Clinical Reports

Epidural analgesia for labour in a patient with Charcot-Marie-Tooth disease

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Purpose: *We report a case of a parturient with peroneal muscular atrophy – Charcot-Marie-Tooth disease type 1 (CMT) – who received epidural analgesia for labour. The effects of pregnancy and labour on the course of CMT are reviewed, together with the current literature on the provision of epidural and general anaesthesia in this disease.*

Clinical features: *A 26-yr-old primipara with CMT was provided with epidural analgesia for labour. She experienced good pain control and suffered no untoward neurological sequelae.*

Conclusion: *Epidural analgesia, after thorough discussion with the patient, may be offered to parturients with CMT.*

Objectif: *Faire le compte rendu de l'analgésie épidurale administrée pour le travail à une parturiente porteuse d'atrophie musculaire péronéale de Marie-Charcot-Tooth type 1 (MCT). Les effets de la grossesse et du travail sur l'évolution du syndrome de MCT sont passés en revue ainsi que la littérature actuelle sur la pratique de l'anesthésie épidurale et générale pendant cette maladie.*

Éléments cliniques: *Une primipare de 26 ans porteuse de MCT a reçu une analgésie épidurale pour son travail. Le contrôle de la douleur a été efficace et aucune séquelle neurologique n'est survenue.*

Conclusion: *Après les explications d'usage, on peut offrir une épidurale à une porteuse de MCT.*

Key words

ANAESTHESIA: obstetric; epidural;
NERVE: Charcot-Marie-Tooth disease.

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Accepted for publication 30 June 1996.

Charcot-Marie-Tooth disease (CMT) – peroneal muscular atrophy – is the most commonly encountered of the hereditary motor and sensory neuropathies. The disease is divided into types 1 and 2, the former having its onset in the first decade, featuring foot deformities and being associated with lower nerve conduction velocities. The usual pattern of inheritance is autosomal dominant but recessive and x-linked patterns also occur.[1-3] Charcot-Marie-Tooth disease is known to pursue an aggravated course during pregnancy and labour in some patients. We present a patient with CMT who received effective epidural analgesia for labour with no untoward neurological sequelae. To our knowledge, this is the first case report of the use of epidural analgesia in CMT for this purpose.

Case report

The patient was a 26-yr-old primipara. Since age two she had had difficulty walking and had foot and toe deformities, including pes cavus. In childhood, she had numerous orthopaedic procedures to correct lower limb anomalies, all under general anaesthesia without incident.

At age eleven, after nerve conduction studies, she was diagnosed as having CMT type 1. There was no family history of this disease. Lower limb reflexes disappeared at the age of twelve. Since puberty, the disease had been stable and the patient worked as a laboratory technician. Apart from tiring more easily than her peers, she had no functional impairment, sensation being normal.

The pregnancy was complicated by type A1 diabetes and a small first trimester vaginal bleed. Notably, the CMT was not exacerbated by pregnancy.

At 30 wk gestation, she consulted the obstetric anaesthesia service about her choice of labour analgesia. She was informed about the lack of data on provision of labour analgesia in CMT and on the influence of epidural analgesia on this condition. The patient understood that her symptoms could become worse after delivery as

part of the natural progression of the disease. After an extensive discussion of the risks and benefits of the procedure, it was decided that epidural analgesia would be used if needed.

At 40 wk gestation, she presented in labour and requested epidural analgesia at 4 cm dilation. An epidural catheter was placed uneventfully at L_{2-3} and excellent analgesia was obtained. After receiving the epidural the patient was confined to bed. Her physiological responses to the blockade, both cardiovascular and neurological, were within normal limits. A total of 70 mg bupivacaine and 80 mg lidocaine was administered during the next six hours, when the patient had a spontaneous vaginal delivery of a 3,521 g infant, with APGAR scores of 9 at one and five minutes.

During the first six months after delivery, there was no worsening of neurological symptoms and the patient remained pleased with her choice of epidural analgesia.

Discussion

Charcot-Marie-Tooth disease usually presents in childhood with difficulty in walking and weakness of the lower limbs. Abnormalities of the feet and toes, including pes cavus, are invariably apparent. The usual course of the disease is a slowly progressive neuropathy with muscle wasting and loss of tendon reflexes early in life.

The potential for CMT to be exacerbated during pregnancy is well documented although this was originally not thought to occur.[4–6] In the series of 21 patients described by Rudnik,[4] 38% reported exacerbation of symptoms and not all returned to the baseline level after delivery.

Early age onset and severity of disease appear to correlate with a higher incidence of pregnancy-associated exacerbations. The labour and delivery process itself may also be linked to increased symptomatology.[4] Four patients in Rudnik's series reported new symptoms and/or worsening of existing symptoms after-delivery.

The cause of the pregnancy-associated exacerbations is a matter of speculation. Pollock *et al.*[5] proposed that endoneurial oedema may be responsible, secondary to pressure effects of the gravid uterus. There may also be hormonally-induced changes in the mucopolysaccharide content of ground substance. They contend that the loss of interstitial fluid post-delivery causes the usual postpartum improvement.

There are reports of the use of general anaesthesia for surgical procedures in patients with CMT. Succinylcholine should be avoided, as this might precipitate hyperkalaemia. Brian[7] reported a patient with CMT who developed extreme weakness and dyspnoea during pregnancy. Thiamylal, atracurium, nitrous oxide and enflurane were used to provide anaesthesia for Caesarean section. The patient's neurological status slowly improved during the first three postpartum months. A report of general anaesthesia for abdominal hysterectomy in a patient with CMT by Roelofse[8] emphasizes the need to avoid depolarizing relaxants and also cautions against the use of volatile anaesthetic vapours, despite the fact that malignant hyperpyrexia has not been noted in these patients.

The provision of epidural blockade for surgery has only been reported in two patients with this condition. Epidural anaesthesia was used for Caesarean section in a patient with CMT and mitral prolapse.[9] There is also a report of epidural anaesthesia for reduction of a femoral fracture in a 75-yr-old man.[10] Neither of these patients developed postoperative neurologic problems.

The arguments against the use of epidural analgesia in CMT are similar to those used in other progressive neurological diseases, such as multiple sclerosis. Data from prospective randomized trials are lacking. Fear of litigation should exacerbation occur could lead some to avoid regional anaesthesia in this situation. The issues must be thoroughly discussed with the patient before labour. It is important that she understand that the natural course of CMT may progress after delivery. However, there is no convincing evidence that epidural analgesia will affect CMT. We feel that a patient with CMT should be able to choose this superior form of analgesia if she wishes.

In summary, epidural analgesia was used without incident for labour analgesia in a patient with CMT.

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