European Journal of Case Reports in Internal Medicine

Do We Have a Culprit? An Association of Giant Cell Arteritis with Pulmonary Embolism

Carolina Miguel Gonçalves[1], Pedro Neves Tavares[1], Fátima Saraiva[1], João Morais[1,2], Maria Jesus Banza[1]

[1] Centro Hospitalar de Leiria, Leiria, Portugal

[2] ciTechCare – Center for Innovative Care and Health Technology, Polytechnic of Leiria, Leiria, Portugal

Doi: 10.12890/2022_003028 - European Journal of Case Reports in Internal Medicine - © EFIM 2022

Received: 02/11/2021
Accepted: 10/12/2021
Published: 31/01/2022

How to cite this article: Gonçalves CM, Neves Tavares P, Saraiva F, Morais J, Banza MJ. Do we have a culprit? An association of giant cell arteritis with pulmonary embolism. *EJCRIM* 2022;**9**: doi:10.12890/2022_003028.

Conflicts of Interests: The authors declare there are no competing interests.

ABSTRACT

Giant cell arteritis is the most common type of systemic vasculitis. An increased risk of venous thromboembolism has been described in these patients. We report the case of a 79-year-old woman with a history of polymyalgia rheumatica, who presented with left thoracic pain radiating to the neck and scapula plus temporal headache. She had no changes on physical examination, but work-up tests showed increased D-dimer levels and computed tomography pulmonary angiography revealed signs of a chronic/subacute embolism in the right inferior lobe. Anticoagulation with edoxaban was started after 5-day bridging with enoxaparin. Three weeks after the initial diagnosis the headache still persisted and she developed scalp tenderness. Giant cell arteritis was diagnosed and treated with prednisolone, with complete resolution of symptoms. Extensive diagnostic work-up was performed to identify an alternative cause of pulmonary thromboembolism; however, the investigations were negative. This case supports the hypothesis that this type of vasculitis could be related to the occurrence of pulmonary embolism.

LEARNING POINTS

- An increased risk of thromboembolism has been associated with giant cell arteritis.
- Early diagnosis is crucial; however, the role of antiplatelets or anticoagulants is not yet established.

KEYWORDS

Giant cell arteritis, pulmonary embolism

CASE DESCRIPTION

A 79-year-old woman presented to our hospital with a 4-hour history of left thoracic pain, radiating to the neck and scapula, as well as headache. She had a past medical history of polymyalgia rheumatica (PMR), treated with 10 mg of prednisolone, obstructive sleep apnoea, arterial hypertension and obesity.

The patient was haemodynamically stable and no change on physical examination was found. Electrocardiography was unremarkable. Blood tests showed increased D-dimer levels (1196 ng/mL) and computed tomography (CT) pulmonary angiography revealed signs of a chronic/subacute embolism in the right inferior lobe. Her Simplified Pulmonary Embolism Severity Index score was 0 and based on the European Society of Cardiology (ESC) guidelines [1] had a low mortality risk.

She was admitted to hospital for etiologic study and management of pulmonary embolism (PE). Further work-up tests were negative, such as thrombophilia testing, leg ultrasonography, thyroid and breast ultrasonography, and thoracic, abdominal and pelvic CT. Based on ESC guidelines [1], our patient was started on 5-day bridging with enoxaparin before edoxaban 60 mg daily was introduced. No intercurrent disease was recorded and she was discharged home after 6 days on 5 mg of prednisolone, with no recurrence of thoracic pain.

European Journal of Case Reports in Internal Medicine

Approximately 3 weeks after the initial diagnosis, in a routine rheumatology outpatient visit, she complained of headache, located over her left temple, and it was observed that she had developed pain and stiffness of her shoulders and hip girdle as well as scalp tenderness. She had no visual disturbances. Based on the 2016 revised American College of Rheumatology (ACR) criteria[2], a diagnosis of giant cell arteritis (GCA) was made. She was subsequently started on high dose prednisolone (60 mg daily). She was reviewed 1 week later, with complete resolution of symptoms noted and the slow tapering of the prednisolone dose indicated.

DISCUSSION

Giant cell arteritis (GCA), or temporal arteritis [3], is characterised by granulomatous lesions of medium and large sized arteries [3,4], more frequently involving the temporal artery but also the aortic arch, iliac, axillary and femoral arteries[3]. With an incidence ranging between 10 and 29 cases per 100 000 person-years and a prevalence of 8–10% per 100 000 persons[3], it is the most common type of vasculitis in adults[4]. The peak onset is in the seventh decade of life, being rare in patients under 50 years[3] and most commonly affecting women[2].

Early diagnosis of GCA is of the utmost importance [3], as this type of vasculitis is associated with several complications, such as blindness, cerebrovascular accident, aortic aneurysm, myocardial infarction [3,4], neuropathy, and vestibular and hearing deficiencies [3]. GCA is associated with PMR in 50–60% of patients, while 16–21% of patients with PMR have a current GCA[3], as in our patient's case.

Temporal artery biopsy is considered the gold standard diagnostic test [2,3]. Nevertheless, it is an invasive test [2] that has 10–15% false negative results [3]. The 2016 revised ACR criteria suggest that the presence of three or more points out of 11 establish a GCA diagnosis[2]. Our patient scored 5/11 points: older than 50 years; absence of exclusion criteria; new headache (1 point); PMR (2 points); anaemia (1 point); and tender temporal artery (1 point). An elevated erythrocyte sedimentation rate (ESR) (59 mm) should be ignored because of PMR[2].

An increased risk of venous thromboembolism (VTE), namely PE and deep vein thrombosis, has been described in GCA [3,4] and is independent of other risk factors, age and sex [4]. This risk is increased throughout the first year of GCA diagnosis and several mechanisms could account for it: stasis, hypercoagulability, and endothelial damage [4,5]. These mechanisms involve impaired mobility, inflammatory endothelial dysfunction, upregulation of procoagulants, downregulation of anticoagulants and fibrinolysis due to systemic inflammation, myointimal thickening, stenosis of vessels and thrombocytosis [4]. The role of glucocorticoids in contributing to the risk of VTE is controversial [4]. Nevertheless, the risks of VTE warrant further research [4], as contradictive results in another retrospective study showed a similar prevalence of VTE in GCA patients and the general population [6].

As our extensive diagnostic work-up to identify an alternative cause of PE was negative, GCA is a probable cause for the occurrence of PE in our patient. Although prophylaxis for these events may be reasonable [3,5], the use of antiplatelets or anticoagulants in the GCA population needs further research [4,5]. While low dose aspirin is used for cardiovascular prevention in these patients [3,5], its efficacy is lower in VTE [5]. Some studies propose the use of anticoagulation in high risk patients[5], whereas others report it as controversial [3].

A good prognosis can be expected in GCA with a similar mortality rate to the general population [3] and indeed our patient recovered completely.

In conclusion, the risk of VTE appears to be increased among GCA patients and further research is required to verify these findings, as well as to investigate the role of antiplatelet and anticoagulant therapies in these patients.

REFERENCES

1. Konstantinides SV, Meyer G, Becattini C, Bueno H, Geersing GJ, Harjola VP, et al. 2019 ESC Guidelines for the diagnosis and management of acute pulmonary embolism developed in collaboration with the European Respiratory Society (ERS). *Eur Heart J* 2020;**41**(4):543–603.
2. Sait MR, Lepore M, Kwasnicki R, Allington J, Balasubramanian R, Somasundaram SK, et al. The 2016 revised ACR criteria for diagnosis of giant cell arteritis – our case series: can this avoid unnecessary temporal artery biopsies? *Int J Surg Open* 2017;**9**:19–23.
3. Guida A, Tufano A, Perna P, Moscato P, De Donato MT, Finelli R, et al. The thromboembolic risk in giant cell arteritis: a critical review of the literature. *Int J Rheumatol* 2014;**2014**:806402.
4. Aviña-Zubieta JA, Bhole VM, Amiri N, Sayre EC, Choi HK. The risk of deep venous thrombosis and pulmonary embolism in giant cell arteritis: a general population-based study. *Ann Rheum Dis* 2016;**75**(1):148–154.
5. Novikov P, Makarov E, Moiseev S, Meshkov A, Strizhakov L. Venous thromboembolic events in systemic vasculitis. *Ann Rheum Dis* 2015;**74**(3):e27.
6. Lo Gullo A, Koster MJ, Crowson CS, Makol A, Ytterberg SR, Saitta A, et al. Venous thromboembolism and cerebrovascular events in patients with giant cell arteritis: a population-based retrospective cohort study. *PLoS One* 2016;**11**(2):e0149579.