Case Report

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Anesthetic Management of a Rare Case of Primary Sjogren's Syndrome for a Gynecological Surgery

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Abstract

A 44 years old female patient with primary sjogren's syndrome presented for surgical repair of third degree uterovaginal prolapse with cystocele and rectocele. Other associated medical problems included hypertension, Type II diabetes mellitus and hypothyroidism. Vaginal hysterectomy with colpoperineorrhaphy and levataroplasty was done under lumbar subarachnoid block and the perioperative course was uneventful. Herein, we describe our experience of anesthetic management of a case of primary sjogren's syndrome for an elective surgical procedure.

Keywords: Sjogren's syndrome; Schirmer's test; Immunosuppressants; Spinal anesthesia.

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Introduction

Sjogren's syndrome is a chronic systemic autoimmune disorder. The overall prevalence of sjögren's syndrome is 0.1% to 0.4% in the general population, with a female to male ratio of 9:1. It is usually diagnosed in the fourth or fifth decade of life eventhough, it can manifest in all age groups.[1] The clinical hallmarks of Sjogren's syndrome are dryness of the cornea and conjunctiva or keratoconjunctivitis sicca and dry mouth or xerostomia due to the lymphocytic infiltration of lacrimal and salivary glands respectively. It is classified either as primary (PSS) when occuring alone or secondary (SSS) when asscociated with other autoimmune diseases.[2] The disease remains fundamentally incurable and treatment is mainly symptomatic.[3] Due to the constantly evolving nature, overlapping qualities of each unique connective tissue disorder along with the multiorgan involvement and associated increased mortality rate the impact of these disorders on different systems and their management needs to be emphasized. Thus, in this case report we discuss about a female patient with primary sjogren's syndrome who underwent an elective gynecological procedure.

Case Report

A 44 years old female patient, homemaker by occupation presented with mass per vagina which was diagnosed to be third degree uterovaginal

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prolapse with Grade 2 cystocele and rectocele for which an elective gynecological surgery was planned. Her obstetric score was P4L4.

About 5–6 months back, she had one episode of fever which was completely relieved by antipyretics following which she developed arthralgia involving both small and large joints of upper and lower extremities associated with skin lesions and photosensitivity. Since then she also complains of persistent sensation of gravel in both the eyes for which she had to clear her eyes with water and dryness of mouth which compelled her to take frequent sips of water. Since, last two months, she had pain in the region of both the parotids and difficulty in swallowing solid foods. There was no history of hematemesis or melena. The patient denied any history of cough, hemoptysis or bleeding from any site. There was no history of oral ulceration, alopecia, analgesic abuse or any recent trauma. There was no history of similar or any major illness in her family. Schirmer's test was done for the patient and was found to be positive. Based on history, clinical examination, relevant laboratory investigation reports and as per 2016ACR/EULAR criteria[4] she was diagnosed as primary sjogren's syndrome and was on treatment with wysolone, methotrexate, hydrochloroquine and topical corticosteroids for skin lesions.

She was a diagnosed case of hypertension on treatment with amlodipine tablets since 5 years. She is recently diagnosed with Type 2 diabetes mellitus on oral hypoglycemic agents and borderline hypothyroidism not on any treatment. On personal history, she had no addiction or promiscuous behavior. Sleep, bowel and bladder habits were normal

Laboratory investigations and test reports were:

Investigations	Patient report	Reference ranges
Total leukocyte count (cells/cumm)	3970*	4000–11000
ESR (mm/hr)	47*	Male: < 15 Female: < 20
Thyroid stimulating hormone (μIU/mL)	4.97*	Adults: 0.3–4.5
Glycosylated hemoglobin (HBA1C)	6.7%*	Normal < 5.7 %
USG-abdomen and pelvis	Grade I fatty liver	
2d-ECHO	Hypertensive heart disease*	
Anti-nuclear antibodies	1: 920*	< 1:40
Anti-Ro/SSA	1.45*	< 1.0
Anti-La/SSB	7.25*	<1.0
C3 (mg/dl)	99	90–180
C4 (mg/dl)	21	10–40
Anti-phospholipidantibodies IgG (U/ml)	1.2	< 10
Anti-phospholipidantibodies IgM (U/ml)	0.9	< 10
Anti-smith antibodies	Negative	
Serum ceruloplasmin (mg/dl)	41.5	20–35

*Elevated/abnormal results

Preanesthetic evaluation was done and on examination, she was found to be moderately built with weight 55 kg, height 155 cm and BMI of 22.9 and well nourished. Erythematous skin lesions (mottled hypo and hyperpigmentation type) were seen on cheeks, neck and extensors of the forearm. Other general physical examination findings were within normal limits. Respiratory system and cardiovascular system examination was within normal limits. Central nervous system examination did not reveal any motor, sensory or autonomic neurological involvement. On airway examination, no features suggestive of anticipated difficult airway were found.

Different anesthetic options along with its risks and benefits were explained to the patient. Nil per oral status of 8 hours was confirmed and pantoprazole tablet was given the night prior to surgery as aspiration prophylaxis. After arrival in the operating room, multi-para monitors were connected and base line parameters recorded as heartrate of 78 beats per minute, noninvasive blood pressure 118/71 mm Hg, respiratoryrate 16 per minute, O_2 saturation of 100%. After placing the patient in sitting position, under aspetic precautions lumbar subarachnoid block was performed by mid line approach at L3-L4 intervertebral space using 25 G quincke needle and 18 mg of 0.5% hyperbaric bupivacaine injected after free flow of clear cerebrospinal fluid through the needle was confirmed. Immediately patient was positioned supine, motor block, sensory block assessed using modified bromage score and pinprick method respectively. Maximum sensory block level achieved was T10 within 5 minutes, then the patient was put in lithotomy position for the surgical procedure. Vaginal hysterectomy with colpoperineorrhaphy and levataroplasty was done and the surgical procedure lasted for 2 hours. The patients vital parameters remained stable throughout the

perioperative period without any pharmacological intervention and the estimated blood loss was about 500 ml. Humidified O_2 supplemented at 5 liters per minute. Total 1500 mL of crystalloids was transfused intraoperatively. Effect of spinal anesthesia lasted for 4 hours. Postoperatively pain was managed with intravenous injections of tramadol.

Postoperatively, foot exercises were started as soon as the motor block receded. Prophylactic anticoagulation and chest physiotherapy was also initiated. She was discharged on the fifth postoperative day without any complication. The patient was followed up regularly on out patient basis for 6 months and no complications were noted.

Discussion

Sjogren's syndrome is a well-known entity with multiple connective tissue disorders and hyper-gamma globulinemia.[5] The pathogenetic mechanism underlying these disorders is an inappropriate and excessive immunological reaction by the patients antibodies. It follows an indolent or slowly progressive course with the disease confined to the exocrine glands. However, in 30% of patients, it can involve neurological function (abnormal gait, autonomic dysfunction, seizures, movement disorder, ataxia/incoordination, insensitivity to pain, hyporeflexia, and even paraparesis or quadriparesis), immunological disorder (immune system anomalies, dysfunction autoimmunity), pharyngeal abnormality, respiratory tract modification (interstitial-like disease), chronic atrophic gastritis, celiac like disease, distal renal tubular acidosis, raynaud phenomenon and other skin anomalies. Occurrence of nonhodgkins lymphoma and mortality are higher in these patients. Secondary Sjögren syndrome occurs in 10 to 20% of patients with rheumatoid arthritis, systemic lupus erythematosus, and scleroderma. Various other diseases can imitate this syndrome including sarcoidosis, lipoproteinemia, and amyloidosis.

Preoperative assessment includes evaluation of constitutional illness, associated rheumatoid arthritis, interstitial pneumonitis, nephritis, vasculitis, peripheral neuropathies and lymphomas. Hypothyroidism and sleep disorders are relatively common.[6] Drug history particularly intake of systemic steroids, immunosuppressants and radiotherapy and its associated adverse effects if present should be recorded. Immunosuppressants are continued perioperatively while monitoring patient's leucocytes and platelet counts. Drugs with anticholinergic side effects like atropine, hyosine, phenothiazines, tricyclic antidepressants and antispasmodics should be avoided. Oral premedicants may provoke oesophageal injury by adhering to dryer walls of oesophagus[7], hence avoided. The enlarged parotid and submandibular glands may make the mask ventilation difficult. Laryngoscopy and intubation are complicated by xerostomia, poor orodental hygiene and temporo-mandibular joint arthritis. Hoarse voice is suggestive of crico-arytenoid joint involvement. Preoperative indirect laryngoscopy or fiberoptic assessment helps to plan intubation strategies or consideration of alternative regional techniques. Anesthesiologist must minimize infection by maintaining equipment cleanliness, using bacterial filters and prophylactic antibiotics. Inspired gases and intravenous infusates are warmed. The administration of anesthetic agents, hypnotics and local anesthetics must be done slowly and in presence of blood pressure monitoring because of the possibility of autonomic nervous system dysfunction. Ocular lubricants should be instilled frequently. Particular care is necessary to prevent positioning trauma associated with rheumatoid deformities. Adhesive tapes and ECG electrodes can cause epidermal ecchymosis or bullous eruptions. Thin and fragile skin makes venous cannulation difficult. Vasospastic irritability may provoke painful cyanotic reaction similar to raynaud's phenomenon.[8] Intraoperative vasodilatation necessitates colloids infusion for hemodynamic stability. Vigilant monitoring of respiratory embarassment due to crico-arytenoid inflammation and glandular enlargement[9] is required. Humidified oxygen facilitates mobilization of tenacious secretions as mucus plug inspissation can lead to bronchospasm and dyspnoea. All the above mentioned precautions were taken care of in our patient. Although, general anesthesia may be the most rapid and appropriate method in emergency surgical situations but in our patient, due to the presence of other associated medical problems and allergic predisposition for which she was on treatment with multiple drugs, we decided to avoid polypharmacy, unnecessary drug interactions and as the neurological examination findings, preoperative reports of coagulation profile were all within normal limits, as there was no surgical emergency conditions, we opted for spinal anesthesia. As the postoperative analgesic requirement in vaginal hysterectomy patients is relatively less, in our institution we do not practice epidural analgesia in these set of patients.

Preoperative aspiration prophylaxis, intravenous steroids administration and optimal preloading enabled us to manage the patient without aggravation of the clinical symptomatology. Effect of spinal anesthesia lasted for four hours, prolonged reversible sensory blockade is expected due to additional binding of local anesthetics and is useful for postoperative analgesia.

Conclusion

Careful preoperative assessment, intraoperative management tailored to the needs of the patient and surgeon reduce the perioperative risk to minimum and improve patient outcome.

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