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Case report

Heterogeneous aetiology and clinical presentation of cardiac involvement in hypereosinophilic syndrome: A case series

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ABSTRACT

Introduction: Hypereosinophilic syndrome (HES) is a heterogeneous group of diseases defined by marked hyperesonofilia (1.5×10^9/l) that persists more than 6 months with organ infiltration and damage. Cardiac involvement in HES is common and is associated with significant morbidity and mortality. To illustrate contemporary diagnostic and therapeutic methods, we reviewed three cases of HES with cardiac involvement, recently diagnosed in our institution.

Description of cases: We present a series of three cases of HES with cardiac involvement and clinical presentation in the form of intracardiac thrombosis. One of these cases was caused by FIP1L1-PDGFRA positive myeloproliferation and successfully treated with imatinib. All cases received corticosteroids and oral anticoagulation with vitamin K antagonists with favourable clinical outcomes.

Conclusions: Although HES with cardiac involvement is a rare condition, it may have severe clinical consequences and needs to be diagnosed early. Subsequent important step is careful differential diagnosis of hypereosinophilia and its targeted treatment.

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Abbreviations: HES, hyperosinophilic syndrome; HE, hyperesonophilia; LV, left ventricle; RV, right ventricle; CMRI, contrast enhanced magnetic resonance imaging.

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Introduction

Hypereosinophilic syndrome (HES) is a rare disease with an incidence of 0.35 cases per 1 000 000 inhabitants and year [1]. Definition of HES requires presence of severe hypereosinophilia (>1.5 × 10^9/l) lasting more than 6 months without any evidence for secondary aetiology and presence of organ damage caused by infiltration of tissues with eosinophils. The aetiology of HES could be very heterogeneous (Table 1). It may be related to clonal production of eosinophils in hematologic diseases, to reactive release of eosinophils in several clinical conditions or it remains unexplained (idiopathic) [1]. Among organ systems, HES affects most commonly lungs, heart, gastrointestinal system and peripheral nerves. Cardiac involvement in HES was first described in 1936 by Loeffler as ''fibroblastic parietal endocarditis with mural thrombi and blood eosinophilia'' and since then is referred to as Loeffler's endocarditis. However, HES can present also as isolated eosinophilic myocarditis [1]. According to pooled data, cardiac involvement occurs in 20% of patients with HES [2]. Cardiac involvement is the major cause of morbidity and mortality in HES with estimated survival of 42% at 10–15 years of follow-up [3–5]. We reviewed three cases of HES with cardiac involvement which have been recently diagnosed in our institution. This case series should illustrate contemporary diagnostic and treatment methods in this condition.

We performed a retrospective analysis of relevant clinical, laboratory and imaging data in three individuals with cardiac involvement in HES (Table 2). The diagnosis of HES was based on the proposed criteria of the Working Conference of Eosinophil Disorders and Syndromes (Vienna, Austria; 27–28 May 2011) [6]. We obtained a written informed consent from the patients to present their cases.

Table 1 – Aetiology of hypereosinophilia.

Primary (clonal): stem cell, myeloid or eosinophilic neoplasm
- PDGFRA (platelet derived growth factor alpha)
- PDGFRB (platelet derived growth factor beta)
- FGFR1 (fibroblast growth factor receptor 1)
- BCR-ABL (chromosome fusion gene in Philadelphia chromosome)
- Chronic eosinopholic leukaemia – not otherwise specified

Secondary (non-clonal, reactive)
1. Malignancies (acute lymphoblastic leukaemia, T-cell lymphoma, certain solid tumours, etc.)
2. Infections (esp. parasitic)
3. Allergy/hypersensitive disease
4. Connective tissue disorders (e.g. Churg–Strauss syndrome)
5. Skin disease (atopic dermatitis, urticaria, eczema, Gleich syndrome)
6. Tropical endocardial fibrosis
7. Metabolic abnormalities (adrenal insufficiency)
8. Immune system disease (e.g. Wiskott–Aldrich syndrome)

Idiopathic HES

Clinical cases

Case 1. A 49-year-old male patient without any prior cardiovascular disease was admitted for subacute occlusion

Table 2 – Clinical and laboratory characteristics of cases.

	Case 1		Case 2		Case 3	
	First visit	Last FU (24 months)	First visit	Last FU (12 months)	First visit	Last FU (48 months)
Clinical presentation	Claudication, peripheral embolism (lower limb)	Arthralgia and chest pain	Pulmonary infiltrates, systemic inflammation			
Eosinophil	7.96 × 10^9/l	1.35 × 10^9/l	2.5 × 10^9/l	0.74 × 10^9/l	27.2 × 10^9/l	0.22 × 10^9/l
(abs/%)	55.3%	17.8%	23%	10.9%	63%	2.3%
FIP1L1-PDGFRA	Present		Absent		Absent	
Hs Troponin T (ng/l)	<10		90		1134	
LVEF (%)	60	60	60		40	
Intracardiac thrombosis	RV apex	RV apex	LV apex	LV apex/ lateral wall	None	LV apex
Valve disease	Mobile aortic valve structures	Fibrotic changes with new aortic stenosis	None	None	None	None
Signs of RCMP	No	No	No	No	No	No
Other thoracic/ abdominal manifestation	None		None		Pulmonary and GIT inflammation	Peripheral embolism
Treatment	Corticosteroids + imatinib + anticoagulation	Corticoids + anticoagulation	Corticoids + anticoagulation			
Final diagnosis	Chronic eosinophilic leukaemia	Idiopathic HES	Idiopathic HES			

Abbreviations: FU, Followup; GIT, gastrointestinal; HES, hypereosinophilic syndrome; LVEF, ejection fraction of left ventricle; RCMP, restrictive cardiomyopathy.

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