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CASE REPORT

Acquired unilateral Brown Syndrome in newly diagnosed Systemic Lupus Erythematosus

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Brown Syndrome is a congenital or acquired ocular movement disorder that is known to be a rare complication of Systemic Lupus Erythematosus (SLE). We report a case of acquired Brown Syndrome in an adolescent girl with newly diagnosed SLE which responded well to oral prednisolone

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Introduction

Brown Syndrome is caused by abnormalities of the superior oblique tendon-trochlea complex that results in difficulty and diplopia on elevating the affected eye when held in adduction. Acquired causes of Brown Syndrome include rheumatological conditions such as SLE and, although rarely reported in the literature, this association may develop and may require treatment.

Case Presentation

A 14 year 11-month-old adolescent girl with a history of asthma and heavy menstrual bleeding, initially presented with a two-month history of lethargy, a malar rash, occasional shoulder pains anorexia and weight loss of 6 kg. Initial investigations revealed lymphopenia, anaemia, and a raised erythrocyte sedimentation rate (ESR) of 40 mm in the first hour. During the week following presentation the patient developed central chest pains and shortness of breath and was admitted in view of deteriorating symptomatology.

Further investigations showed a raised rheumatoid factor (29 U/ml), anti-nuclear antibody (ANA) positivity (>1/1000), raised anti-double stranded DNA (>800) with homogenous anti-nuclear (ANF) pattern and low complement levels (C3 589mg/L and C4 58mg/L). A diagnosis of Systemic Lupus Erythematosus (SLE) was made; a dose of 80mg intramuscular methylprednisolone was administered and the patient was started on regular analgesia including paracetamol and ibuprofen.

Figure 1 This shows an axial T2 weighted MRI image demonstrating a subtle increased signal intensity of the right lateral rectus muscle

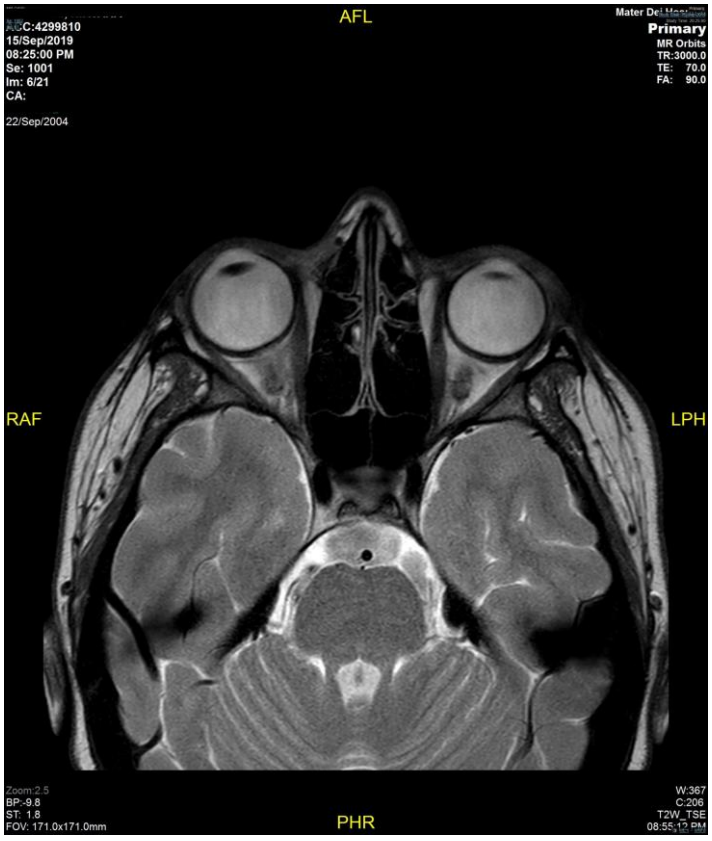
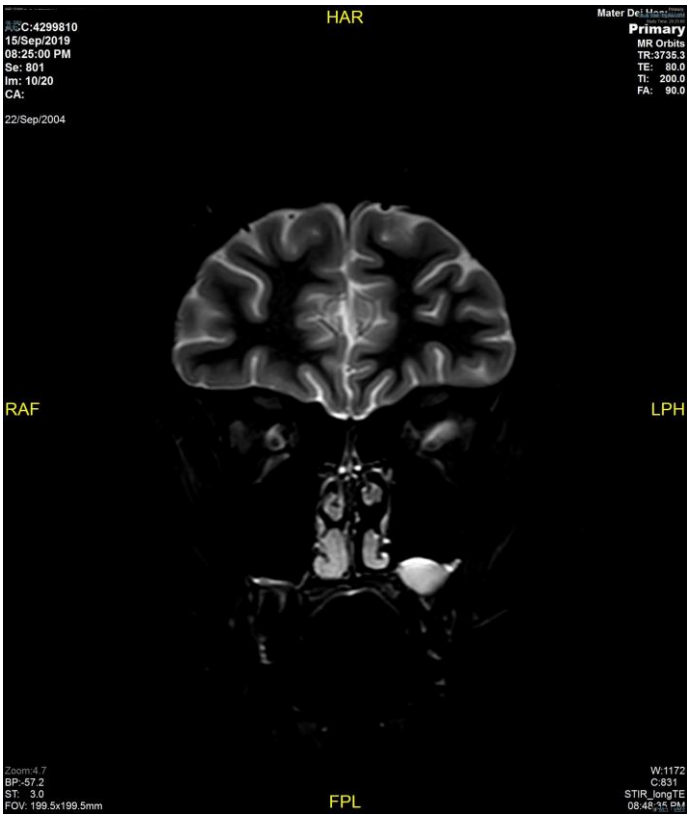


Figure 2 This shows a coronal STIR MRI image also demonstrating a subtle increased signal intensity of the right lateral rectus muscle



During this admission the patient continued to complain of myalgias, arthralgias and chest pain, and also developed new onset blurred vision on upward gaze. Vertical diplopia of the right eye was reported on upward gaze especially in the superior-nasal direction suggestive of a right-sided acquired Brown's syndrome, confirmed after an urgent ophthalmological assessment. An MRI of the head and orbits confirmed a high intensity signal of the oblique and rectus muscles, affecting the right extraocular muscles more than the left as depicted in Figures 1 and 2. The patient was started on hydroxychloroquine 200mg twice daily and a four-week course of prednisolone by mouth, starting with 20mg daily tailing down by 5mg per week; with close follow up from the multidisciplinary team. The patient remained in good health and, in view of this, further imaging was not deemed to be clinically indicated.

Discussion

Brown Syndrome is characterized by an upward gaze impairment that occurs when the affected eye is held in adduction and was first described in 1950 by Dr. Harold W. Brown.[1] The pathogenesis of the disease involves congenital or acquired abnormalities of the superior oblique (SO) tendon-trochlea complex that restricts the mobility of the SO muscle.[2] Congenital Brown syndrome is the commonest form, is of unknown aetiology and confers a worse prognosis.[3] Rheumatic and non-rheumatic causes of acquired Brown Syndrome have been described including trauma and a complication of peri-orbital surgery, sinusitis, myopathies, rheumatoid arthritis, juvenile chronic arthritis, and, as in this case report, systemic lupus erythematosus.[4]

A review of the medical literature only discovered three case reports associating the development of Brown Syndrome with the diagnosis of SLE. The first reported case, published in 1990, described a 30-year-old man with a 5-month history of generalised arthralgia and acute alopecia who developed variable diplopia on upward gaze, which was diagnosed as Brown Syndrome; further investigation confirmed an underlying diagnosis of SLE.[5] The other two cases concerned young women who were known to suffer from SLE and who presented with acute diplopia, also diagnosed as Brown Syndrome.[6-7] It is postulated that rheumatic conditions may cause stenosing tenosynovitis of the SO tendon with impingement of the muscle and resultant decreased ocular motility.[8] The increased signal intensity at the level of the SO muscle reported on MRI and the response to oral steroids shown by our patient suggests that this was the most likely underlying pathophysiology. The treatment of acquired Brown Syndrome varied in the literature from conservative management, locally injected steroids and systemic steroids, non-steroidal anti-inflammatories and sometimes even surgery.[3, 8] Among the three case reports of SLE-associated Brown syndrome, one patient responded to oral ibuprofen, one responded to oral prednisolone the third case did not receive any treatment for this complication.

The exact incidence, prognosis and management of acquired Brown Syndrome in cases of SLE is unknown and limited data has been reported and published. Diagnostic difficulties and mild or transient symptomatology of this rare complication have likely resulted in under-reporting.[5]

Conclusion

This case report describes how a 14-year-old girl, newly diagnosed with SLE, developed concomitant acquired Brown Syndrome which responded well to oral steroids. Although it is a scarcely reported association, Brown Syndrome should be considered as a possible cause of diplopia in patients known to suffer from SLE and its management may involve MRI, ophthalmology input and steroids.[6]

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