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Y. Emad¹ · Y. Ragab² · A. El-Marakbi³ · A. Saad⁴ · O. Ibrahim⁵ · A. Abd-Elhalim⁶ ·
H. El-Santawi⁶ · J. J. Rasker⁷

¹ Rheumatology Department, Faculty of Medicine, Cairo University, Cairo, Egypt

² Radiology Department, Faculty of Medicine, Cairo University, Cairo, Egypt

³ Vascular Surgery Department, Faculty of Medicine, Cairo University, Cairo, Egypt

⁴ Internal medicine Department, Faculty of Medicine, Cairo University, Cairo, Egypt

⁵ Morecambe Bay University Hospitals Lancaster, Lancashire, UK

⁶ Vascular Surgery Department, Dr.Erfan and Bagedo General Hospital, Jeddah, Saudi Arabia

⁷ Faculty of Behavioral, Management and Social Sciences, Department Psychology, Health and Technology, University of Twente, Enschede, The Netherlands

A case of Hughes–Stovin syndrome (incomplete Behçet’s disease) with extensive arterial involvement

Unmasking the true face of a rare syndrome

Introduction

Hughes–Stovin syndrome (HSS) was named after two British physicians, Drs. John Patterson Hughes and Peter George Ingle Stovin. They described the syndrome in two male patients with deep venous thrombosis and segmental pulmonary artery aneurysms in 1959 [1]. The current consensus is that HSS results from a vasculitis similar to that in Behçet’s disease (BD). Several investigators have suggested that HSS is actually a variant of BD rather than a discrete clinical entity [2–4]. However, the link between HSS and BD remains speculative because the etiologies of both disorders are still uncertain. The association stems from the observation that HSS is occasionally seen as a feature in BD and may even be the presenting manifestation of the syndrome, but HSS lacks the classical triad of BD (oral ulceration, genital ulceration, and eye disease) [2]. In this report we describe a young man with the features of HSS presenting with extensive arterial involvement of the legs as well as extensive venous thrombophlebitis and pulmonary arterial aneurysms with in situ thrombosis.

Case presentation

A 35-year-old male patient presented in our emergency clinic with acute onset of bilateral lower limb pain and features of acute ischemia more evident on the left side with absent pulsations of the dorsalis pedis and popliteal arteries on the left side and absent dorsalis pedis artery pulsation on the right. The patient gave a past history highly suggestive of ischemic claudication of both lower limbs, especially on the left side, and low-grade fever. He also had a history of recurrent thrombophlebitis of the lower limb veins for 1 year. Urgent power Doppler study of both lower limb veins showed marked luminal attenuation of the great saphenous veins bilaterally, with thickened wall and areas of non/partial compressibility denoting underlying phlebitis without active thrombosis or venous occlusion. The patient was admitted to our facility for further assessment and investigations. Initial laboratory investigations showed; ESR first hour 44 mm/h, CRP 4.3 mg/dl, Hgb 11.7 gm/dL, WBC count (8.8), platelet count (197,000), normal values for the factor V Leiden mutation gene, factor II-

prothrombin mutation, normal homocysteine level (12.36 mcmol/L; normal range 6.26–15.01 mcmol/L), normal protein C, and normal protein S values. Immunological profile showed negative ANA, no anti-DNA antibodies, negative anti-cardiolipin antibodies (IgG and IgM), negative P-ANCA, and C-ANCA. Normal D-dimer levels (1.01 mg/L) and normal procalcitonin levels (0.04 ng/ml) were found. Urgent computed tomography (CT) angiography for the aorta and arterial trees of the lower limbs demonstrated segmental occlusion of the superficial femoral artery with collateral refilling of the popliteal artery. In addition, acute diffuse thrombosis was seen affecting the distal 1/3 of the abdominal aorta and left common iliac artery with refilling of its distal 1/3 via collateral circulation (■ Fig. 1). Furthermore, the left anterior and posterior tibial arteries were markedly attenuated, denoting occlusive disease along their courses, apart from distal short segmental collateral refilling of the distal left posterior tibial arterial segment with poor distal runoff (■ Fig. 1). The patient was put on anti-coagulation therapy with sc heparin necessitated by the critical

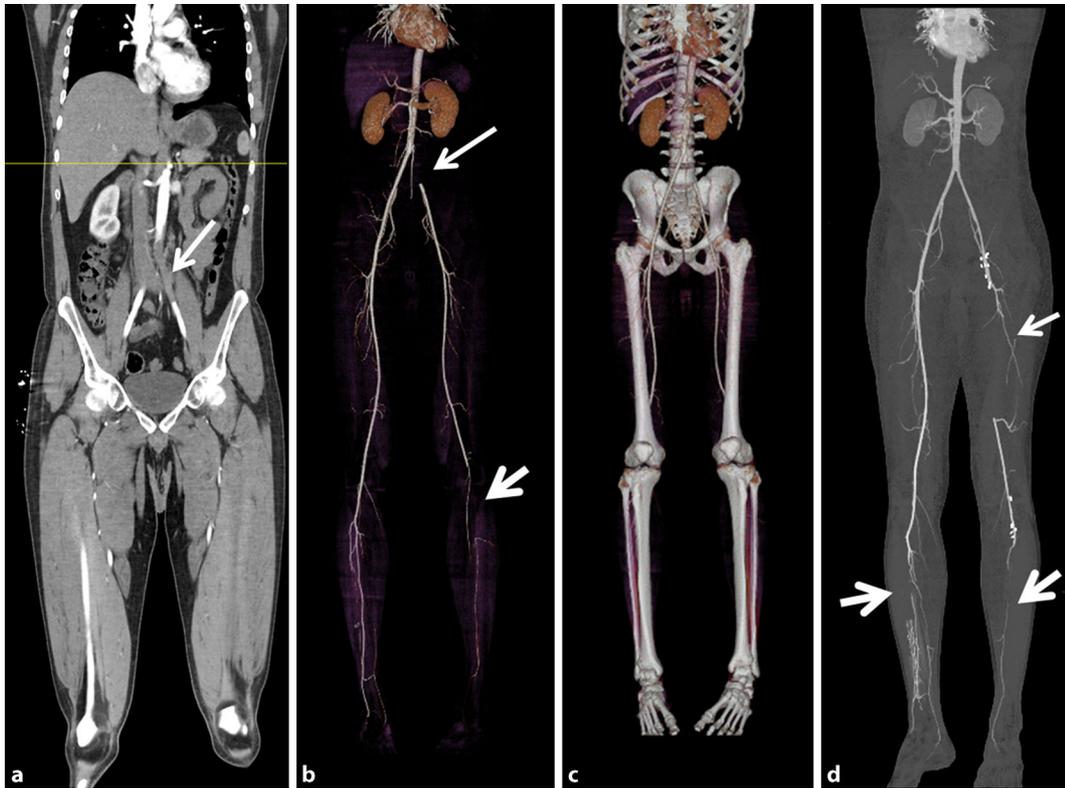


Fig. 1 ◀ CT angiography showing the aorta and lower limb arterial tree (a,b), aorta, and left common iliac thrombosis (white arrow); c CT angiography (3D) with bony skeleton for level localization; d CT angiography post-operative with recent occlusion of the superficial femoral artery (proximal 2/3), an occlusion of the left leg arteries denoting poor distal runoff (short thick arrows) and also interrupted right leg arteries are also seen (short thick arrow)

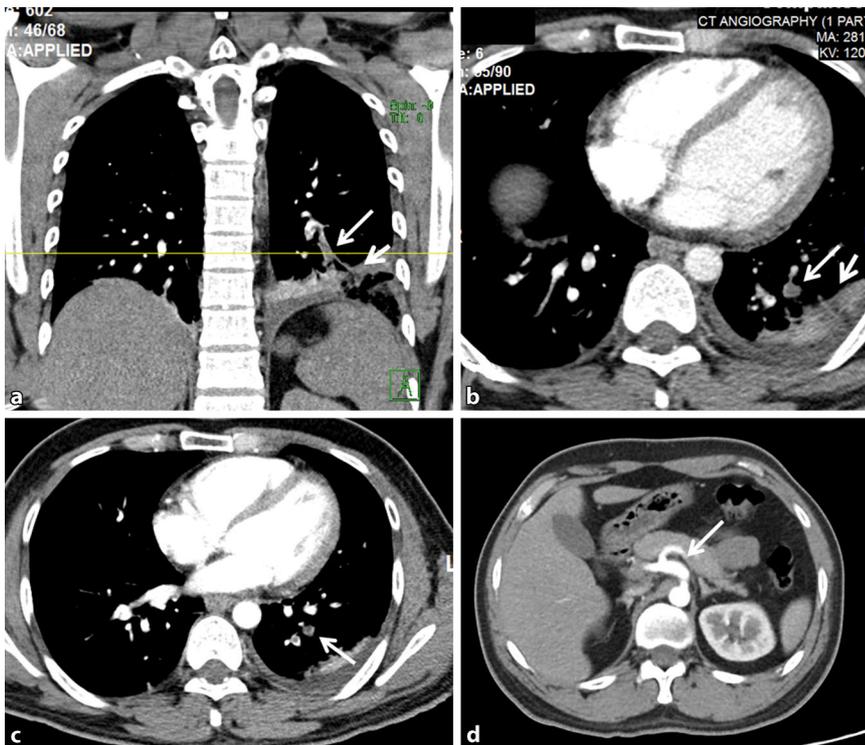


Fig. 2 ▲ Pulmonary CT angiography (mediastinal window) showing a–c left lower lung lobe arterial ectasia and in situ thrombosis (white arrows) and left basal pulmonary infarction and pleural effusion (short thick arrows); d celiac trunk aneurysmal dilatation shortly after its origin (white arrow)

ischemia, and after 1 week developed a cough and mild respiratory distress without concomitant hemoptysis. Pulmonary CT angiography was ordered and revealed filling defects in the left pulmonary lower lobar segmental and sub-segmental arterial branches with associated left lung lower lobe consolidation (■ Fig. 2) and celiac trunk aneurysmal dilatation shortly after its origin (■ Fig. 2d). After rheumatological consultation and taking into account the critical ischemia of the left lower limb, the patient underwent urgent vascular intervention in the form of left lower limb angioplasty and aorto-iliac, popliteal, and tibio-peroneal thromboembolectomy, and received post-operative anti-coagulation with warfarin. The diagnosis of HSS was made with extensive venous and arterial affection as there were no findings indicating Behçet's disease (BD). The arterial as well as venous affection and the pulmonary CT angiography showing filling defects represent in situ thrombosis, as there was no deep vein thrombosis (DVT) that might have caused pulmonary thromboembolism. Eye investigation by a specialist excluded

uveitis and the patient gave no history of recurrent and/or mouth ulcers.

Shortly after, the patient started with pulse methylprednisolone therapy (1000 mg/day) iv bolus infusion for 3 consecutive days, followed by pulse cyclophosphamide 750 mg/iv bolus infusion over 30 min with ample hydration. Later, the patient was scheduled to receive pulse cyclophosphamide 750 mg on a monthly basis for at least 1 year. Oral prednisolone 40 mg/day was started after the pulse steroid therapy. The patient showed much improvement with no ischemic insults during the 6-month period of follow-up without any reported new ischemic events and the oral prednisolone dosage could be tapered.

Discussion

Although the exact etiology and pathogenesis of HSS is unknown, the current consensus is that vasculitis is the primary pathologic process underlying HSS [5]. The population-based incidence of HSS cannot be exactly determined. It usually affects young adults, especially males [6, 7]. Being an extremely rare disease, there are no formally described diagnostic criteria or pathognomonic laboratory investigations for this syndrome.

Generally, HSS is characterized by thrombophlebitis and multiple pulmonary aneurysms associated with in situ thrombosis, as in our case [5, 8]. Thus, if a patient presents with this set of findings and the clinician is able to exclude other causes, the patient has either HSS or BD. However, BD can be ruled out by its classic distinctive features (eye inflammation, recurrent mouth and genital ulceration) which are absent in HSS. This is how HSS was diagnosed in the majority of the case reports in the literature [4].

We performed a PubMed search of the available HSS case reports with typical and atypical vascular manifestations and also case reports where it is discussed whether HSS is an outcome of BD or a separate clinical entity; findings are summarized in [Table 1](#).

Recurrent phlebitis in HSS frequently involves the large vessels, resulting in thromboembolism, with reports of

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Y. Emad · Y. Ragab · A. El-Marakbi · A. Saad · O. Ibrahim · A. Abd-Elhalim · H. El-Santawi · J. J. Rasker

A case of Hughes–Stovin syndrome (incomplete Behçet’s disease) with extensive arterial involvement. Unmasking the true face of a rare syndrome

Abstract

Hughes–Stovin syndrome (HSS), characterized by the combination of multiple pulmonary artery aneurysms and deep vein thrombosis, is a rare and an under-recognized clinical entity with less than 40 published cases in English medical literature. Vascular venous thrombotic events, as occurring in the course of Behçet’s disease (BD), are also described in HSS, e. g., vena cava, intracardiac, jugular vein, iliac vein, femoral vein, and dural sinus thrombosis. We describe a 35-year-old man with HSS showing classical features of the syndrome in the form of recurrent thrombophlebitis of the lower limb veins, pulmonary arterial aneurysms, and left lower limb ischemia with extensive arterial tree involvement. The patient presented with critical arterial ischemia in the left lower limb together with aortic and left common

iliac artery thrombosis, occlusion of the left superficial femoral artery, and occlusion of both lower limb arteries. Urgent vascular surgeries were carried out for limb salvage. Shortly after, the patient started on pulse corticosteroid/cyclophosphamide therapy, followed by monthly cyclophosphamide for 1 year, with much improvement. We discuss arterial involvement in HSS and similarities of HSS and BD regarding thrombotic events. We summarize the current management options of HSS.

Keywords

Hughes–Stovin syndrome · Incomplete Behçet · Arterial vasculitis in Hughes–Stovin syndrome · Aortitis · Thrombophlebitis · Pulmonary artery aneurysms

Hughes-Stovin-Syndrom (unvollständige Ausprägung des M. Behçet) mit extensiver arterieller Beteiligung. Das wahre Gesicht eines seltenen Syndroms

Zusammenfassung

Das Hughes-Stovin-Syndrom (HSS) ist durch die Kombination von multiplen Pulmonalarterienaneurysmen und tiefer Venenthrombose charakterisiert; es stellt eine seltene sowie unterdiagnostizierte klinische Entität mit weniger als 40 publizierten Fällen in der englischsprachigen medizinischen Literatur dar. Vaskuläre Ereignisse im Sinne von Venenthrombosen, wie sie im Verlauf des M. Behçet auftreten, wurden auch bei HSS beschrieben, z. B. Vena-cava-, intrakardiale, Jugularvenen-, Iliakalvenen-, Femoralvenen- und Duralsinusthrombosen. Hier wird der Fall eines 35-jährigen Mannes mit HSS beschrieben, der klassische Zeichen des Syndroms in Form rezidivierender Thrombophlebitiden der Beinvenen, Pulmonalarterienaneurysmen und eine Ischämie des linken Beins mit extensiver Beteiligung der arteriellen Äste aufwies. Bei dem Patienten zeigten sich eine kritische arterielle Ischämie im linken Bein in Kombination mit einer Thrombose der

Aorta sowie der linken A. iliaca communis, Okklusion der linken A. femoralis superficialis und Okklusion beider Beinarterien. Zur Rettung der linken unteren Extremität wurden notfallmäßig Gefäßoperationen durchgeführt. Kurz darauf begann für den Patienten eine Kortikosteroid-Cyclophosphamid-Stoßtherapie mit erheblicher Besserung unter anschließender monatlicher Gabe von Cyclophosphamid über ein Jahr. In der vorliegenden Arbeit werden die arterielle Beteiligung bei HSS sowie Ähnlichkeiten von HSS und M. Behçet in Hinblick auf thrombotische Ereignisse erörtert. Zusammenfassend werden die aktuellen Optionen zur Therapie des HSS dargestellt.

Schlüsselwörter

Hughes-Stovin-Syndrom · Unvollständig ausgeprägter M. Behçet · Arterielle Vasculitis bei Hughes-Stovin-Syndrom · Aortitis · Thrombophlebitis · Pulmonalarterienaneurysmen

Table 1 Summary of the Hughes–Stovin syndrome (HSS) case reports sharing some features of the classic triad of Behçet's Disease (BD), e. g., mouth and/or genital ulcers and/or eye inflammation and other cases with HSS lacking any of the classical features of BD

Summary of HSS case reports revealing some of the classical features of BD	
Kechida et al. [14]	Case 1: aortic aneurysm in a 55-year-old man who initially presented with deep venous thrombosis; the diagnosis of HSS revealing BD was made given the history of recurrent oral and genital ulcers
Bennji et al. [3]	Case 2: multiple pulmonary aneurysms with life-threatening hemoptysis; pulmonary artery coil embolization and right lower lobectomy were performed; extensive bilateral femoral deep vein thrombosis extending into the inferior vena cava, massive hemoptysis. A final diagnosis of BD was made after extensive investigations
El Jammal et al. [15]	Case 3: 19-year-old man with hemoptysis; tongue ulcers; CT angiography revealed femoral vein thrombosis, large threatening aneurysm of a left pulmonary artery segment. The aneurysm was embolized and simultaneously a vena cava filter was inserted
Robinson et al. [16]	Case 4: A 21-year-old male; recurrent oral ulcers; no genital ulceration; superficial thrombophlebitis; pulmonary CT angiography a 35 mm right lateral segmental PAA with multiple pulmonary artery in situ thromboses
Demirkan and Gültekin [17]	Case 5, 6: two patients with HSS who presented with pulmonary artery aneurysm, thrombophlebitis, hemoptysis, and oral ulcers
Al-Jahdali [18]	Case 7: 23-year-old Saudi woman; recurrent oral ulceration; right-lower lobe PAA; papilledema; DVT
Yagi et al. [19]	Case 8: A 32-year-old male; multiple PAA; DVT of the right leg and the right femoral vein; thrombosis of the vena cava; aphthous ulcer in the oral cavity, an ulcer in the genital region
Madiha and Sami [20]	Case 9: DVT; oral ulcers; giant aneurysm in the left lower lobe pulmonary artery
Summary of HSS case reports without features of the classic triad of BD	
Hughes and Stovin [1]	Case 1, 2: two male patients; segmental PPA with peripheral venous thrombosis
Kopp and Green [21]	Case 3, 4: two male patients; PAA and recurrent thrombophlebitis
Fabi et al. [22]	Case 5: A 12-year-old boy; right atrium endocardial mass; jugular vein and cerebral venous thrombosis; deep venous thromboses; PAA
Abdelbary et al. [23]	Case 6: A 35-year-old Egyptian female lower limb deep vein thrombosis; pulmonary aneurysm
Ribeiro et al. [24]	Case 7: A 43-year-old male; superficial thrombophlebitis and DVT of the lower limbs; PAA
Pankl et al. [25]	Case 8: A 41-year-old man; deep venous thrombosis of the right leg, and PAA
Al-Zeedy et al. [26]	Case 9: A 53-year-old man; DVT and PAA
Kably and Reveron [27]	Case 10: A 41-year-old male; massive hemoptysis; ruptured PAA; DVT; cardio-venous thromboembolism; pulmonary infarction
El Aoud et al. [28]	Case 11: A 42-year-old woman; DVT; PAA
Jaramillo et al. [29]	Case 12: A 47-year-old male; dilated main pulmonary arteries, multiple right bronchial artery aneurysms and a splenic artery aneurysm
Silva et al. [30]	Case 13: A 25-year-old male; DVT; PAA
Grembale et al. [31]	Case 14: DVT; PAA; Budd–Chiari syndrome; a thrombotic occlusion of inferior vena cava
Amezyane et al. [32]	Case 15: A 28-year-old female; right ventricular thrombus, PAA, iliac vein thrombosis; caval thrombosis
Kim et al. [7]	Case 16: A 45-year-old man; massive hemoptysis; DVT; bilateral PAA and inferior vena caval thrombosis
Chalazonitis et al. [9]	Case 17: A 18-year-old, Greek male patient; DVT; PAA; superior sagittal and transverse sinuses
Emad et al. [2]	Case 18, 19: two male patients; DVT, PAA, superior sagittal sinus thrombosis in one case
Balci et al. [33]	Case 20: A 41-year-old patient; multiple PAA; thrombus in both the inferior and superior vena cavae
Herb et al. [34]	Case 21: A 25-year-old man; PAA; multiple aneurysms of the bronchial arteries; severe hemoptysis; aneurysm of the left hepatic artery
Margolesky et al. [35]	Case 22: A 38-year-old woman; DVT, PAA; right ventricle thrombus; transverse myelitis

PAA pulmonary artery aneurysms, DVT deep vein thrombosis, BD Behçet's disease, HSS Hughes–Stovin syndrome, CT computed tomography

thrombosis of the cardiac chambers, jugular vein, iliac vein, femoral vein, and dural sinuses as previously described in detail by Khalid and Saleem [4]. Various venous thrombotic events occurring during the course of BD have also been described in HSS, e. g., inferior sagittal sinus thrombosis [2], superior sagittal and transverse sinus thrombosis

[9], inferior vena cava thrombosis [7], inferior vena cava (IVC) and intra-cardiac mural thrombosis [8], basilic vein thrombosis [5], and IVC and portal vein thrombosis [10].

No previous report described such extensive arterial involvement in HSS as observed in our case. This fits with the finding that pulmonary artery aneurysms

in HSS proved to be vasculitis of the pulmonary arteries upon histopathological examination [11]. An important detail that merits consideration here is that the clot in the pulmonary arteries in HSS or BD is mostly due to arterial vasculitis rather than to venous thromboembolism, especially in patients without DVT. Also, the thrombi in the lower extremities are

tightly adherent to the inflamed veins with no tendency for propagation in BD and HSS patients [5]. Likewise, in our case, no saphenous vein thrombosis was found, just luminal attenuation on both sides, denoting underlying phlebitis. At the same time, small pulmonary artery aneurysms were seen associated with in situ thrombosis which are not due to pulmonary embolism and/or thromboembolism from the peripheral deep venous system. BD, being a systemic vasculitis, may affect virtually all types and sizes of vessels involving pulmonary arteries, veins, and septal capillaries [12].

It is generally agreed that the treatment of pulmonary vasculitis in HSS and BD follows the same lines, because, at present, these are the only two conditions known to predispose to pulmonary artery aneurysms with underlying pulmonary arterial vasculitis [2, 6]. The new updated EULAR recommendations for the management of Behçet disease advise the use of high-dose glucocorticoids and cyclophosphamide for treatment of pulmonary artery aneurysms, while monoclonal anti-TNF antibodies should be considered in refractory cases. For patients who have or who are at a high risk of major bleeding, embolization should be preferred to open surgery. For the management of acute deep vein thrombosis in BD, glucocorticoids and immunosuppressives such as azathioprine, cyclophosphamide, or cyclosporine-A are recommended. For both aortic and peripheral artery aneurysms, medical treatment with cyclophosphamide and corticosteroids should be applied first, before considering intervention to repair. Surgery or stenting should not be delayed if the patient is symptomatic [13].

In BD patients, pulmonary hemorrhage is one of the main causes of death and prognosis is poor if pulmonary aneurysms are left untreated [2, 6]. But if the patient also has extensive acute arterial ischemia with thrombosis, starting anticoagulation may be necessary, as in our case; this has to be decided in every single case. The issue of anticoagulation in patients with HSS and BD is complex and further studies are needed before definite recommendations can be made, as stressed by the fact that the use of an-

ticoagulants and antifibrinolytic agents in BD is not currently recommended by EULAR [13].

Conclusions

- **To the authors' knowledge, our report is the first to report extensive arterial involvement with major arterial vasculitis in a case of HSS.**
- **Though most cases of HSS present with hemoptysis, also arterial vasculitis of the lower limbs and abdominal aorta should alert the clinician to think of this rare syndrome, provided that other causes are ruled out. This is especially the case in patients with the typical classical features of HSS, notably recurrent thrombophlebitis or deep vein thrombosis or thrombosis elsewhere, in association with pulmonary artery vasculitis and aneurysm formation with in situ thrombosis.**
- **For early diagnosis the radiologist may play an important role.**
- **The management of pulmonary vasculitis in HSS should follow the same lines used for its treatment in BD. The issue of anticoagulation in these patients is challenging and requires further deliberation and should be individualized according to the clinical presentation.**

Corresponding address

Prof. Y. Emad, MD, PhD
Rheumatology Department, Faculty of Medicine, Cairo University
Cairo, Egypt
yasseremad68@gmail.com

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Compliance with ethical guidelines

Conflict of interest Y. Emad, Y. Ragab, A. El-Marakbi, A. Saad, O. Ibrahim, A. Abd-Elhalim, H. El-Santawi, and J.J. Rasker declare that they have no competing interests.

This article does not contain any studies with human participants or animals performed by any of the authors. For images or other information within the

manuscript which identify patients, consent was obtained from them and/or their legal guardians.

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