The Greek E-Journal of Perioperative Medicine 2022;21(a): 45-50
Ελληνικό Περιοδικό Περιεγχειρητικής Ιατρικής 2022;21(a): 45-50
ISSN 1109-6888

Case Report

Caesarean section in a parturient with Behçet's Disease: Case report and literature review

Tsianos V[1a*], Stamatakis E[1b], Valsamidis D[1b]

[1]*MD, MSc, PhD Anesthesiology*

[a]*Department of Anesthesiology, Evaggelismos General Hospital of Athens, Greece*

[b]*Department of Anesthesiology, Alexandra General Hospital of Athens, Greece*

**Correspondence: Tsianos Vasileios, Ipsilantou 45-47, Athens, Greece, PC10676, email: vtsiano@gmail.com, tel: 00306947815783*

ABSTRACT

Caesarean section in a parturient with Behçet's Disease: Case report and literature review.

Tsianos V, Stamatakis E, Valsamidis D

Behçet's Disease is a chronic autoimmune disorder manifesting mainly with mucocutaneous ulcers (mostly in the oral cavity and the genitalia), ophthalmic lesions and systemic vasculitis. Neuro-Behçet is a severe though rare form of the disease. Behçet's Disease effect on pregnancy is controversial with higher rates of miscarriage, but with symptoms remission during peripartum period. Mouth scarring from ulcers can complicate general anesthesia rendering intubation impossible and neuro-Behçet's CNS complications can avert general techniques. Here, we present the case of a C-section in a parturient with this uncommon disease and the anesthetic considerations. Literature review is also discussed.

Keywords: Behçet's Disease, Neuro-Behçet, caesarean section, general anesthesia, airway management

INTRODUCTION

Behçet's Disease (BD) is considered a relapsing chronic autoimmune inflammatory disorder manifesting mainly with mucocutaneous ulcers, ophthalmic lesions and systemic vasculitis. Mucocutaneous ulcers predominantly occur in the oral cavity and the genitalia[1]. Alt-

hough rarer, any organ system can be affected, with reports of Central Nervous System (CNS), lungs and the cardiovascular system being involved as well[2]. Involvement of the CNS, reported also as neuro-Behçet, constitutes the most serious form of the disease[3]. Neuro-Behçet is not common, but due to its difficult diagnosis, cases may be underdiagnosed[4].

Pregnancy, as it does with many autoimmune inflammatory disorders, influences disease's course with each women experiencing various manifestations of the disease during each trimester[5]. In some, flair-ups occur in the second or third trimester or in the immediate postoperative period, while in others an increase in drug requirements is reported[6]. Behçet's Disease effect on pregnancy is controversial with higher rates of miscarriage on the one hand, but with symptoms' remission during peripartum period on the other[7]. While the disease may be remitted during pregnancy, complications should always be considered prior to an operation. Mouth scarring from ulcers could possibly complicate or even make impossible to ventilate and/or intubate[8] and if neuro-Behçet is present, major CNS complications, such as myelitis, could become life-threatening[9]. Pharmacotherapy is another factor that can complicate management; i.e. colchicine can impair liver and kidney function[10].

Here, we present the case of a C-section in a parturient with this uncommon disease and the anesthetic considerations. Literature review is discussed. Written consent was taken from our patient for publication of this case report.

CASE REPORT

A 41-year-old nulliparous woman, at a gestation age of 37 weeks; 3 days, 168 cm tall and 90 kg, was presented for elective caesarean section. The gestation was the result of the fourth in vitro fertilization (IVF) attempt. Her American Society of Anesthesiologists Physical Status (ASA-PS) was considered as II. From her history; she had BD, factor V Leiden thrombophilia (heterozygote) and was an active smoker. Behçet's Disease was diagnosed 14 years before when genital ulcers were developed along with lymphadenopathy, oropharyngeal ulcers and CNS symptoms reported as a burning sensation in the spine. She was initially treated with cortisone and then colchicine up until the day of the CS. The last 10 years she reports only spinal symptoms, with last flair being 2 years ago. Though, MRI showed no active lesions in the CNS. During pregnancy she reported sporadic episodes of spinal symptoms, with completely absence of them the last four gestational weeks. Factor V Leiden thrombophilia was diagnosed 4 years ago when the IVF attempts started. During pregnancy she was treated with acetylsalicylic acid and low molecular weight heparin; last doses were omitted as per institutional protocols.

The parturient had not been seen in a pre-

anesthetic assessment clinic, so assessment was performed on the day of the operation. Extensive discussion with the parturient and the surgical team was made regarding the anesthetic options and due to the possible CNS involvement; the decision was made in favour of general anesthesia. Mallampati score was II, 5 cm mouth opening, thyromental distance 6 cm and no obvious or reported mouth lesions. After 3 min preoxygenation Induction was performed with propofol 2,5 mg/kg and rocuronium 1mg/kg. Endotracheal intubation was achieved uneventful (Cormack-Lehane I) Five minutes later; a healthy 3460gr boy with Apgar score 8 was delivered. Total operation time was 50 minutes. Patient was discharged three days post-operatively with no evidence of BD exacerbation.

DISCUSSION

Behçet's Disease is a rare autoimmune disorder of the connective tissue and small vessels[1,2].

Few cases have been published in literature reporting the anesthesia management of the BD patient, and only a handful of them are concerning the pregnant population. In one of them the general anesthesia approach was preferred[11], while in others neuraxial techniques were more applicable[12,13]. Either approaches are appropriate in certain circumstances, yet individualisation is the key. Behçet's Disease itself can guide management. For example, presence of genital ulcers is a good reason for caesarean delivery. Regarding anesthesia management in such patients, careful considerations must be taken due its multifactorial nature. Whether a general or regional anesthesia approach will be preferred, decision should be based upon clinical and laboratory findings and it must always be individualised. CNS involvement due to its difficultness in diagnosing should never be excluded and taken always into consideration when deciding the type of anesthetic technique.

In the pre-operative period, if time enables, deep investigation of the organs involved in each patient should be top priority, especially if CNS involvement is likely. Thorough review of MRIs or CTs could help exclude active neuro-Behçet with cord compression, raised ICP and risk of herniation[14]. Echocardiography and ECG need to be reviewed as cardiovascular involvement is also possible and anesthesia management can become quite challenging. There is a report in bibliography of a parturient with BD and arrhytmogenic right ventricular dysplasia in which regional technique was preffered[13]. Arterial Blood Gas (ABG) analysis along with spirometry and chest X-ray could help clarify the presence and severity of respiratory involvement. Thorough oral examination should be performed for detection of ulcers or scarring. Oropharyngeal ulcers and lesions may bleed during laryngoscopy and the presence of scar tissue can minimise mouth open-

ing[8]. Gupta et al reported a case of a BD requiring repeated surgeries with general anesthesia and endotracheal intubation and found that airway anatomy was compromised due to recurrent lesions and subsequent scarring[15]. A retrospective study of Noel et al found that BD symptoms seem to improve during pregnancy, especially in patients treated with colchicine[16]. Attention need to be paid though in drug side effects such as renal and liver functions that can be altered with long term pharmacotherapy[1]. Blood urea nitrogen (BUN) levels are helpful in identifying nephrotoxicity. Moreover, chronic use of corticosteroids requires supplemental dosing intra-operative. Skin puncture, when placing intravenous catheters or performing regional techniques, can lead to a phenomenon called pathergy, which is the formation of an inflammatory nodule at the site of the puncture. Thus, it should be minimised and by some experts, regional techniques are better to be avoided. Deshpande et al reported a case of transverse myelitis as a pathergy response to spinal anesthesia[9]. Additionally, neuro-Behçet is a contraindication for neuraxial techniques. Patients that require regional anesthesia should be evaluated in detail with neuroimaging techniques. On the other hand, general anesthesia with endotracheal intubation can be quite challenging if extensive oral lesions and scarring is present. For elective cases an awake fibreoptic approach through the mouth or the nasal cavity is the best option[8]. In addition, careful choice of the anesthetic agents to be used should be made. If spinal cord lesions are symptomatic, use of succinylcholine can lead to hyperkalaemia.

In our patient, despite the negative MRI, symptoms from the CNS, reported as burning spine sensation, were the main BD sign. We considered that as a clinical contraindication for regional anesthesia. This led us to general anesthesia with endotracheal intubation, since preoperative tests for prediction of difficult intubation were negative and also oral lesions or scarring were not present.

CONCLUSION

Uncommon medical conditions can pose a great challenge for the anesthetist. Behçet disease due to its versatile clinical nature needs to be assessed on a case-by-case manner and anesthesia management should be individualised. Both regional and general anesthesia are appropriate if meticulous investigation of signs and symptoms is performed. Special attention needs to be given for oral lesions and CNS involvement.

Additional materials: No

Acknowledgements:

Not applicable

Authors' contributions:

TV drafted the paper and is the lead author. SE contributed to planning and the critical

revision of the paper. VD contributed to planning and the critical revision of the paper.

Funding: Not applicable.

Availability of supporting data:

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Ethical approval and consent to participate: No IRB approval required.

Consent for publication:

Patient consent obtained

Competing interests:

The authors declare that they have no competing interests.

Received: February 2022, Accepted: February 2022, Published: May 2022.

REFERENCES

1. Sakane T, Takeno M, Suzuki N, et al. Behçet's Disease. N Engl J Med 1999; 341:1284–91.
2. Kaklamani V, Vaiopoulos G, Kaklamanis P. Behçet's Disease. Seminars in Arthritis and Rheumatism 1998; 27:197–217.
3. Al-Araji A, Kidd DP. Neuro-Behçet's disease: epidemiology, clinical characteristics, and management. The Lancet Neurology 2009; 8:192–204.
4. Kalra S, Silman A, Akman-Demir G, et al. Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. J Neurol 2014; 261:1662–76.
5. Orgul G, Aktoz F, Beksac MS. Behcet's disease and pregnancy: what to expect? Journal of Obstetrics and Gynaecology 2018; 38:185–8.
6. Marsal S, Falga C, Simeon CP, et al.Behcet's disease and pregnancy relationship study. Rheumatology 1997; 36:234–8.
7. Jadaon J, Shushan A, Ezra Y, et al.Behçet's disease and pregnancy. Acta Obstet Gynecol Scand 2005; 84:939–44.
8. Bhardwaj M, Singh K, Taxak S. Oral Scarring in Behcet's Disease - An Airway Concern. J Anesthe Clinic Res 2012; 03.
9. Deshpande DM, Krishnan C, Kerr DA. Transverse myelitis after lumbar steroid injection in a patient with Behcet's disease. Spinal Cord 2005; 43:735–7.
10. Finkelstein Y, Aks SE, Hutson JR, et al.Colchicine poisoning: the dark side of an ancient drug. Clinical Toxicology 2010; 48:407–14.
11. Koca E, Şayan H. Management of Anesthesia in a Pregnant with Behçet's

Disease. Kafkas J Med Sci 2017; 7:166–8.

12. Metodiev Y, Walker T. Anaesthetic considerations in a parturient with Behcet's disease. International Journal of Obstetric Anesthesia 2018; 33:93–4.
13. Giuri PP, Gonnella GL, Catarci S, et al. Anesthetic management of a parturient with Behcet's disease and suspected arrhythmogenic right ventricular dysplasia. Korean J Anesthesiol 2020; 73:75–6.
14. Jorizzo JL: Behçet's Disease. Neurologic Clinics 1987; 5:427–40.
15. Gupta A, Banerjee N, Sharma J, et al. Repeated surgeries in a patient with Behçet′s disease, what changes to expect? Anesth Essays Res 2013; 7:279.
16. Noel N, Wechsler B, Nizard J, et al. Behçet's Disease and Pregnancy: Retrospective Study of BD and Pregnancy. Arthritis & Rheumatism 2013; 65:2450–6.

Publisher's Note

The publisher remains neutral with regard to jurisdictional claims in published maps and institutional afliations.

Citation: Tsianos V, Stamatakis E, Valsamidis D. Caesarean section in a parturient with Behçet's Disease: Case report and literature review. Greek e j Perioper Med. 2022;21(a): 45-50.