MENINGIOMA AND LUPUS – A DEADLY DUO! – A REPORT OF 2 CASES

Sriram Sankaran¹, Rajeswari Sankaralingam²

¹Postgraduate, Institute of Rheumatology, Madras Medical College, Chennai
²Director and Head, Institute of Rheumatology, Madras Medical College, Chennai
ara12283@yahoo.co.in

ABSTRACT

Living with lupus is deadly! Adding fuel to fire is the occurrence of meningioma in these patients. We describe 2 cases of meningioma from a large cohort of around 400 systemic lupus erythematosus patients. Both the patients were middle aged females diagnosed to have lupus by Systemic Lupus International Collaborating Clinics (SLICC 2012) classification criteria with disease onset at 29 and 39 years respectively. One patient had class V lupus nephritis and was on high dose steroids, hydroxychloroquine and mycophenolate mofetil. The other patient was on low dose steroids, hydroxychloroquine and azathioprine as steroid sparing agent. Both patients presented with gradually worsening headache. Magnetic resonance imaging of brain showed a mass lesion in both the patients suggestive of meningioma. Both were referred to neurosurgery department and were operated successfully. Occurrence of meningioma in patients with lupus is a matter of pure coincidence. We report this deadly combination as the occurrence of both in a given patient is rare.

Key words: Systemic lupus erythematosus, Meningioma, Magnetic resonance imaging

INTRODUCTION

Systemic lupus erythematosus (SLE) is an autoimmune connective tissue disorder with unknown etiology, diverse pathogenesis and an array of clinical features. Long term follow up of these patients have shown a moderately increased risk of malignancies (particularly lymphoma). Few case reports have been reported till date showing the occurrence of both in a same patient. Early recognition of such malignancies can lead to a better management and good clinical outcome, thereby reducing the morbidity and mortality occurring as a result of malignancy. We report 2 cases of meningioma in our lupus patients.

CASE 1

30 years female was diagnosed to have SLE in our lupus clinic 1 year back using SLICC 2012 Classification Criteria. She had malar rash, oral ulcer, polyarthralgia, photosensitivity, leucopenia and thrombocytopenia. Her Antinuclear antibody (ANA) by indirect immunofluorescence showed homogenous (4+) pattern. Extractable Nuclear Antigen (ENA) profile by immunoblot showed positivity for anti dsDNA and anti Sm antibodies. Antiphospholipid antibodies were negative. As she had class V lupus nephritis, she was treated with high dose steroids, hydroxychloroquine and mycophenolate mofetil. Since the patient had a gradually increasing persistent headache, which had increased in intensity disturbing her routine, she consulted us. She was advised MRI brain which showed a well defined T2 isointense, flair
non suppressible lesion measuring 3.4*2.9cm with no diffusion restriction in the right parafalcine region suggestive of a meningioma(fig 1). After controlling lupus activity, she was referred to neurosurgeon and was operated successfully.

CASE 2

41 years female, a known patient with lupus, who was diagnosed in our lupus clinic 2 years back using SLICC 2012 criteria. She had oral ulcers, discoid lesions in the scalp and face with scarring alopecia and polyarthritis. ANA by indirect immunofluorescence showed speckled pattern(3+). ENA by immunoblot showed anti SS A, anti SS B and anti dsDNA positivity. She was on low dose steroids, topical steroid, hydroxychloroquine and azathioprine. She had an intense headache over the last 3 months with frequent vomiting. MRI brain showed T1 iso T2 hypointense extra axial lesion of size 2.2*1.8*2cm in the left frontal convexity with intense homogenous enhancement. There was adjacent frontal lobe compression with edema(fig 2). The lesion was suggestive of meningioma. She was also referred to neurosurgeon and was operated successfully.

DISCUSSION AND REVIEW OF LITERATURE

Systemic lupus erythematosus is a multi system autoimmune disease with diverse clinical features. There is increased risk of hematological malignancies like Non hodgkin’s and Hodgkin’s lymphoma. There is a moderately increased risk of lung cancer, and possibly for rarer cancer types such as hepatobiliary and vulvar/vaginal malignancies[1]. The incidence of brain tumours in lupus is very low.

Meningiomas are the most common tumours of the brain. They are non glial tumours which are extra-axial. They arise from meningiocytes. They are classified based on their location(intradural and extradural). They are more common in females(2:1). Small tumours are found incidentally and may be asymptomatic. Large tumours compress on adjacent structures and present with headache, hemiparesis and change in mental symptoms. MRI is the diagnostic modality of choice. Signal characteristics in MRI include isointense lesions in both T1 and T2 weighted images with homogenous enhancement in T1 contrast.

Few imaging signs(MRI) help in diagnosing meningioma. They include CSF vascular cleft sign[2](representing a thin rim of Cerebrospinal fluid between tumour and brain parenchyma), dural tail sign[3](thickening of the dura adjacent to an intracranial pathology on contrast-enhanced T1 MR Images), ginkgo leaf sign[4] of spinal meningiomas on axial post contrast T1 imaging( leaf representing the distorted spinal cord, pushed to one side of the theca by the meningioma, and the stem representing the stretched dentate ligament), mother in law sign(lesions that enhance early during the arterial phase and remain opacified well after the venous phase – “mother in law comes early and goes late”), sunburst sign(characteristic vascular supply when viewed from side).


We report this combination of lupus and meningioma for its rarity, which can mimic the headache which is one of the manifestations of neuropsychiatric lupus. Fortunately, in both our cases, the presentation was classical and was amenable to treatment.
REFERENCES


Fig. 1 MRI brain of patient 1 (description in text)

Fig. 2 MRI brain of patient 2 (description in text)