Pheochromocytoma in association with focal dermatomyositis

Rheumatology key message

- Detailed malignancy screening is of paramount importance in patients presenting with features consistent with dermatomyositis.

Sn, DM is a rare autoimmune inflammatory muscle disease with a known increased risk of associated malignancy [1]. Pheochromocytomas are very rare tumours for which the reported incidence is 0.2% in a hypertensive population [2]. We report a patient with a focal pattern of DM and an associated pheochromocytoma.

A 53-year-old lady presented to the Salford Royal neuromuscular clinic with a 10-month progressive history of red marks, pain and swelling of the right forearm. She was unable to fully move her forearm and grip was also affected. No other muscle groups were involved. A month later, her symptoms progressed further, with development of a rash over her arm and face and an accompanying periorbital swelling. There were no associated features to suggest an underlying CTD, although at age 7 she was treated with steroids for suspected SLE. Past medical history included hypothyroidism, diet-controlled diabetes mellitus and hypertension. Medications included an angiotensin-converting-enzyme inhibitor, a calcium channel blocker and thyrroxine.

On clinical examination, her right arm was swollen in the flexor compartment, with associated limitation and weakness of wrist flexion at Medical Research Council grade 4/5 using manual muscle testing. There was no evidence of weakness contralaterally or in other muscle groups proximally or distally. There was a subtle, but not definite, heliotrope rash over her eyes. The reported periorbital oedema had subsided by that point. Respiratory, cardiovascular and abdominal examinations were normal and blood pressure was 137/85 mmHg. Initial blood screen results were within normal limits: CRP <5 (n < 10 mg/l), creatine kinase 135 (<170 U/l), ANA positive 1:100 speckled pattern. A further immunological screen was normal (centromere, Mi-2, TIF1g, MDA5, NXP2, SAE1, Ku, PM-ScI100, Pm-ScI75, SRP, PL-7, PL-12, EJ, OJ, ENA, Jo-1, Ro52, dsDNA all negative). A magnetic resonance scan of the right forearm using short tau inversion recovery sequences showed high signal changes in the T1 sequences.

An open muscle biopsy of the right forearm at this point had not been diagnostic. A targeted open muscle biopsy in the right arm showed evidence of dense and patchy endomyositis and perivascular inflammation without fibrinoid necrosis as well as widespread HLA-1 upregulation and dense Membrane Attack Complex deposition along the capillary walls—consistent with inflammatory myopathy (features of DM). As the patient remained symptomatic with localised pain and weakness, she was commenced on oral prednisolone EC at 30 mg/day. AZA was subsequently started, and within 2 months her rash fully resolved and manual muscle testing indicated normal strength in the right arm. The steroids were gradually tapered and, following worsening liver function tests, AZA had to be withheld.

Histopathological confirmation of DM prompted a screening for underlying paraneoplastic disease. A CT thorax/abdomen/pelvis, mammogram and upper and lower endoscopies were requested to rule out underlying malignancy. A right-sided 42-mm adrenal mass was observed on CT scanning, suspected to be a pheochromocytoma. At the time she was being treated for resistant high blood pressure with two different antihypertensive medications. Subsequent blood tests of metanephrines and catecholamines confirmed the findings of a functioning pheochromocytoma. Surgical resection was planned and phenoxybenzamine commenced to ensure adequate pre-operative blood pressure control. The patient underwent a right laparoscopic adrenalectomy with no complications. Histology confirmed a 46 × 38 × 52 mm tumour confined to the adrenal gland and microscopically described as pheochromocytoma with complete resection margins. Post-operatively, the patient remained normotensive and phenoxybenzamine was stopped. The DM remained in remission and steroids were tailored down, with a plan to reinstate AZA if required. At 18 months following surgery, the patient remains well, her DM remains quiescent and the prednisolone dose is being slowly reduced (currently 10 mg/day).

DM is associated with malignancy, usually within 3 years of diagnosis, and cancer screening is vital, especially in patients over the age of 50 with no detectable antibodies on routine antibody screening [3, 4]. In our case, screening was conducted following confirmation of diagnosis by histopathology. Pheochromocytomas are rare tumours of the adrenal medulla that can be benign or malignant, and up to 90% of tumours are functioning. Cure is possible in 90% of cases by surgical resection after initial preparation with alpha blockade [5]. Cardiac muscle damage due to excess catecholamines has been previously reported [6]. It is possible that our patient presented with localized skeletal muscle inflammation and damage in part due to excess catecholamine production. A literature search found only two reported cases of DM and pheochromocytoma coinciding together: a post-mortem finding of myositis associated with pheochromocytoma, and an inactive pheochromocytoma with confirmed DM [2, 5, 7].

In summary, both DM and pheochromocytoma are extremely rare conditions, and the temporal coexistence of these two diseases is extremely rare. Identification of malignancy should prompt screening for other associated malignancies.
of the two conditions makes our case even more unusual. The case presented reinforces the requirement for detailed malignancy screening of patients presenting with features consistent with DM.

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**References**