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A CASE REPORT OF BROWN SYNDROME COMPLICATING THE MANAGEMENT OF SCLEROMYOSITIS

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Background: Brown syndrome is a rare ocular motility disorder which has been reported in JRA, RA and SLE but never in a patient with scleromyositis.

Objectives: To report the first case of Brown syndrome in a patient with scleromyositis and increase awareness of this condition.

Methods: A case report and discussion.

Results: The patient was diagnosed with scleromyositis, at the age of 34, after presenting with arthralgia, sclerodactyly, skin pigmentation, Raynaud's phenomenon, mild muscle weakness and dyspnoea. His labs were CRP 47 mg/L, CK 868 IU/L, ANA strongly positive; anticentromere Ab negative and Anti-PM/Scl-75 and Anti-PM/Scl-100 Ab positive. HRCT chest showed extensive pulmonary fibrosis with lower lobe honeycombing. TLCO was 3.98 (33% of predicted).

He was initially managed with high dose steroids and pulsed IV cyclophosphamide with azathioprine for maintenance therapy. His lung disease stabilised and myositis resolved but he continued to develop calcinosis cutis so was switched to 6 monthly IV rituximab.

6 years later, he developed morning headaches with intermittent diplopia, described as double vision in vertical gaze with one image being above the other. Episodes lasting 10 minutes to 2 hours. Examination showed normal visual acuity and fundoscopy, no peripheral or eye muscle weakness.

Investigations to exclude myasthenia gravis, cerebral vasculitis and atypical infection were organised (MRI, AChR antibody, lumbar puncture, MRA) and were normal.

Because of intermittent nature of his episodes, his eye examination was always normal but he captured images in disconjugate gaze with right eye looking upwards and outwards when trying to look straight (Figure 1). Occasionally this was associated with orbital pain and an audible click. These features are suggestive of Brown syndrome.

He continues to have recurrent episodes despite immunosuppression but prednisolone 20mg daily for 1-2 days at onset of each attack causes rapid resolution of symptoms.

Figure 1. Right eye looking upwards and outwards when trying to look straight

Conclusion: Scleromyositis is an overlap syndrome of scleroderma and dermatomyositis. Muscle involvement is mild and clinical presentation can be variable. The PM/Scl antibodies are highly characteristic of the syndrome. (1) Brown syndrome is an ocular motility disorder, first described in 1950, characterized by the inability to fully elevate the affected eye in adduction due to pathology of the superior oblique tendon sheath. (2)

It can be congenital or acquired, viz, trauma, surgery or sinusitis and also been described in RA, JIA and SLE. (3) If superior oblique tendon cannot relax or slide freely through the trochlea then the affected eye cannot depress completely, leading to diplopia on upward gaze. (4) In inflammatory disease it is thought that swelling of the posterior part of the superior oblique tendon or tenosynovitis are likely causes of the tendon sheath abnormality. (4) This is likely to be the case in this patient because his symptoms are recurrent, respond to steroids and tend to occur more towards the end of rituximab cycles.

Recognition of this syndrome is important because invasive investigations can be avoided. Also, intermittent diplopia in a patient with autoimmune disease is suggestive of myasthenia gravis which maybe incorrectly diagnosed.

Finally, this case demonstrates the syndrome can be easily managed with short courses of oral steroids, although patients who are already on immunosuppressant treatment may need this in addition.

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FIBROUS ARTHROPATHY AS THE KEY FEATURE OF JUVENILE SCLERODERMA - CASE REPORT

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Background: Systemic sclerosis (SSc) is an autoimmune disease characterized by endothelial dysfunction, immunological disorders, and excessive synthesis of collagen and its deposition in various tissues and organs. The juvenile onset SSc before the age of 16 is very rare, with annual incidence of 0.27-0.5 cases per million children according to English and Finnish authors [1,2].

Objectives: To present a clinical case of juvenile onset SSc, manifesting from the childhood predominantly with fibrous contractures.

Methods: Patient K., 30 yo. Clinical presentation on admission to the Institute of Rheumatology in September 2020: thickening of the trunk and limbs skin (mRSS 10 scores), pronounced induration of subcutaneous tissues and muscles; contractures of the elbow, shoulder, hip and knee joints, short stature (height 142 cm) with proportional shortening of the limbs. ANA (HEP-2) 1/320, a-Scl-70, a-RNP-70 and ACA tests were negative. Ultrasonography revealed left-sided coxitis, esophagogastrroduodenoscopy - Barrett's esophagus. Chest CT, echocardiography, electrocardiography and capillaroscopy yielded no specific findings.

The patient has been ill since the age of 3, when SSc manifested with skin thickening, "dry" arthritis and rapid development of contractures of the large joints. Thorough diagnostic elaboration ruled out such potential causes as phenylketonuria, glycogenosis, mucopolysaccharidoses, primary amyloidosis, and porphyria. Histological findings (2007) of a biopsied skin specimen containing subcutaneous fat and muscle tissue included focal vacuolization of keratinocytes, poor perivascular lymphocytic and histiocytic infiltration, fibrosis and hyalinosis of collagen fibers of varying intensity in the mid- and deep dermis, infiltration of collagen fibers by fibroblasts, skin appendages atrophy – all of them representing a pattern of morphological changes characteristic of SSc. Therapeutic regimens including prednisone at 5-15 mg/day and D-penicillamine were ineffective.

Results: In this case, in view of fibrotic arthropathy, a differential diagnosis was made with deep morphea and stiff skin syndrome. Visceral involvement, immunological disorders and biopsy findings substantiated a diagnosis of juvenile onset SSc. Oral MTX was initiated at 15 mg to target skin lesion and osteoarticular symptoms.

Conclusion: Predominance of fibrotic arthropathy in presented case caused difficulties in establishing SSc diagnosis, as this patient did not have such inherent features as the Raynaud's phenomenon, interstitial lung disease or pulmonary hypertension. Juvenile onset SSc manifesting before the age of 16 has its own clinical features, usually persisting through the adulthood, and therefore, such one-of-a-kind appearances of juvenile onset SSc should not be missed or misinterpreted.

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CHRONIC INTESTINAL PSEUDO-OBSTRUCTION WITH HYDRONEPHROSIS: A CASE REPORT ON SUCH DISABLING AND RARE COMPLICATION OF LUPUS

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