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Anaesthetic consideration in a ruptured ectopic pregnancy with neurofibromatosis

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Abstract

The neurofibromatosis are autosomal dominant diseases that have widespread effects on ectodermal and mesodermal tissue. The most prevalent member of the group is neurofibromatosis type 1 (NF1), which can impact all physiological systems and has a range of severity. The distinctive lesions of the condition are called neurofibromas, and they can develop in the larynx, oropharynx, and neuraxis. These lesions can make laryngoscopy and tracheal intubation challenging. Pulmonary pathology encompasses cystic lung disease and pulmonary fibrosis. The cardiovascular manifestations of NF1 include hypertension, which may be associated with phaeochromocytoma or renal artery stenosis. The gastrointestinal tract may also be affected by neurofibromas, and the duodenum may include carcinoid tumours. When assessing and managing patients for surgical operations, anesthesiologists must be aware of and take into account each of the multisystemic consequences of the condition. We report a successful case of spinal anesthesia for salpingectomy in ruptured ectopic pregnancy with NF 1.

Keywords: Neurofibromatosis, Salpingectomy, Neurologic

Introduction

Neurofibromatosis is a multisystem genetic disorder that is associated with cutaneous, neurologic and orthopedic manifestations. Type 1 neurofibromatosis (NF 1)is characterized by café-au-lait spots and benign cutaneous neurofibromas. Type 2 neurofibromatosis (NF 2) affects the central nervous system (CNS) via spinal cord tumors and bilateral vestibular Schwannomas.[1,2]

Neurofibromatosis is characterized by different types of mutations of the NF-1 gene[3,4] .Approximately 50% of the NF-1 gene mutations result from de novo mutations[5-7]. The reported incidence of

neurofibromatosis (NF) in pregnancy varies from 1:5000 to 1:18.500[8].

Anesthesiologist must perform a thorough preoperative evaluation of every patient and have a well formed and comprehensive anesthetic plan to be prepared for any complication. Anaesthesia for pregnant patient with neurofibromatosis is not well documented.Pregnancy in patients with neurofibromatosis can result in multiple complications. During pregnancy, hypertension may be exacerbated, neurofibromas may increase in size and large pelvic or genital neurofibromas may cause preterm labour .

A study of Segal et al showed a significantly higher rate of extrauterine growth restriction, still births and need for caesarean sections in pregnant women with neurofibromatosis type 1 [9] Additionally, the increase in neurofibroma size may cause an increase in intracranial pressure. According to Dounas et al, the presence of increased intracranial pressure and spinal neurofibromas should be evaluated using CT or MRI before spinal or epidural anaesthesia despite the radiation risk to fetus. [10]

Case Report

A 30year-old primigravida with 8th week of gestation diagnosed with ruptured ectopic pregnancy posted for emergency salpingectomy.Past medical history revealed – NF 1, which was diagnosed 10 years ago with the characteristic Café-au-lait spots and benign cutaneous neurofibromas. There was no history of previous surgeries and drug allergies. On examination, her pulse was regular with 96 beats per minute, blood pressure 110/60 mmHg and respiratory rate 22 per minute. Her mallampatti grade was three with normal extension and flexion of the neck. There were no oral mass. Auscultation revealed normal heart and breath sounds. Inside the operating room, monitoring included 5-lead ECG, noninvasive blood pressure and SpO2 . sub-arachnoid space was located in the L3-L4 space with a midline approach. Lumbarpuncture was performed with a 25G Quincke's spinal needle and subarachnoid block established Using 2.5ml of 0.5% hyperbaric bupivacaine. Intra-op and post-op was uneventful.

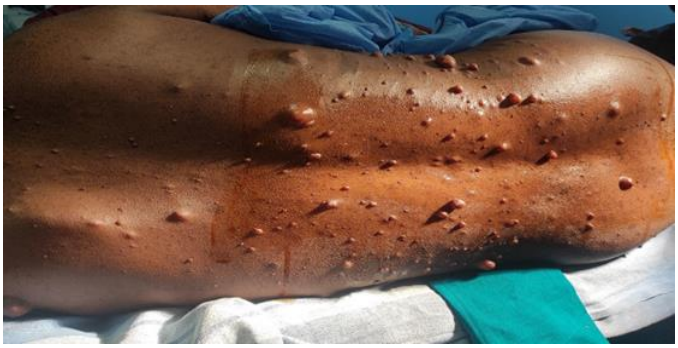

Fig 1: Benign cutaneous neurofibromas sized 1-2 cm spread throughout the entire body as well as the back

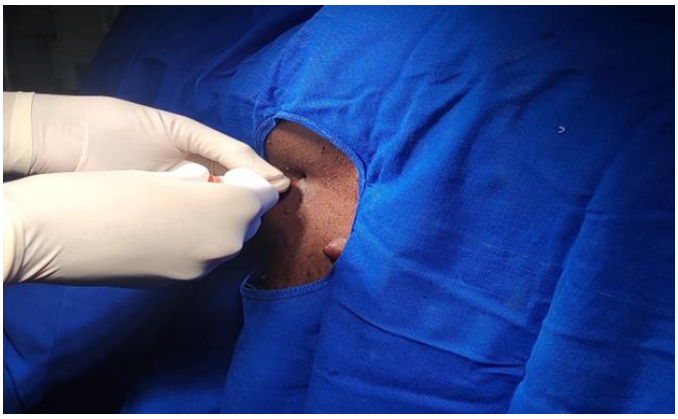

Fig 2 : spinal needle insertion in midline Btw L3-L4

Discussion

Neurofibromatosis is an inherited autosomal dominant disease , classified as type 1 and type 2. It is caused by a mutation of different chromosomes, type 1 in chromosome 17q11.2 and type 2 In chromosome 22q12.1. NF 1 is characterized by dermatological Lesions such as benign neurofibromas of the skin and café-aulait spots, is more common than NF 2. Neurofibromas also found in the oropharynx and larynx

and can produce difficulties with laryngoscopy and tracheal intubation .

Anaesthetic considerations of NF1 :

Central nervous system	Cerebral and spinal neurofibromas common. Increased incidence of epilepsy and learning disorders Cerebrovascular disease may co-exist
Airway	Neurofibroma of tongue, pharnyx or larynx may interfere with tracheal intubation Suspicion raised by history of dysphagia, dysarthria, stridor or change of voice
Cardiovascular system	Mediastinal tumours may result in superior vena caval obstruction Hypertrophic cardiomyopathy may occur
Respiratory system	Intrapulmonary neurofibroma, pulmonary fibrosis may produce cough and dyspnoea Scoliosis/kyphosis may compromise lung function
Gastrointestinal tract	Intestinal tumours may present with pain, gastrointestinal haemorrhage or perforation. Carcinoid tumours occur in duodenum and may result in jaundice and carcinoid syndrome
Genitourinary system	Neurofibromas may cause ureteric/urethral obstruction
Musculoskeletal system	Vertebral deformities or spinal cord tumours may make spinal/extradural techniques difficult

NF 2 characteristically has bilateral vestibular Schwannomas leading to gradual hearing loss . Other clinical features such as meningioma of the brain, Schwannoma of the cranial, spinal or peripheral nerve and juvenile cortical cataract can also exist. Neurofibromatosis appears to have no intrinsic effect on fertility; a high rate of spontaneous abortion and stillbirth has been reported. Because of the involvement of the CNS, regional anesthesia in NF 2 without careful preoperative examination can be extremely dangerous, many anesthesiologists prefer general anesthesia. On the other hand, regional anesthesia could be useful in NF 1 because CNS involvement is rare. Additional masses in the tongue, pharynx and larynx in NF 1 may interfere with intubation during general anesthesia . Regionalanesthesia is relatively safe in NF 1 so we opted for subarachnoid block in this patient.

Conclusion

The manifestations of neurofibromatosis are often mild, but there may be associated pathology of direct relevance. Anesthesiologists should consider the complex and diverse associated factors to provide optimal anesthesia. Whether to proceed with general or regional anesthesia in these cases must be viewed within the appropriate clinical context, like associated system involved and type of surgery.

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