

## Subinguinal Varicocelectomy Under Spinal Anesthesia in a Fahr's Syndrome Patient with Non-Obstructive Azoospermia

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### Abstract

Previously, Fahr's syndrome diagnosed 21 year-old male patient, consulted to urology outpatient clinic suffering from left scrotal pain. Physical examinations and Doppler tests results indicated left grade III varicocele. Left subinguinal varicocelectomy was performed safely under spinal anesthesia. This is the first and unique case showing co-existence of Fahr's syndrome and varicocele and also the first spinal anesthesia procedure in a Fahr's syndrome patient. In this report we tried to show that varicocelectomy can be performed under spinal anesthesia safely on Fahr's syndrome patients.

**Keywords:** Fahr's syndrome; Varicocelectomy; Spinal anesthesia

### Introduction

Fahr's syndrome is a rare disease that was identified by Fahr in 1930 for the first time, depending on calcium and phosphorus metabolism disorder, characterized by double sided, symmetrical intracranial calcification [1]. Although its clinical findings are parkinsonism, dystonia, tremor, chorea, dementia and mood disorders, it can occur with urological symptoms such as overactive bladder [2-3]. Previously, operation under general anesthesia in a Fahr's syndrome patient was reported in the literature [4]. However, no operation under spinal anesthesia was mentioned in any the literature.

In this report we intended to assess and evaluate varicocelectomy and infertility case and operation under spinal anesthesia for a Fahr's syndrome patient after his consent.

### Case Report

15 year-old male patient consulted to neurology outpatient clinic with dizziness, nausea, vomiting and hypertension complaints. Listening his story we realised that his parents were relatives. Physical examinations showed no evidence of neurological disorder and there was no sign of urinary incontinence or overactive bladder. Routine test results showed that Calcium (Ca) levels were in normal range, slightly low ionized Ca and high level of phosphorus. Patient's parathormone level was in the lower limit.

Calcifications were detected in the bilateral thalamus, globus pallidus and putamen by cranial magnetic resonance imaging and computed tomography. Urinary tract ultrasonography result was normal in terms of urinary stone disease. In 24-hour urine sample, calcium and phosphorus levels were low. Creatinine clearance and

Doppler ultrasound results were normal, that was done because of patient's high level of blood pressure during headache.

Bender Gestalt visual perception and number sequence learning tests were administered to assess the cognitive functions and there were no signs of cognitive impairment. Depending to all these findings, and results the patient was diagnosed as Fahr's syndrome. Then, follow-up procedure was scheduled for the patient for regressive complaints. At 21 age, the patient came to our urology outpatient clinic complaining for left scrotal pain and the left grade III varicocele was detected after physical examination.

Left testicle size was smaller than the right one. Doppler ultrasonography showed left sided dilated veins, reaching 4.5 mm in diameter at the widest part and detected prominent reflux flow, correlated with varicocele. Patient's spermogram result indicated azoospermia and ejaculate volume was 4 milliliters. These findings were evaluated as non-obstructive azoospermia. Hormonal evaluation results were; FSH: 6.59 IU/L, LH: 6.95 IU/L, Estradiol 27.96 pg/ml, Prolactin: 10.94 ng/ml, Total Testosterone: 351.07 ng/dL.

Genetic analysis was evaluated as 46XY (normal karyotype). Left subinguinal varicocelectomy surgery was planned due to all these findings. The patient was persuaded for spinal anesthesia and after getting his written consent he was scheduled for surgery. Blood pressure was 126/72 mmHg, heart rate 89 beat/min, and SpO<sub>2</sub> was 97%, mallampati score was 1, and ASA score 2 in the operation room. Preventive measures and preparations were ready for any possible intubation problems: with various of size masks, tubes, blades and lma. The patient was positioned carefully for spinal anesthesia and 0.2 mg/kg. Bupivacain was administered intrathecally at the level of L4-L5. Left subinguinal varicocelectomy operation lasted for 45 minutes, and hemodynamic course was stable. Patient was observed in recovery unit and was sent to clinic after motor block was ended. Routine catheter was not inserted.

The patient urinated after six hours and was discharged from the hospital on postoperative second day. There was not any remarkable problem associated with wound healing.

## Discussion

Fahr's syndrome, associated with calcifications in the basal ganglia, is thought to be a rare neurological disease with 1/1.000.000 prevalence [5]. Because of abnormal accumulation of calcium in the basal ganglia, thalamus, dentate nucleus, cerebral cortex, cerebellum and structures such as the hippocampus, that controls movement and many cases start with extrapyramidal disorder. Speech disorders, cerebellar symptoms, neuropsychiatric symptoms are observed in more advanced stages [6]. There were cases with no neurological findings. The disease usually tends to start in the 3<sup>rd</sup> and 4<sup>th</sup> decades. Any disorder is not detected in routine blood tests and hormonal tests [5]. Genetic studies of Fahr's disease may suggest that abnormalities in the chromosomes 2, 8 and 14 [7-9].

Previously coexistence of Fahr's syndrome and varicocele was not reported in the literature. This is the first case showing coexistence of Fahr's syndrome and varicocele. In our case varicocele was detected by physical examination along with Doppler ultrasonography. Hormonal evaluation because spermogram result indicated non-obstructive azoospermia and genetic evaluation were normal. There is a need to evaluate Fahr's syndrome patient's without varicocele spermograms to find out if non-obstructive azoospermia is due to Fahr's syndrome or varicocele. Yet considering Fahr's syndrome may increase calcium stone formation due to disturbances in the metabolism of calcium, urinary tract ultrasonography was performed, but the stone in the urinary tract could not be screened.

Due to these metabolic disturbances, anesthesia management should be planned carefully taking the risk of malignant hyperthermia into account. In addition, the probability of difficult intubation owing to calcium metabolism disturbance should be borne in mind. For early diagnosis of malignant hyperthermia due to volatile anesthetics and cardiac effect, careful monitoring, invasive blood pressure follow up and blood gas investigation should be carried out. As personality changes and impairment in mental functions is marked in Fahr's syndrome and the etiology of intracerebral calcification cannot be identified, we believe that general anesthesia is safer. In these patients,

preparations should be made for difficult intubation. Previously inguinal herniorrhaphy surgery under general anesthesia was reported in the literature in a patient with Fahr's syndrome [4]. Our patient underwent spinal anesthesia safely and no complication was observed. Spinal anesthesia application for varicocelelectomy surgery in a patient with Fahr's syndrome is the first case in the literature.

## Conclusion

This is the first case showing coexistence of Fahr's syndrome and varicocele and also first spinal anesthesia application in a Fahr's syndrome patient. In this report we showed that varicocelelectomy under spinal anesthesia can be performed for Fahr's syndrome patients safely.

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