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Gynecological surgical approach to a patient with Sneddon's syndrome

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Abstract

In this case report, the management of a patient with Sneddon's syndrome who underwent laparoscopic hysterectomy for menometrorrhagia and multiple leiomyoma uteri is discussed. The clinical management of Sneddon's syndrome is based on four distinct aspects: Neurological, hematological, dermatological, and psychiatric. Their analysis can be a valuable tool in the preoperative and postoperative monitoring of these patients.

Keywords: Sneddon's syndrome, Menorrhagia, Laparoscopy

Introduction

Sneddon's syndrome, a non-inflammatory thrombotic vasculopathy which involves small and medium diameter arteries, manifests with Livedo reticularis and cerebrovascular diseases. The relationship between livedo reticularis and cerebrovascular symptoms was first defined by Kimming in 1959. Sneddon named this as a syndrome in 1965 with the reporting of 6 patients showing all the characteristics of a syndrome. The primary underlying event in the pathogenesis of the disease is a reduction in blood flow to the brain and skin, caused by small and medium diameter arterial occlusions. With an incidence of 4/1,000,000, it is usually observed in middle-aged females [1-3].

The etiopathogenesis of Sneddon's syndrome is unknown. Anti-cardiolipin antibodies (ACA) have drawn the most attention. In some reports, ACAs, especially IgG, were high in Sneddon's syndrome patients. Therefore, it is thought that ACAs could play a role in the pathogenesis of the disease [4-9]. Findings similar to those of antiphospholipid antibody syndrome, such as recurrent, low thrombocytopenia, have also been observed in various cases with Sneddon's syndrome [5, 10, 11]. In numerous reports, Sneddon's syndrome is evaluated under the main heading of antiphospholipid antibody syndrome [2]. However, the ACA levels of many Sneddon's syndrome patients remain normal, and elevated ACA levels are not deemed necessary for diagnosis [1, 12, 13]. This subject remains a matter of debate.

Livedo reticularis lesions, which are a characteristic finding of Sneddon's syndrome, have the appearance of a persistent, blue-purple colored, irregular mesh. They extend particularly from the trunk (gluteal region, and the lower lumbar region) towards the extremities (thighs, dorsal surface of the arms). Livedo reticularis generally emerges a few years or even a few decades before the onset of central nervous system (CNS) symptoms. As diagnosis is usually made after CNS symptoms, the age at diagnosis is the fifth decade. These lesions are progressive and exacerbated in the acute phase of a neurological attack [6, 14-16].

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Cerebrovascular symptoms usually occur secondary to ischemic attacks. In approximately 77% of patients, headache, seizures, hemiparesis, sight disorders and psychiatric disorders such as depression may be seen. The common CNS pathology in patients include temporary ischemic attack (TIA) and/or ischemic stroke. Despite widespread lesions in the CNS, clinical findings are not very severe due to the involvement of small and medium diameter arteries. In approximately 75% of patients, the symptoms are recurrent, generally in a similar form to the first symptoms [1, 10-14]. The first symptoms in Sneddon's syndrome generally occur in the reproductive years and disease activity tends to stabilize with menopause. The use of oral contraceptives can increase the disease. As Sneddon's syndrome is usually seen in females, female reproductive hormones are thought to play a role in the pathogenesis of the disease. It has also been reported that hypertension, observed in some patients with Sneddon's syndrome, may have a triggering role in the development of symptoms [1-6]. No effective definitive treatment has yet been defined. The use of oral anticoagulants, such as warfarin and clopidogrel, is widely accepted in the treatment of the disease [17-19].

We herein present the management of a patient with Sneddon's syndrome.

Case presentation

A 41-year old female presented with complaints of irregular menstruation and intermittent bleeding. She was diagnosed with Sneddon's syndrome priorly, and there had been several miscarriages associated with this disease (Gravidity:10, Abortus:9, Parity:1). While investigating the neurological findings of the patient, livedo reticularis lesions were noticed on the lower extremities, and the diagnosis had been previously made because of skin biopsies obtained from these lesions (Figure 1) and neurological findings. The patient was taking warfarin 5 mg once a day. Physical examination revealed livedo reticularis lesions on the whole trunk, gluteal region, and hands (Figure 2). The second toe on the right foot was amputated from the middle of the proximal phalanx (Figure 3), due to an arterial circulation disorder when the patient stopped taking warfarin for a lengthy period. In pelvic examination, there was passive bleeding, the cervix was of nulliparity type and normal. Transvaginal ultrasound examination revealed that the uterus was larger than normal. There was a subserous myoma of 8 cm in the fundus, extending 3 cm intramurally, which was compressing the cavity, along with numerous myoma. The patient was admitted for a planned laparoscopic hysterectomy. In routine tests, Hemoglobin was 6.7 g/dL, platelet count was $135x10^3/mm^3$ INR was 2.6, and blood pressure was normal. Erythrocyte suspension was transfused preoperatively for anemia. INR was checked daily to assess bleeding risk; warfarin was terminated, and enoxaparin treatment of 2 x 0.6 ml was started.

To reduce the risk of intraoperative bleeding to a minimum, INR was expected to fall to below 1.5. However, after starting the enoxaparin treatment, a progressive decrease was observed in thrombocytes as a side-effect. In addition, during this period when warfarin was terminated, the patient showed psychiatric problems such as insomnia and anxiety. Headaches became more frequent and the skin lesions became more evident. The planned operation was scheduled earlier due to the exacerbation of the syndrome. With Hb 11.8 g/dL and platelet count $46x10^3/mm^3$, three units of thrombocyte suspension were transfused, and the patient was admitted for surgery. Laparoscopic hysterectomy was performed because of the lower morbidity taking into consideration potential problems which could be experienced in the postoperative period (wound site problems, thrombosis caused by late mobilization). No complication developed intraoperatively.

Figure 1: Skin biopsy from livedo reticularis lesion of the patient (HEx100)

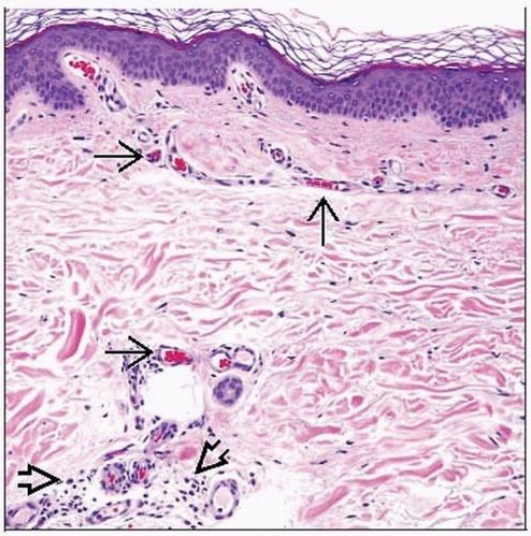


Figure 2: Livedo reticularis lesions on the patient's hand

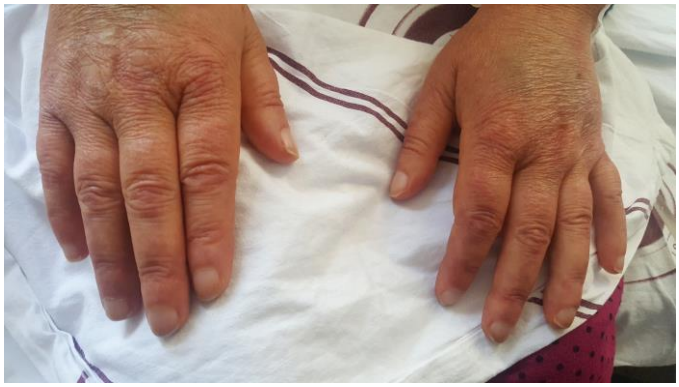
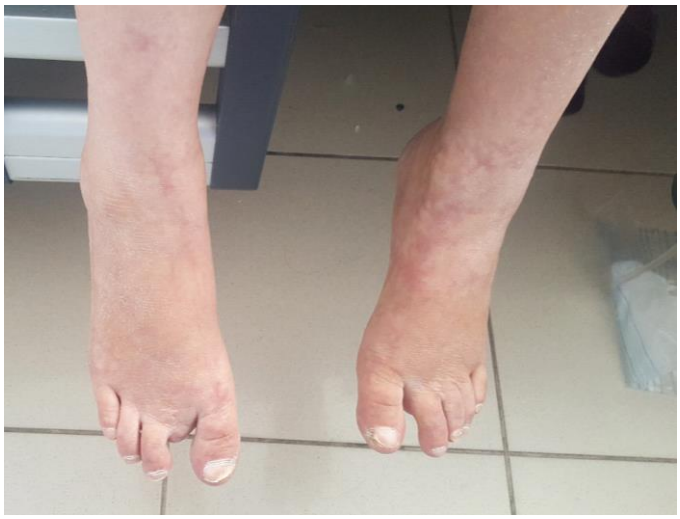


Figure 3: The second toe on the right foot was amputated from the middle of the proximal phalanx.



In the postoperative period, 200 milliliters of hemorrhagic drainage was observed in intra-abdominal drain in the first 24 hours. At 8 hours postoperatively, enoxaparin was

started and continued at 2 x 0.6 ml. Despite thrombocyte transfusion, platelet count was $33x10^3$ /mm^3, which necessitated six more units of thrombocyte suspension transfusion. The follow up PLT count was $27x10^3$/mm^3. Precautions of frequent mobilization and anti-embolism stockings were taken, and as bleeding control had not been achieved, enoxaparin was terminated, and warfarin was re-started. For the purposes of immunosuppression, 100 mg dexamethasone was administered. The vital signs of the patient were closely monitored, which revealed a sudden increase in blood pressure. Systolic blood pressure was 170 mmHg, and nifedipine 60 mg twice a day was started, after which the blood pressure decreased and stabilized. A short time after the addition of nifedipine to the treatment, the color of the livedo reticularis lesions on the hands and feet of the patient was observed to fade. The patient was monitored for 5 days without the administration of enoxaparin and warfarin. Blood tests obtained after terminating enoxaparin revealed a progressive elevation in thrombocytes. During this period of observation without enoxaparin, the patient reported double vision. Neurological examination and cranial magnetic resonance imaging **(**MRI) were performed, which revealed no neurological pathologies. Crying episodes started to accompany the complaints of insomnia and anxiety which had begun preoperatively. Thinking that this could be a psychiatric and cognitive symptom related to the syndrome, the Psychiatry Department was consulted. As the patient stated that she saw insects on the walls, especially at night, delirium was suspected, and 2 x 1 drops of haloperidol was administered daily. In the evaluation made after 2 days of haloperidol use, the complaints of the patient had receded. After ensuring that bleeding control had been obtained, warfarin was re-started. Blood follow-up was continued until the INR returned to the desired level. When thrombocyte value reached 129,000 and the INR value was 1.8, the patient was discharged with recommendations.

Discussion

Sneddon's syndrome is rare with clinical manifestations of various systems and well-defined clinical and laboratory findings. Based on our experience in this patient, the symptomatology is based on neurological, hematological, dermatological, and psychiatric aspects, as defined in literature. Therefore, essential points requiring special attention in the clinical and surgical management of a patient with Sneddon's syndrome were noted.

Anticoagulants are included in the treatment of Sneddon's syndrome [17-19]. In the adjustment of the INR level to reduce the risk of intraoperative bleeding, warfarin used in the treatment of the syndrome must be terminated preoperatively. In the current case, warfarin was terminated by checking INR daily. To reduce the risk of intraoperative bleeding to a minimum, we waited until the INR fell to below 1.5.

Low molecular weight heparin (LMWH) should be initiated to protect the patient against thrombosis when warfarin is stopped [1-3]. In the current patient, 2 x 0.6 ml enoxaparin was started when warfarin was terminated.

After starting enoxaparin, a progressive drop was observed in thrombocyte count as a side-effect. Appropriate blood products were prepared preoperatively as a precaution against the possibility of bleeding. It must not be forgotten that thrombocytopenia could develop in patients as a complication of LMWH, so platelet levels must be closely monitored with blood tests and blood products should be prepared preoperatively.

Before ceasing warfarin treatment, the lesion borders must be marked for more accurate monitoring of the lesions. In the current patient, there was no change in the size of the lesions, but they became more evident than before.

Taking into consideration wound site problems that may develop and the risk of thrombosis when the patient is immobile, the operation process should be accelerated and if possible, laparoscopy should be preferred [1-3]. With the consideration of potential problems that may be experienced postoperatively, a laparoscopic hysterectomy was performed in our patient for lower morbidity.

We placed one intra-abdominal drain during the operation and monitored it for postoperative bleeding. Within the first 24 postoperative hours, 200 milliliters of hemorrhagic content drained. Taking a history of warfarin use and the risk of thrombocytopenia into consideration, the drain should be placed intraoperatively for the purpose of monitoring intra-abdominal bleeding.

Vital signs must be closely monitored for a hypertensive status that can occur with the combination of the nephrological effects of the syndrome and the effects of the anesthetic substances administered intraoperatively [1]. We observed a sudden increase in blood pressure in our patient. As systolic blood pressure was around 170 mmHg, 60 mg of nifedipine was administered twice a day, after which regulation of blood pressure was achieved. A brief time after the addition of nifedipine to the treatment, the livedo reticularis lesions on the hands and feet began to fade.

The psychiatric leg of the syndrome must not be forgotten, and if necessary, psychiatric consultation should be requested [1, 10, 11]. In the current patient, crying attacks started to accompany the complaints of insomnia and anxiety which had begun preoperatively. During the psychiatric interview, the patient stated that she saw insects walking up the wall, especially at night, which led to the diagnosis of delirium and appropriate treatment was administered.

To avoid missing neurological problems, frequent neurological examinations should be performed [12-14]. On the seventh postoperative day, our patient reported double vision. She was evaluated with neurological examination and MRI, which revealed no neurological pathologies.

Conclusion

The results obtained from close clinical monitoring of the current patient in the preoperative and postoperative periods will be of guidance for the management of other patients with Sneddon syndrome. Nevertheless, there is a need for examination of a greater number of patients to confirm the data obtained.

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