TÜBİTAK

CASE REPORT

Turk J Med Sci
2008; 38 (5): 477-480

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The Anesthetic Approach in a Patient with Type I Neurofibromatosis with Multiple Deformities

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Abstract: von Recklinghausen disease is a type I neurofibromatosis originating from the nervous system. In this syndrome, there is abnormal deposition of neural tissue in the nervous system, endocrine system, visceral structures, and skin. We report the anesthetic management of a 31-year-old female patient with multiple thorax deformities who was diagnosed as von Recklinghausen disease. The patient was admitted to the Plastic and Reconstructive Surgery Department because of a huge subcutaneous neurofibroma on her right upper extremity. General anesthesia was considered a high risk for this patient on the basis of her laboratory results and physical findings. We planned brachial plexus block with axillary approach under sedation. The subcutaneous neurofibroma was not in contact with the nerve sheath; thus, regional anesthesia was our preference for this particular patient.

The anesthetic management of this patient was satisfactory and uneventful, and there were no complications.

Key Words: Anesthesia, neurofibromatosis, peripheral block

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Çoklu Deformiteli Tip I Nörofibromatozis Hastasında Anestezi Yaklaşımı

Özet: Von Reclinghausen hastalığı, sinir sisteminden kaynaklanan tip I nörofibromatozisdir. Bu sendromda sinir sistemi, endokrin sistem, viseral yapılar ve deride sinir dokusunun anormal birikimi mevcuttur.

Von Reclinghausen hastalığı tanısı olan, multipl toraks deformiteli, 31 yaşında kadın hastanın anestezik yaklaşımını sunmayı amaçladık. Plastik ve Rekonstrüktif Cerrahi bölümüne başvuran hastada sağ üst extremitede çok büyük subkutan nörofibrom mevcuttu. Fizik muayene ve laboratuar sonuçları ile genel anestezi bu hasta için büyük risk taşımaktaydı. Bu hastaya sedasyon eşliğinde aksiller yaklaşımla brakial pleksus bloğu planladık. Subkutan nörofibromun sinir kılıfı ile bağlantısının olmaması rejyonal anestezi kararı almamızı sağladı.

Restriktif pulmoner patolojisi olan hastada cerrahi için yeterli ve komplikasyonsuz anestezi uygulamayı başardık.

Anahtar Sözcükler: Anesthesia, neurofibromatosis, peripheral block

Introduction

Neurofibromatosis, which is a neuroectodermal disease, is characterized by congenital malformations of ectodermal tissues and their extensions. Neurofibromatoses are genetic disorders of the nervous system primarily affecting the development and growth of neural tissues and causing their subsequent growth. It is a syndrome caused by the abnormal deposition of neural tissue within the nervous system, endocrine system, visceral structures, and skin. Neurofibromatoses are divided into type I (NFI), also known as von Recklinghausen's disease, and type II (NFII) (1). While type II form of the disease affects the central nervous system, NFI has multi-organ system involvement. In these patients, the decision of anesthetic management is not easy. In the presented case with NFI, there was severe restrictive pulmonary pathology due to the significant deformities of the chest wall and lung. In this case report, anesthetic management during excision of the mass on the patient's right arm is discussed.

Received: November 12, 2007
Accepted: July 07, 2008

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Case Report

Physical examination of this patient, a 31-year–old female, revealed cachexia, severe kyphoscoliosis, and diffuse hyperpigmented skin eruptions (Figure 1). There was a mass

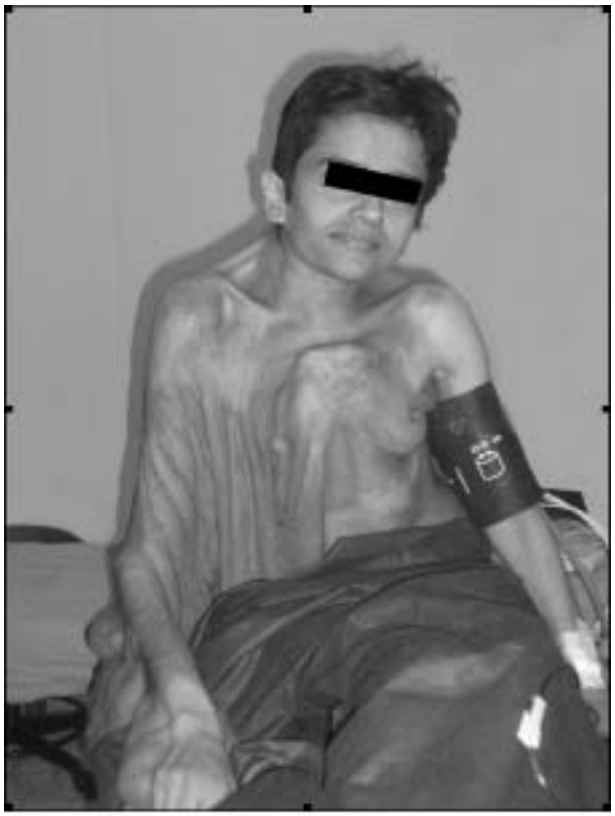

Figure 1. Patient diagnosed as neurofibromatosis type I

on the right arm surrounding the whole extremity and extending to the right part of the thorax. The patient was unable to lift her arm (Figure 2). Evaluation of the respiratory system determined inadequate inspiration and breath sounds were diminished (Figure 3). Three attempts to complete spirometric examination failed, but on the last attempt there were findings of severe restriction (FVC 0.35, FEV1 0.84, FEV1% 82.9, PEF 0.35, FEF25% 3.27, FEF50% 1.25, FEF75% 0.29). Blood gas analysis revealed pH: 7.41, PCO2: 41.9, PO2: 68.8, and SpO2Hb: 92.6%; the other values were within normal limits. Thorax computerized tomography (CT) showed severe kyphoscoliosis at the cervicothoracic column, corpus deformities at the cervical vertebrae and posterior fusion defect. Particularly on the right side, cutaneous neurofibromas at bilateral axillae were present. Mediastinal vascular structures were distorted and no mediastinal or hilar mass was observed. There was no active infiltration of pulmonary area bilaterally on radiographic examination. The right lobe of the liver was larger and scoliosis was seen at lumbar vertebrae on abdominal CT. Spinal canal anteroposterior and lateral diameters were considerably wider at lower cervical and upper thoracic levels of the vertebral column. In bilateral paravertebral areas of lower cervical and upper thoracic

Figure 2. Subcutaneous neurofibroma on the patient's right arm

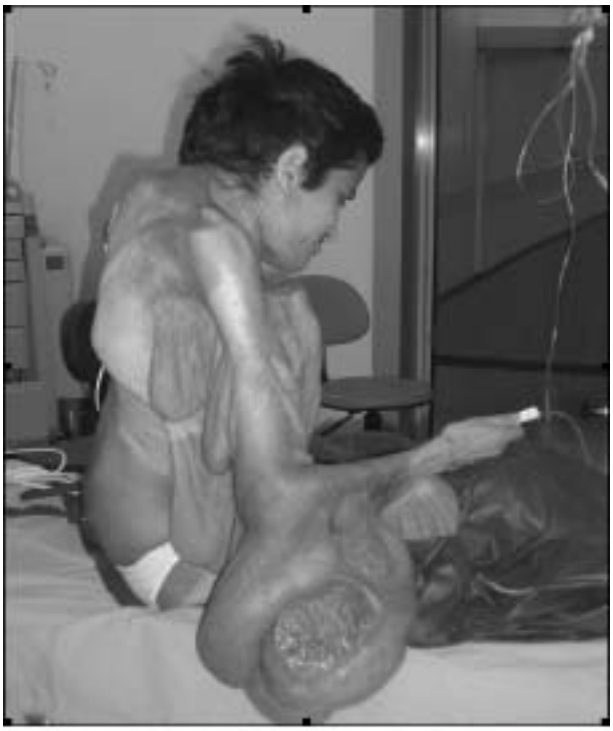

Figure 3. Thorax deformities of the patient.

levels, there were cystic densities up to approximately 5 cm on the right side and 9 cm on the left side.

The patient was evaluated for pheochromocytoma but surrenal CT scan and blood catecholamine levels were normal. Cranial CT scan was defined as normal and in consultation with the Neurology Department, no other pathologies were determined. For the anesthetic management, we planned brachial plexus block with axillary approach and sedation with propofol infusion during the excision of the neurofibroma (weight 2.5 kg) on the right arm.

The median and radial nerves were located by eliciting the maximal flexor and extensor response in the fingers of the hand, respectively. The response to the neurostimulation was weak. Totally 40 ml local anesthetic, 20 ml 2% prilocaine and 20 ml 0.5% bupivacaine, was applied. Due to the language barrier, we could only follow the patient's signals to speculate the pain level when controlling the block success. Surgery in the areas where there was insufficient blockage was achieved with 6 mg/kg/h propofol infusion and 50% 02-N20 mixture given by laryngeal mask airway. Hemodynamic values of the patient were stable intraoperatively. Sp02 saturation was between 93-96%. Postoperative blood gas analysis revealed pH: 7.4, PC02: 36.7 mmHg, and P02: 74 mmHg. In the postoperative period, the patient was observed for possible adverse effects; 1 mg bolus doses of morphine were given for pain relief until visual analogue scale (VAS) <3 was reached.

Discussion

The multiple sites of involvement in advanced neurofibromatosis determine the priorities for surgical preparation. Perioperative issues in patients with neurofibromatosis are: cranial CT/magnetic resonance imaging (MRI), pulmonary function tests, echocardiogram, blood urea nitrogen/creatinine measurement, detection of abnormal electrolytes, difficult airway management, respiratory compromise with high neuroaxial block, temperature control, and abnormal response to muscle relaxants (3). To identify masses, midline shift, or increased intracranial pressure and to demonstrate any potential risk of herniation, CT or MRI is recommended (3,4). In our patient, cranial CT and neurologic examination were normal. Because of kyphoscoliosis and vertebral anomalies in cervical and thoracal vertebrae, spirometry and arterial gas analysis was made (5). In view of the obstructive and restrictive signs, brachial plexus block was preferred to general anesthesia. When advanced pulmonary compromise is present, the possibility of prolonged postoperative mechanical ventilation must be considered (5). As the huge mass on her arm could cause significant tissue and blood loss, the reconstructive plastic surgeon decided not to excise the mass in the chest wall. In patients with neurofibromatosis disease in whom regional anesthesia is considered, the site for the block must be free of lesions and sufficiently normal anatomically to perform the block (3). After consultation with the surgeon, our final decision was to perform brachial plexus block that would provide sufficient anesthesia for this patient. We performed brachial plexus block with axillary approach using a combination of 2% prilocaine and 0.5% bupivacaine. We preferred prilocaine for its rapid onset and bupivacaine for the long duration of its local anesthetic effect. Based on the CT scan results of this patient and the physical examination by the surgeon, it was determined that the neurofibromas were not originating from the nerve sheath (6,7). There are five forms of neurofibroma; three of them - localized

cutaneous neurofibroma (when multiple), plexiform neurofibroma, and massive soft-tissue neurofibroma - are highly specific for NFI. Massive soft-tissue neurofibromas are worrisome in that they may mask malignant peripheral nerve sheath tumor arising from one of the mentioned neurofibromas (6-8). In this case, neurofibromas were localized subcutaneously and were without peripheral nerve sheath connection. With these findings, there was no contraindication for peripheral nerve block, so our decision was brachial plexus block with axillary approach. In this patient, blockage was performed at the border of the axillary subcutaneous neurofibroma according to the CT scan. We did not attempt interscalene brachial plexus block because of anatomic deformities. To our knowledge, there is no case report in the literature discussing peripheral nerve blocks in NFI patients.

For those patients in whom postoperative pulmonary support may be necessary because of severe deformities, regional anesthesia with brachial plexus block and sedation can serve as suitable anesthetic management to prevent perioperative and postoperative complications.

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