereosinophilia associated with hepatosplenomegaly and pulmonary or cardiac symptoms in 1968. Based on the analysis of these and other more recent observations, four criteria were proposed by Chusid and al. and are still valid today (3): The patient must have a hypereosinophilia greater than 1500/mm3 for more than six months, associated with visceral lesions in the absence of any cause of hypereosinophilia(1).

The initial presentation can be variable, from flu-like symptoms to acute cardiac or neurological complications. Cardiac involvement can evolve through three stages: Necrosis, thrombosis and fibrosis (4) :
- The first phase corresponds to necrosis of the endocardium and myocardial infiltration by lymphocytes and eosinophilic micro abscesses and is observed in the first five weeks (5)
- The second phase corresponds to the formation of intraventricular thrombi along the injured endocardium and occurs on average after ten months of evolution
- The last phase is characterized by myocardial fibrosis responsible for restrictive cardiomyopathies and mitral and/or tricuspid valve regurgitation

This chronology is not always respected, since cases of acute necrotizing eosinophilic myocarditis have been described.

Our patient presented at the second stage of the evolution, with thrombi formation and before the installation of a restrictive cardiomyopathy or major valvular damage, with as main mode of revelation a cerebral and pulmonary embolic accident:

- Neurological complications are common in HES and can be of three types.

The first is the embolic stroke, often of cardiac origin may originate from the left ventricle or, more rarely, in the case of persistent foramen ovale, from the right ventricle. In some cases, local intravascular thrombosis may develop in the cerebral vessels, although the mechanism is not well identified.

Embolic or transient cerebral vascular accidents may be multiple and recurrent, and can develop even before cardiac involvement is identified on echocardiography and may thus be the first mode of revelation of HES. It should be noted that recurrence of embolisms in properly anticoagulated patients is not uncommon (6,7).

The second and the third types of neurological complications are the primary central nervous system dysfunction and the peripheral neuropathies.

- Concerning the pulmonary complications which represent 40% (4), both emboli from right ventricular thrombi and congestive heart failure may be responsible for pulmonary involvement, which may also be the result of primary eosinophilic infiltration of the lungs.

Pulmonary fibrosis develops over time, mostly in patients with cardiac fibrosis.(8)

Regarding the therapeutic aspect, the products most commonly used are hydroxyurea, corticosteroids and interferon a (9).Corticosteroids, including prednisone at doses of 20 to 60 mg/day (10), used in the early stages prevents the risk of fibrosis, whereas anticoagulants are indicated for thrombotic complications.

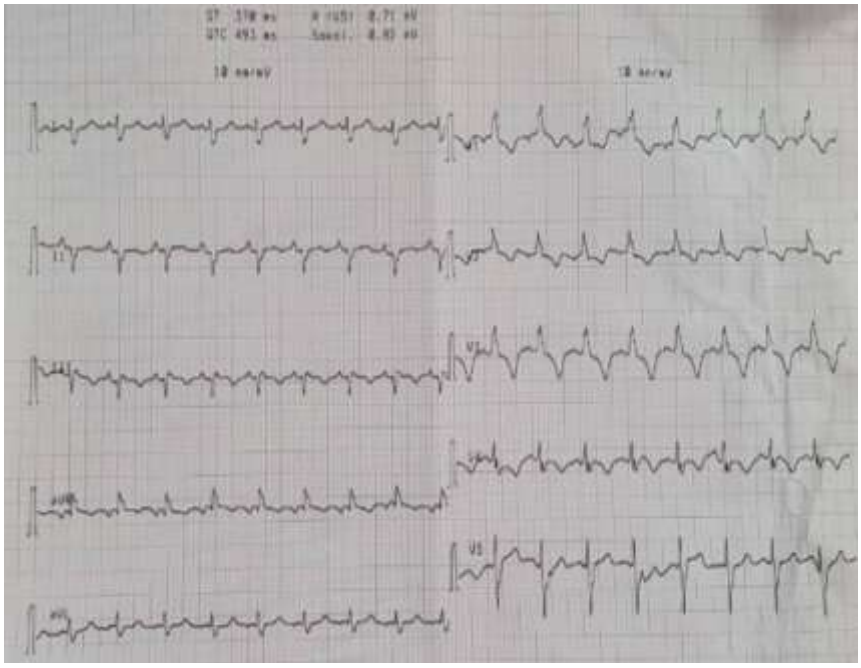

Figure 1:- ECG on admission with sinus tachycardia and appearance of right bundle branch block

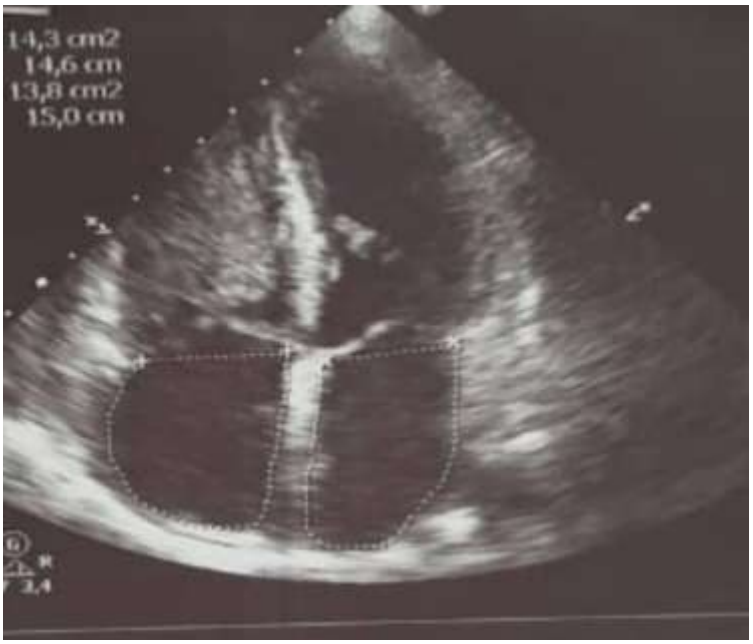

Figure 2:- Echocardiographic image in apical 4-chamber showing right and left intraventricular thrombus.

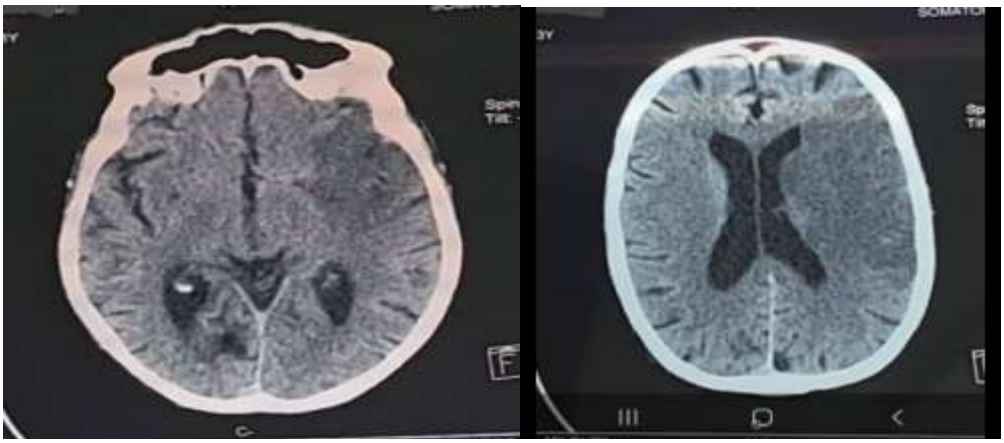

Figure 3:- CT scan brain in favor of a left sylvianischemic stroke.

Conclusion:-

Depending on the stage of the disorder, two-dimensional echocardiography can be used to visualize cardiac thrombi. The prognosis is related to the stage of the disease and not to the eosinophil count. Cardiac manifestations including ventricular thrombi are responsible for several embolic manifestations and increase the risk of morbidity and mortality in HES.

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