An unusual case of refractory Behçet's disease presenting with co-occurrence of thrombosis and thymoma

Sir,

Behçet's disease (BD) is a chronic vasculitis with mucocutaneous, articular, and vascular manifestations in 20-40% of cases (1). We report a rare case of refractory BD complicated by deep vein thrombosis (DVT) and an incidental thymoma, notable for its absence of myasthenia gravis (MG), highlighting unique immunopathogenic and therapeutic challenges.

A 30-year-old woman presented with a 5-year history of recurrent painful oral aphthous ulcers (4-5 times yearly, lasting 7-10 days), vulvar ulcers, and inflammatory polyarthralgia (knees, ankles, wrists; morning stiffness ~30 minutes). Physical examination revealed three buccal/tongue aphthae, two erythematous vulvar ulcers, and positive pathergy test, fulfilling International Study Group criteria for BD (2). Initial laboratory findings showed elevated erythrocyte sedimentation rate (ESR, 45 mm/h; reference: 0-20 mm/h), C-reactive protein (CRP, 18 mg/L; reference: <5 mg/L), and negative antinuclear antibody and rheumatoid factor. Colchicine (0.6 mg twice daily) failed to prevent 3-4 annual flares, requiring prednisone bursts (10-20 mg daily for 7-14 days). Postpartum exacerbation after an uncomplicated pregnancy prompted continued corticosteroids.

Four years post-diagnosis, acute left lower extremity swelling led to Doppler ultrasound confirming popliteal and peroneal DVT. Hypercoagulable workup was negative (normal protein C, protein S, antithrombin III; negative anticardiolipin and beta-2 glycoprotein I antibodies; no factor V Leiden/prothrombin mutations) except for positive lupus anticoagulant (ratio 1.8; reference: <1.2), suggesting inflammation-driven thrombosis (3). Adalimumab (40 mg every two weeks) and apixaban were initiated, resolving symptoms and thrombus within months.

Six months later, substernal chest pain prompted chest CT, revealing a 2.1 cm anterior mediastinal mass and splenomegaly (splenic length 14 cm, likely reactive). PET/CT showed size reduction to 1.8 cm with low fluorodeoxyglucose uptake (SUV max 2.0), favouring thymoma over lymphoma. Video-assisted thoracoscopic thymectomy, per 2025 NCCN guidelines (4), confirmed a Masaoka-Koga stage I, WHO type B1 thymoma (lymphocyte-rich, non-invasive). No MG or paraneoplastic syndromes were noted. Concurrent hyperthyroidism (TSH 0.1 mIU/L; reference: 0.4-4.0 mIU/L; elevated free T4) was managed with methimazole (10 mg daily), achieving euthyroidism, and perioral dermatitis was treated with topical metronidazole. At one-year follow-up, adalimumab maintained BD under control (normalised ESR/CRP, no flares), with no thymoma recurrence.

Thymomas are associated with autoimmune disorders (30-40%), typically MG, but rarely vasculitides (5). Literature reports one case of thymic carcinoma with BD and IgA nephropathy (6) and rare BD-MG overlaps, often thymoma-related (7, 8). This case is unique for its benign thymoma, absence of MG, and incidental detection in a young woman with vascular BD. Defective thymic T-cell regulation may contribute to BD's vasculitic phenotype (5, 9). Colchicine's limitations in refractory BD necessitated adalimumab, aligning with EULAR recommendations (10). This case underscores BD's systemic complexity and the need for vigilant screening for rare comorbidities like thymoma, which may inform immunopathogenesis. Multidisciplinary management, including biologics and surgical resection, was pivotal. Further genomic studies (*e.g.*, HLA-B51) could elucidate BD-thymoma links.

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