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Case report

Subarachnoid block in a patient with extensive neurofibromatosis at the back

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Abstract Neurofibromatosis is an autosomal dominant disease characterized by widespread effects on different systems of the body. A 65 year old male who had to undergo surgery on the left thigh had neurofibromatosis type 1 with extensive neurofibromas all over his body, including the entire back. Patient was given spinal anaesthesia, at L3–L4 intervertebral space. Successful subarachnoid block was obtained and the surgery was performed successfully without any untoward complications.

1. Introduction

Neurofibromatosis is a group of hereditary diseases transmitted in autosomal dominant manner. It has extensive effects on ectodermal and mesodermal tissue and is the most common type of neurocutaneous syndromes. On clinical and genetic grounds, neurofibromatosis has been classified into two different types – neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2). Von Recklinghausen disease, also known as neurofibromatosis 1 (NF1), is characterized by multiple cafe-au-lait spots in the skin, multiple peripheral nerve tumors, and a variety of other dysplastic abnormalities of the skin, nervous system, bones, endocrine organs, and blood vessels [1]. The birth incidence of NF1 lies between 1 in 2500 and 1 in 3300 and its prevalence in the population is 1 in 5000 [2]. NF2 is defined by the presence of bilateral vestibular schwannomas. The birth incidence of NF2 lies between 1 in 33,000 and 1 in 40,000 with a prevalence within the population of 1 in 210,000 [3]. Both NF1 and NF2 can present as a non-generalized form in which only one body part is affected.

All this requires astute decisions on part of the anaesthesiologist confronted with such a case posted for surgery, as unique obstacles must be overcome for success.

2. Case report

A 65 year old male with body weight 58 kg and height 166 cm presented with a fungating ulcer for the past two months

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gradually increasing in size, which developed from one nodule present on the anterior aspect of his left thigh.

In the preoperative laboratory assessment all the investigations were within normal range. A pulmonary Function Test was done which was within normal range. A cranial and spinal MRI was advised to rule out central nervous system involvement, which showed no abnormality (Fig. 1).

On physical examination, the patient's entire body was covered with nodules of different sizes (Fig. 2). Oral examination revealed lesions on the tongue. To rule out deeper involvement of the airways, indirect laryngoscopic examination was done, the results of which were normal. The rest of the physical examination was normal. There was no associated deformity of the spine. There was no history of any other associated comorbidity. The patient consented for regional anaesthesia only, so it was decided to conduct the case under spinal anaesthesia. A written informed consent was obtained. On arrival in the operating room, an 18-gauge intravenous line was secured and all the standard monitors were attached. 500 ml lactated ringer's solution was administered, and the patient was placed in sitting position for subarachnoid block. After preparing the site, spinal anaesthesia was successfully performed between L3 and L4 intervertebral space, with a 26-gauge needle and 3 ml of 0.5% hyperbaric bupivacaine (Fig. 3). The patient was immediately placed in supine position and oxygen was supplemented via Hudson's mask at a flow rate of 6 l/min. After 5 min, the level of anaesthesia was checked and the surgery was started after 10 min. 1.5 mg of midazolam IV was administered to sedate the patient.

The surgery lasted for 1 h. All hemodynamic parameters were stable during the operation. The urinary output was 150 ml. The patient was then shifted to the postoperative recovery room where he was observed for 3 h and subsequently shifted to the surgical ward, with no postoperative complications reported. He was discharged on the 5th postoperative day.

3. Discussion

Neurofibromatosis is an autosomal dominant disease that has widespread effects on ectodermal and mesodermal tissue. Neurofibromatosis-1 (NF1) is the commonest member of the group which may affect different systems with varying severity. Neurofibromin, encoded by the gene NF-1 located on chromosome 17, is involved in tumour suppression. Inactivation of the NF-1 gene results in the production of a shortened version of

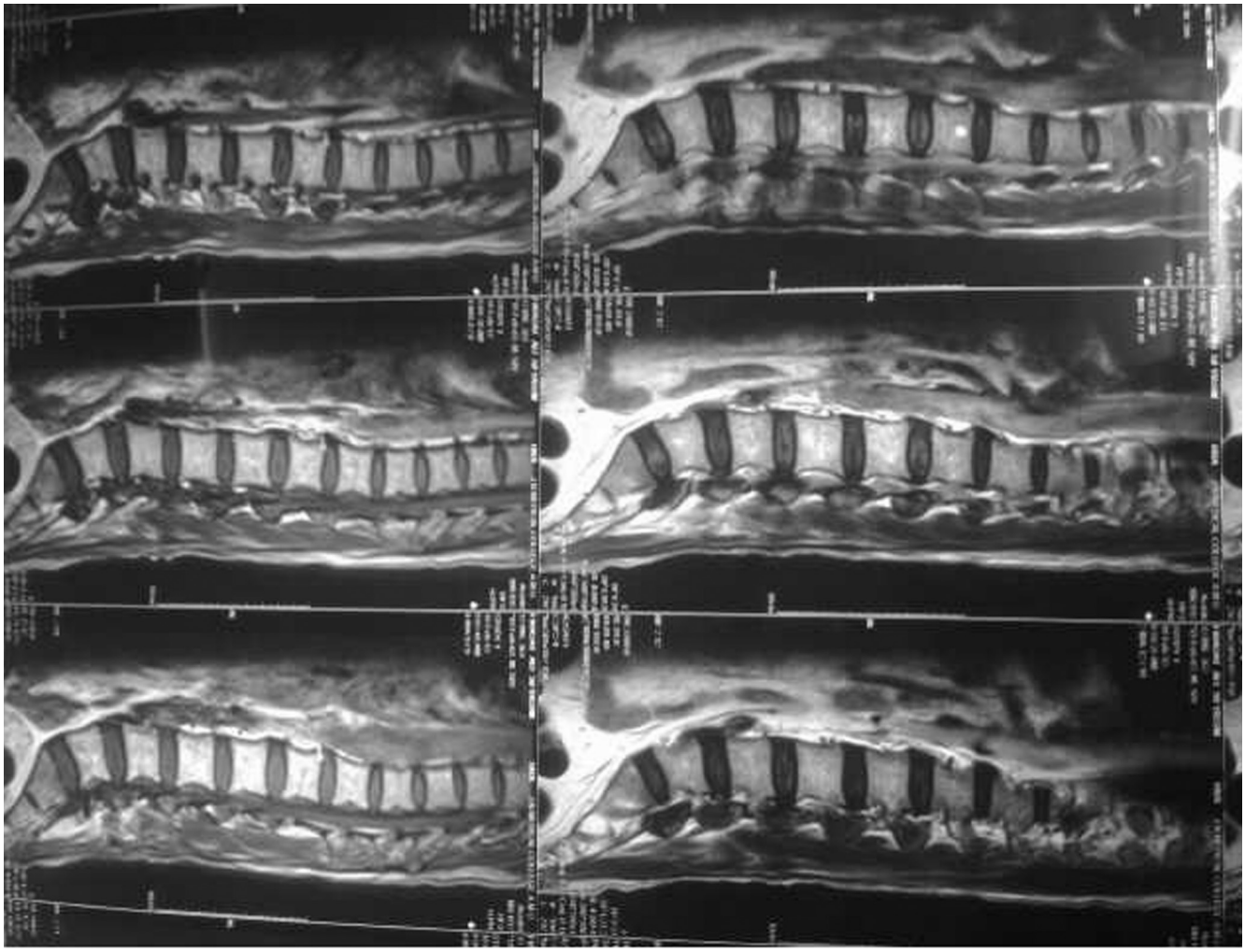

Figure 1 MRI showing no apparent abnormality.