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RESEARCH ARTICLE

SPONTANEOUS CERVICAL HEMATOMA IN A PATIENT WITH NPSLE AND APL SYNDROME

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ABSTRACT

A 31-year-old female patient who was receiving warfarin, who was being followed up with neuro-psychiatric systemic lupus erythematosus (NPSLE) and accompanying anti-phospholipid syndrome (APL), who underwent magnetic resonance imaging (MRI) due to complaints of numbness in upper extremity and neck pain in emergency room, who developed cardiac arrest and respiratory arrest during MRI examination and recovered with cardio-pulmonary resuscitation will be discussed with the findings of NPSLE which are suggestive for cerebral involvement.

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INTRODUCTION

Anti-phospholipid syndrome (APLS) is a multi-systemic autoimmune disease complex which develops due to acquired antibody-dependent thrombosis susceptibility (thrombophilia). While no accompanying, precipitating or underlying disease is present in primary autoimmune APLS, secondary autoimmune APLS is seen together with systemic lupus erythematosus (SLE) or another collagen tissue disorder (Fischer, 2007). SLE is a chronic connective tissue disorder with unknown origin, characterized by autoimmune and immunologic disorders and involves many organs and systems. SLE which is a chronic autoimmune disorder and which may present with different clinical manifestations is more common among females aged between 15-25 years, NPSLE which is characterized by central nervous system development may develop in 12-75% of the patients (Ozbek *et al*., 2003; Tokunaga *et al*., 2007; Brey *et al*., 2002). While evidence is available suggesting that this wide range may develop from different ethnic origins, it is also suggested to result from neglecting that the common symptoms like headache may develop from NPSLE (Stojanovich *et al*., 2007).

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NPSLE includes many clinical manifestations. Single or multiple neurologic and/or psychiatric manifestations may accompany with the disease (Hanly, 2005). These clinical manifestations include acute confusion, aseptic meningitis, myasthenia gravis, mono-neuropathy, Guillian-Barre syndrome, demyelinizing syndrome, personality disorders, anxiety disorder, poly-neuropathy, headache, cerebro-vascular disease, seizures, psychosis and more. Acute spinal subdural hematoma (ASSH) is a rare condition which develops secondarily to spinal vascular disorder and leads to compression. ASSH is usually iatrogenic but it may rarely develop spontaneously. Iatrogenic causes include spinal puncture, spinals surgery and trauma. A case was reported to develop following weight lifting in literature (JiEun Park *et al*., 2011). Although spontaneous ASSH is rare, cases were reported to develop from anti-coagulant or anti-platelet use due to primary disease (Russell, 1983).

Case report: A 31-year-old female patient was admitted to emergency room due to numbness in arms. The patient whose complaints started about 1-2 hours ago had been diagnosed with NPSLE and secondary APLS 10 years ago. We have learned that the patient was not going for regular rheumatology controls, she had the history of eight intra-uterine deaths and

she did not have a living baby. She had widespread cerebral atrophy and frontal ischemia fields inconsistent with her age for 3 years, SLE-related nephropathy and diabetes which was suggested to develop from steroids which she sometimes used. She was learned to develop NPSLE and to be assigned to rear service from nursing services. She was using warfarin however she was not going for controls regularly for dose adjustment. The patient was consulted with the neurologist, lateralizing findings were not detected and magnetic resonance imaging (MRI) was planned. Her general condition was good and Glasgow coma scale score was 14-15 however she developed respiratory and cardiac arrest during MRI examination, cardio-pulmonary resuscitation (CPR) was applied, she recovered and transferred to intensive care unit (ICU) thereafter. She was monitored on mechanic ventilator (MV), Glasgow coma scale score was 3-4, APACHE II (Acute Physiology and Chronic Health Evaluation) score was 26, pupils were fixed, dilated, light reflex was quite weak bilaterally, she had livedo reticularis in lower limbs. She was administered vitamin K and fresh frozen plasma as INR (international normalized ratio) value was found as 5.2.

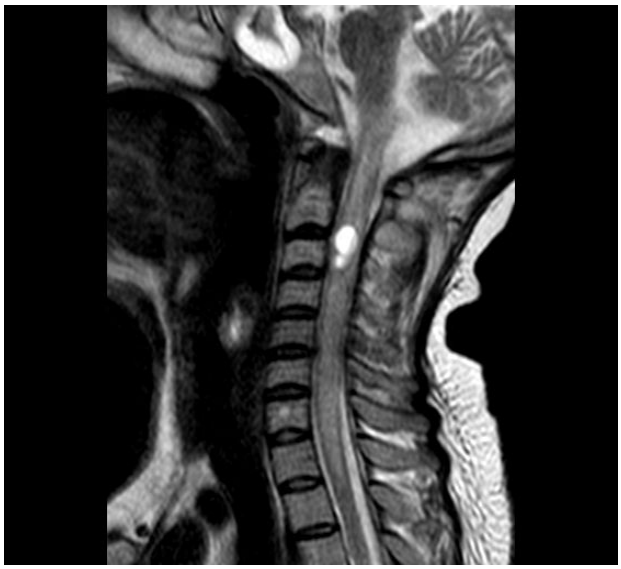

Image 1. Suspected band-shaped hemorrhage field at C2-3 level on cervical MRI

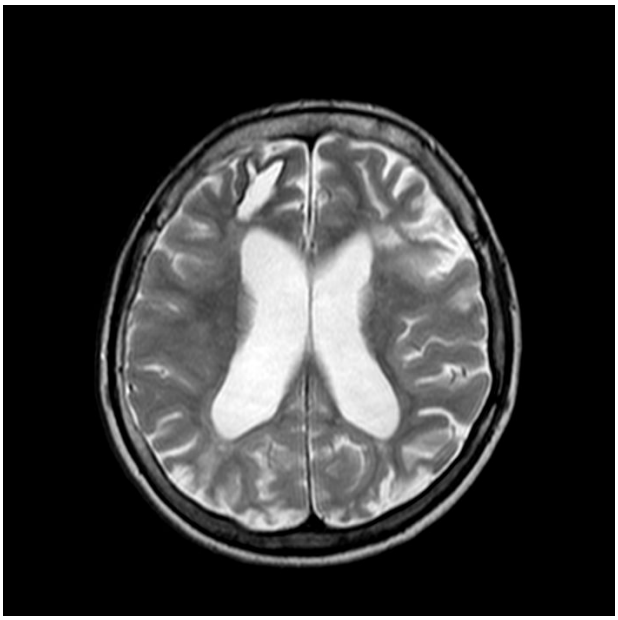

Image 2. Ventricular system and external sub-arachnoid fields are large, cystic area in the right frontal region

Chest X-ray findings were normal, cerebral and cervical MRI revealed a suspected band-shaped hemorrhage field at C2-3 level, a cystic area in the right frontal region, enlargement in ventricular system and external sub-arachnoid area and this finding was interpreted in favor of cerebral parenchymal atrophy (Images 1-2). The patient was consulted with neuro-surgery specialists and operation was not planned. She was followed up at ICU and started 1 gr pulse steroid daily for 5 days after recommended by the rheumatologist. Her clinical condition did not improve so she was administered intra-venous immune globulin (IVIG) for 5 days as recommended by the neurologist. Her clinical condition did not improve and she needed hemo-dialysis (HD) and received HD for three times. Cardiac arrest developed on day 20 of her hospitalization and she died as she did not respond to CPR.

DISCUSSION

American Rheumatology College (ARC) defined 19 different syndromes which influence central, peripheral and autonomic nervous system. None of these syndromes is SLE-specific. So diagnosis of NPSLE is mainly made based on clinical findings. Prevalence may rise up to 37-95% according to the criteria of ARC (Mikdashi, 2004; Hanly *et al*., 2004). A patient with NPSLE may have normal MRI findings while she/he is clinically symptomatic. NPSLE has various MRI findings. The most common MRI finding is small, punctate foci located in subcortical white matter and reported in the ratio of 15-60% followed by cortical atrophy, peri-ventricular white matter changes, ventricular enlargement and large cerebral infarcts (Sibbitt *et al*., 2012; Govoni *et al*., 2004). This vasculitic appearance is proposed to develop from cerebral hypo-perfusion due to widespread vascular changes and early atherosclerosis (Lopez-Longo *et al*., 2003).

Atrophy, small sub-cortical lesions and infarct areas were reported in a study evaluating MRI findings of 81 patients (Lopez-Longo *et al*., 2003). Another study which evaluated 312 NPSLE patients reported no MRI changes in 42% of the patients and pathologic MRI changes were reported in 58%. Pathologic MRI changes included atrophy, vasculopathy, vasculitis, chronic hypo-perfusion, diffuse cortical gray matter lesions as in the previous study (Luyendijk *et al*., 2011). Our patient had known cortical atrophy and ventricular enlargement on MRI for 3 years inconsistently with her age and disease duration besides schizophrenia- like clinical condition, consistently with the previous studies A cystic area in frontal region and spontaneous cervical hemorrhage were detected in our case besides these findings. Coexistence of spontaneous subdural spinal hematoma (SSSH) with coagulation defect, anticoagulant use, surgical intervention, spinal puncture was defined in rare cases before. Spontaneous subdural spinal hematoma is rare in cervical region compared to the other regions and different etiologic factors were reported in literature (Oh *et al*., 2009; Grobovschek, 1989; Reynolds, 1978). The preferred surgical procedure is decompression, laminectomy and hematoma drainage (Calhoun, 1991). Decision for intervention should be made based on life expectation, potential benefit of the therapy and disease severity scores. Spontaneous resorption was seen with conservative approach in some cases (Oh *et al*., 2009). One study has reported successful outcomes with pulse methyl prednisolone (Tae-Jin Song, ?).

A total of 106 non-traumatic patients (49% male, 51% female)were retrospectively evaluated with regard to SSSH which is quite effective on mortality and morbidity; etiologic factor was detected as hemostatic mechanism disorder in 54% of the cases and history of spinal puncture in 47% of the cases (Domenicucci *et al*., 1999). In the same study, 12 out of 106 patients were found to have cervical spinal hematoma. All but two patients (one with cystic fibrosis and one with leukemia) out of 12 were above 47 years. Four patients had the history of anticoagulant use, 3 had spinal puncture, 3 had spinal cord tumor and one had radicular artery aneurysm. Nine patients underwent surgery and 3 did not undergo surgery after SSSH development. Two out of 3 patients who did not undergo surgery died. However Glasgow coma scale score or APACHE scores were not reported in that study. Cervical SSSH developed acutely in our patient, she had admitted to emergency room with complaints of numbness and neck pain and arrest developed during her follow up. While atrophy, infarct areas, ventricular enlargement may be the MRI findings seen in NPSLE, the cystic lesion in the right frontal region and band-shaped hemorrhage field at cervical 2-3 level were not usual MRI findings.

While intra-cranial cystic areas is not a common finding in NPSLE, the cystic areas may be interpreted as cystic areas accompanied by infarct and atrophy and also they may be old resorbed cortical hematoma areas; the cystic areas detected on MRI were reported as cystic areas also histo-pathologically in the ratio of 100% (Sibbitt *et al*., 2009). Our patient's having SSSH which is rare in general population and cervical region's being a rare location makes this case special. We should state that vasculitis and vasculopathy should not be overlooked in NPSLE patients besides our suggesting anti-coagulant use and elevated INR as the first etiologic factor. We consider that this case report is important as it remembers us that unexpected complications may develop in patients who are being followed up due to NPSLE, only psychological disorders should not be suggested but also it should be kept in mind that many different cerebral lesions and complications may be seen together in vast majority of the patients. Image 2. Ventricular system and external sub-arachnoid fields are large, cystic area in the right frontal region

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