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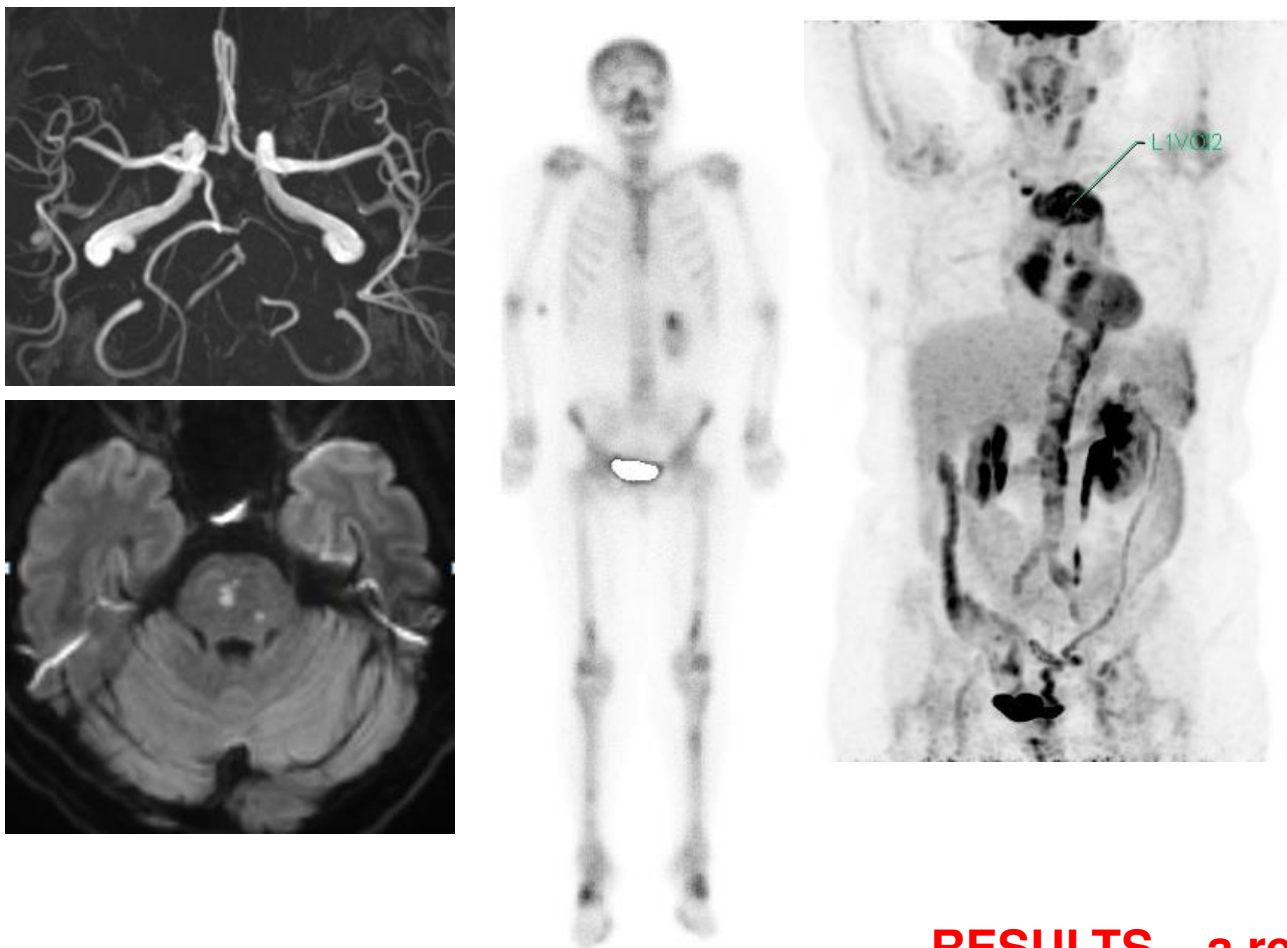
## INTRODUCTION

Erdheim-Chester disease (ECD) is a rare, non-Langerhans form of histiocytosis of unknown origin, which was first described by Jakob Erdheim and William Chester in 1930.<sup>1</sup> This multisystemic disease is currently considered as an inflammatory myeloid neoplasia.<sup>2,3</sup> Central nervous system (CNS) involvement is present in up to 50% of ECD cases and represents a strong prognostic factor and an independent predictor of death in patients with ECD.<sup>4-6</sup> Central nervous system manifestations of ECD are usually related to infiltration of the brain parenchyma by abnormal cells, with a predominance of lesions occurring in the posterior fossa.<sup>5,7,8</sup> Stroke is an exceptional manifestation of this disease.

We aimed to describe cerebrovascular diseases in patients with ECD.

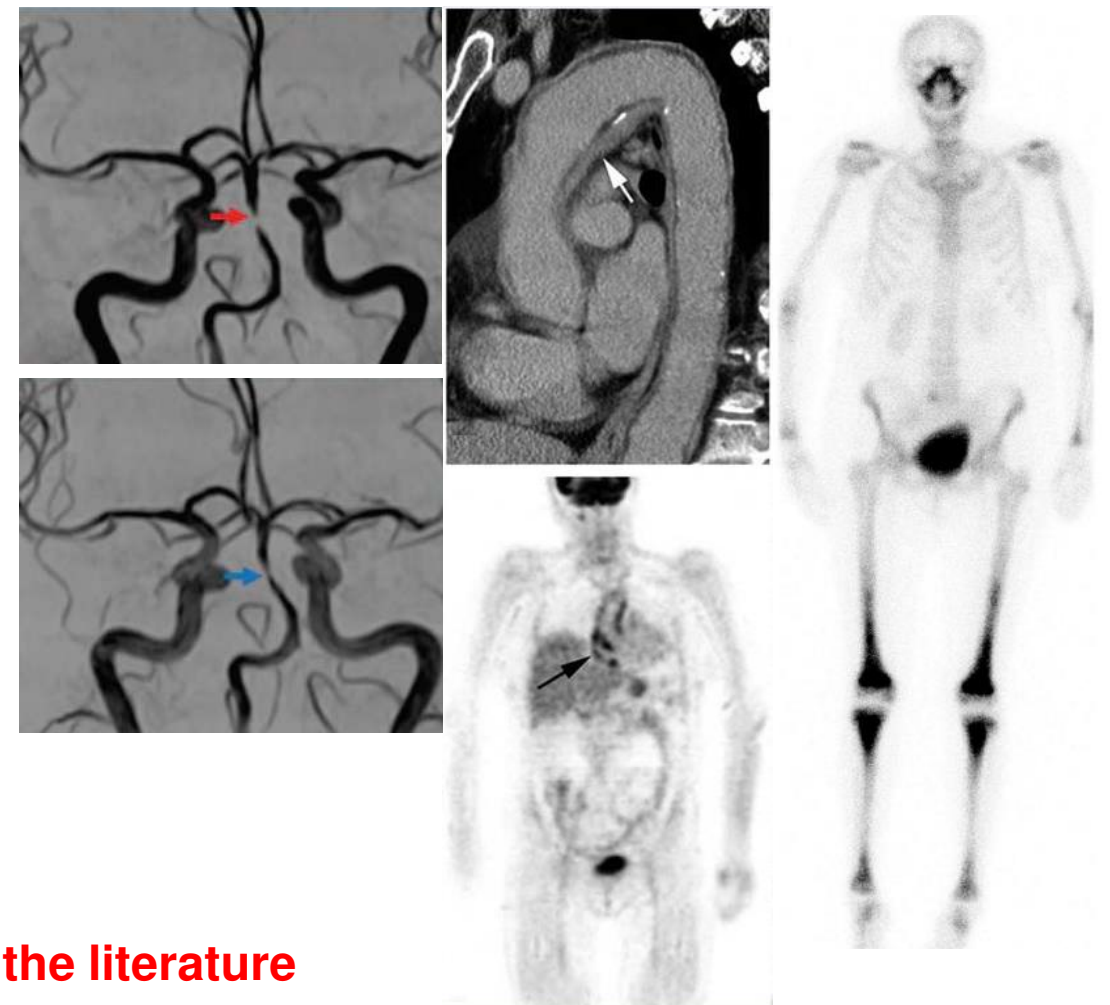
### Patient 1

We report the case of 71-year-old woman with aortic and renal involvement who was hospitalized for recurrent TIAs and a vertebrobasilar stroke secondary to infiltration and severe stenosis of the basilar artery as well as infiltration of other intracranial and extracranial arteries. She stabilized after cobimetinib and steroid treatment.



### Patient 2

We also report the case of a 59-year-old woman who presented with a vertebrobasilar stroke secondary to infiltration and severe stenosis of the basilar artery, who clinically improved after interferon-alpha therapy. We performed a review of the relevant literature and reported the few other cases described.



## RESULTS – a review of the literature

Including our patients, we have found only 8 cases of cerebrovascular disorder in ECD. Most of them had supra-aortic artery involvement.

Patients with stroke and Erdheim-Chester disease							
Reference	Sex	Age of diagnosis of ECD and Stroke	Stroke topography	Supra-aortic vascular involvement	Other Vascular involvement	Cardiovascular risk factors	Other features
Mergancova et al, 1998 [10]	F	68	Vertebrobasilar stroke	Basal brain arteries	Aorta Coronary arteries		The patient died The diagnosis of ECD was established on autopsy
Gauvrit et al, 2004 [11]	M	41	Transient ischemic attack	Left carotid artery	No		Transient monocular blindness of the left eye
Ameziane et al, 2009 [12]	F	59	Focal right precentral infarction	No	No		Right hemiparesis due to left nodular lesion of the brain Asymptomatic stroke
Choi et al, 2013 [13]	F	69	Transient ischemic attack	Carotid arteries	No	Arterial hypertension Type 2 diabetes	Recurrent monocular blindness of the left eye
Fargeot et al, 2014 [14]	F	68	Right middle cerebral artery infarction	Carotid arteries	Aorta Renal arteries	Type 2 diabetes	
Mélé et al, 2015 [15]	F	83	Three strokes in the middle cerebral artery (2) and vertebrobasilar territories (1)	Carotid artery	Aorta		Mixed Langerhans and non-Langerhans histiocytosis in the same patient
Our observation [9]	F	59	Bilateral cerebral posterior infarction	Basilar artery	Aorta Renal arteries	Arterial hypertension	Dide-Bolcazo syndrome
Our observation [unpublished]	F	71	Vertebrobasilar transient ischemic attacks and stroke	Basilar artery, middle cerebral artery, carotid artery	Aorta	Arterial hypertension	

## CONCLUSIONS

Acute ischemic stroke and TIA are rare complications of ECD, which are mainly due to infiltration and stenosis of the cerebral arteries.

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