CASE REPORT
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Brain meningioma in a patient with systemic lupus erythematosus

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Brain meningioma, the most common benign brain tumor, has been reported to account for 13-26% of all intracranial tumors, with a crude incidence rate of 2.3 per 100,000 persons for all types of meningiomas. The prevalence of neuropsychiatric lupus erythematosus is 15-91% and its clinical manifestations are diverse: from mild cognitive dysfunction to serious neurological or psychiatric symptoms. Here, we report the first Korean patient with brain meningioma and systemic lupus erythematosus who had undergone surgical tumor resection.

Keywords: Systemic lupus erythematosus; Meningioma; Headache

INTRODUCTION

Brain meningioma is a common benign brain tumor originating from non-neuroepithelial progenitor cells [1]. Meningiomas account for 13-26% of all intracranial tumors, with a crude incidence rate of 2.3 per 100,000 persons [2]. The incidence increases in the 6th and 7th decades of life, and meningiomas are predominantly found in women, with a gender ratio of 3:2 or 2:1 [1]. A positive association of meningioma with pre-existing diseases has been reported, including diabetes mellitus and pulmonary hypertension, and a negative association with rheumatoid arthritis [3].

Systemic lupus erythematosus (SLE) is a prototypic inflammatory autoimmune disease with multi-organ involvement, varied manifestations, and an unpredictable course. The prevalence of neuropsychiatric SLE ranges from 15-91% of lupus, and its clinical manifestations are diverse: from mild cognitive dysfunction to serious neurological or psychiatric symptoms [4,5]. Brain involvement is a significant cause of morbidity and mortality, and the primary cause of death in 4-8% of patients with SLE [6]. Early detection and proper treatment of brain involvement in SLE with neuropsychiatric features might decisively influence prognosis.

Few occurrences of brain meningioma in patients with SLE have been reported [7,8]. To the best of our knowledge, no SLE patients with brain meningioma have yet been reported from Eastern Asia. In this report, we describe a Korean patient with SLE and concurrent brain meningioma.

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CASE

We report on a 49-year-old female patient diagnosed with SLE who presented with a brain meningioma. She was referred to our hospital for recurrent oral ulcers, malar rash, and arthritis. On physical examination, several ulcers were observed on the lower lips and bilateral buccal mucosa, and a maculopapular rash affected the bilateral malar area. There

was mild effusion in the right knee joint. Laboratory findings showed a hemoglobin concentration 9.2 g/dL, white blood cell 3,450/mm^3 (lymphocytes, 750/mm^3), platelet 187,000/mm^3, erythrocyte sedimentation rate of 45 mm/h, and C-reactive protein 2.3 mg/L. Blood urea nitrogen, creatinine, total protein, albumin, aspartate aminotransferase, alanine aminotransferase, and lactate dehydrogenase were within the normal range. Anti-nuclear antibodies were detected, with a titer 1:640. Reactions for anti-dsDNA, anti-Sm, anti-SSA/Ro, and anti-SSB/La antibodies were also positive. Complement C3 and C4 levels were decreased to 56 mg/dL and 6.3 mg/dL, respectively. Anti-cardiolipin and anti-beta2 glycoprotein 1 antibodies and lupus anticoagulant were not detected, and no proteinuria or red blood cell casts were detected using urine test strips.

The patient was diagnosed with SLE, and treated with low-dose prednisolone (10 mg/day) and hydroxychloroquine (300 mg/day). One week later, she visited the hospital with a severe headache of 3-day duration. On neurological examination, no decreased motor strength or sensory changes were detected, and her mental status was alert. To exclude central nervous system (CNS) involvement in SLE, brain magnetic resonance imaging (MRI) was performed, which demonstrated an extra-axial tumor in the left parietal region, based on the parietal convexity with a dural tail and showing homogeneous signal hypo-intensity on T1-weighted images and hyper-intensity on T2-weighted images (Fig. 1). The tumor measured 13×18 mm in size. The radiological features were suggestive of a brain meningioma. Because her meningioma was large enough to provoke a persistent headache, complete surgical resection was indicated and performed. Histopathological findings from the resected tumor revealed a brain meningioma of the meningothelial type (Fig. 2). No neurological complications were observed. Follow-up brain MRI performed 4 months later showed obliteration of the sulcal spaces in the left parietal lobe without evidence of residual or recurrent tumor. The patient has since remained free of symptoms.

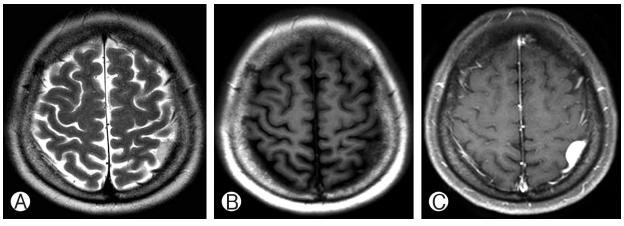

Fig. 1. A 13×18 mm-sized dural-based extra-axial mass is shown in the left parieto-occipital area as hyperintensity on an axial T2-weighted image (A), hypointensity on an axial T1-weighted image (B), and homogeneous enhancement on an axial enhanced T1-weighted image (C).

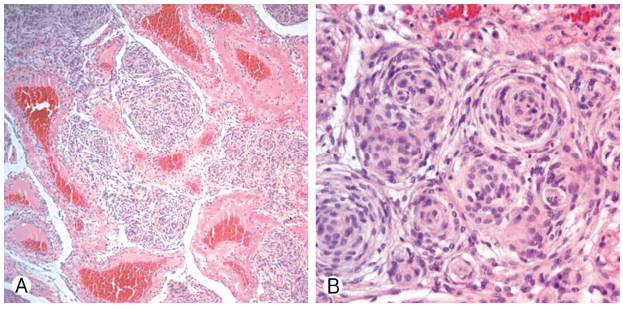

Fig. 2. Microscopic findings demonstrating tightly whorled clusters of tumor cells, suggesting meningioma of the meningothelial type (H&E stain, A×100; B×400).

DISCUSSION

In the Korean population, the overall crude rate of CNS tumors is 11.69 per 100,000 person-years, and meningioma is the most common primary CNS tumor (31.2%). The female-to-male ratio has been reported as 3:2 or 2:1 and the mean age of patients at diagnosis is 57.6 years old. Most meningiomas are benign [1,9].

Most meningiomas are discovered incidentally during examination for unrelated symptoms. If symptoms present, they are determined by the location and size of the mass. Common manifestations of intracranial meningiomas include seizure disorders, cranial nerve dysfunctions, and visual disturbances [10]. Although symptoms of both lupus and meningioma vary and are not specific, imaging study is required for patients who complain of recurrent and intractable symptoms among patients with suspected neuropsychiatric lupus.

Castellino et al. reported three cases of meningioma with lupus erythematosus [8]. The prevalence of meningioma in patients with lupus erythematosus was 1.65% (3 among 181 patients). All three patients were women complaining of neuropsychiatric symptoms characterized by headache, mild cognitive dysfunction, anxiety, depression, and panic attacks, whic h were unrelated to the location of meningioma. One of the patients had undergone surgical removal of meningioma, but showed no improvement of symptoms. The authors concluded that their findings were pure coincidence. Richardson and Cohen reported on a 44-year-old female patient who

developed a meningioma after the onset of subacute cutaneous lupus erythematosus [11].

In previous studies, the risk factors for development of brain meningioma included deletions in the neurofibromatosis type-2 gene, ionizing radiation, head injury, and defective hormonal receptors [1]. Among the risk factors, the role of hormones is expected to have most association between meningioma and SLE as SLE is more prevalent in women and the disease activity of SLE is often associated with oral contraceptive use or pregnancy. Estrogen receptors are particularly associated with aggressive meningiomas. Estrogens are also regarded as potent accelerators of disease in both male and female murine models of SLE [12].

The association between meningioma and autoimmune disease is not yet well established. Meningioma may be more prevalent in patients with lupus erythematosus than in the general population. Although further studies using more extensive data are required, we should be alert to the possibility of meningioma in patients with SLE who present with neuropsychiatric symptoms and use of imaging modalities like MRI should be considered for these patients.

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