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CASE REPORT

Oral Management of Steinert's Disease and Role of Anxiolysis

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ABSTRACT

Background: Myotonic dystrophy type I (DM1) is a genetic autosomal dominant disorder; malignant hyperthermia is a possible complication. It may occur following administration of some halogenated general anesthetics, muscle relaxants, or surgical stress.

Aim: The purpose of this case report is to evaluate the dental management of patients with Steinert's disease.

Case report: The patient needed dental extraction. A locoregional paraperiosteal anesthesia was performed using bupivacaine without vasoconstrictor and sedation with nitrous oxide. The syndesmotomy of the elements 3.1, 4.1, and 4.2 was executed. The elements were dislocated through a straight lever and avulsed with an appropriate clamp. The socket was courted, washing with saline solution, inserting a fibrin sponge, and applying sutures (silk 3-0).

Conclusion: Dental treatment of the patient with Steinert's dystrophy must be carried out under a hospital environment and the use of local anesthetic without vasoconstrictor and with use of nitrous oxide; anxiolysis is recommended.

Clinical significance: This case report describes the precautions to perform oral surgery in patients with Steinert's disease and emphasizes the role of anxiolysis to avoid episodes of malignant hyperthermia.

Keywords: Anxiolysis, Myotonic dystrophy, Oral management.

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BACKGROUND

Myotonic dystrophy (DM) is classified into two subtypes: DM type I or Steinert's disease and DM type II caused by the alteration of the zinc finger 9 gene on chromosome 3. Myotonic dystrophy type II or Steinert's disease is a genetic autosomal dominant disorder. The pathology is characterized by myotonia and progressive muscular weakness. It is associated with cardiac conduction disturbances and central nervous system defects. In 1992, the genetic mutation of the DMPK gene on chromosome 19 was identified as responsible for DM1, which codes for myosin kinase. The prevalence of the disease is 1/8,000. The expansion of the CGT triplet over 50 pairs in the noncoding region of chromosome 19 determines the pathology. The severity of the pathology is determined by the amount of expansion of the triplets.[1] In an unaffected individual 5 to 34 CTG repeats are present; in a situation of premutation 35 to 49 CTG repeats are present. They have an increased risk of inheriting the pathology. In affected individuals, more than 50 CTG repeats are present. Full penetrance alleles are associated with disease manifestations. Myotonic dystrophy type I is divided according to clinical severity and age of onset as congenital, childhood-onset, adult-onset, and late-onset/asymptomatic.[2] The congenital form has onset at birth and is characterized by reduced fetal movement, hypotonia infantile, feeding difficulties, and cardiorespiratory changes. In general, death occurs around the ages of 30 to 40 years. The childhood form is very difficult to diagnose as it has a negative familiarity. The patient has cognitive and learning deficits and a reduced intelligence

quotient (IQ). Alterations of cardiac conduction are frequent. The patient has weakness of the facial muscles and myotonia. The adult onset has an outbreak between 30 and 40 years. The main symptomatology is weakness of the distal striated musculature with reduction of manual dexterity. Facial weakness and deterioration are also present, giving the typical myopathic facial appearance. The flexor muscles of the neck and limbs are commonly involved. Muscle myotonia is very common; however, myotonia affects bulbar, tongue, or facial muscles, causing talking, chewing, and swallowing problems. There are changes in heart rhythm, cataract problems, and a higher frequency of insulin resistance. Moreover, alterations to the psychic sphere with alteration of IQ, apathy, lack of initiative, and social integration are present. The late onset presents with muscle weakness and myotonia.[3] Patients with myopathies, such as DM, are more susceptible after the administration of certain drugs or some stressful situations to malignant hyperthermia, a fatal complication.[4] Malignant hyperthermia is a skeletal muscle disorder given by a massive release of calcium from the endoplasmic reticulum usually caused by a genetic alteration of the RYR gene that codes for a protein that regulates calcium release. However, some neuromuscular diseases can alter the cellular control of calcium with mechanisms that have not already discovered, giving a susceptibility to malignant hyperthermia.[5] According to recent studies, there are few neuromuscular disorders associated strongly with malignant hyperthermia.[6] It is usually triggered by some inhalatory anesthetics (sevoflurane, halothane, etc.) and some specific neuromuscular relaxants, succinylcholine. Some studies show how intense physical activity or major stress, such as surgery, can trigger this fatal complication. Therefore, anxiolysis is important to reduce the surgical stress during a surgery and reduce the incidence of malignant hyperthermia crisis. These substances cause a massive contracture of skeletal muscles, glycogenolysis, increasing lactate and body temperature. The main clinical manifestations are acidosis, hypercapnia, tachycardia, hyperthermia, muscle rigidity, compartment syndrome, rhabdomyolysis with subsequent increase in serum creatine kinase concentration, hyperkalemia with a risk for cardiac arrhythmia or even arrest, and myoglobinuria with risks of renal failure. Therefore, this complication manifests itself mainly in the operating room during surgical interventions, in which the trigging drugs are used, or in the immediate postoperative period. It is treated by administration of dantrolene sodium that blocks calcium release at the initial dose of 2.5 mg/kg.[7]

CASE REPORT

The patient S.B. 36 years old with DM1 childhood-onset (of Steinert) needed multiple extractions of root residues at the "Tor Vergata" University Hospital in Rome. The patient was included in dentistry day surgery for the severity of the disease. After performing a dental examination, a preoperative examination conducted by anesthesiologists and a cardiologic survey was performed. The patient's parents brought into view the documentation released by the center of reference for the study of muscular dystrophy. The patient showed the first clinical disorder at the age of 18 (myotonic phenomenon in the hands with difficulty in holding). A genetic analysis highlighted that the patient had a positive paternal familiarity. The patient had limitation for the functional motor autonomy with difficulty in walking and an easy fatigue, both motor and respiratory. In relation to the related difficulty of breathing, the patient revealed a picture of initial restrictive respiratory insufficiency (framework lung volume 3.45 L; forced vital capacity 3.48 L; forced expiratory volume in 1 second 2.94 L; peak expiratory flow 4.54 L/s). From the cardiological point of view, the patient underwent a reveal implant in 2011. In 2016, an episode of atrial fibrillation with asymptomatic course was highlighted, for which cardioaspirin therapy was recommended. Since the last echocardiographic check, an ejection fraction of 46% has emerged. In addition to the underlying pathology, the patient underwent a thyroidectomy for thyroid carcinoma, cataract surgery, and surgery for disk herniation. The patient's collaboration was evaluated by the Frankel's Behavior Rating Scale and it was considered safer to treat the patient without sedation or general anesthesia. Blood examinations were performed; alteration of the normal parameters was not detected. He had to take eutirox and cardioaspirin. After a week the oral surgery was scheduled by fulfilling the following instructions: obtaining informed consent and warning the possible complications and performing the case study. The study complied with the Declaration of Helsinki. The patient presented with multiple decayed teeth and abundant plaque accumulation and a situation of diffuse parodontopathy. The patient has the presence of all 28 teeth. Preoperative indications were given to the patient: prophylactic antibiotic therapy with amoxicillin + clavulanic acid 1 gm to be started with a daily dose of 2 gm from the day before; perform fasting from midnight of the previous day; without interrupting the usual therapy. For the duration of the patient's hospitalization dantrolene sodium was present in the hospital.

On the morning of the operation, the patient was taken to the dental chair and a locoregional paraperiosteal anesthesia was executed by using bupivacaine without vasoconstrictor. The medical anesthesiologist provided sedation with nitrous oxide. The appropriate flow rate was established while the patient was breathing 100%

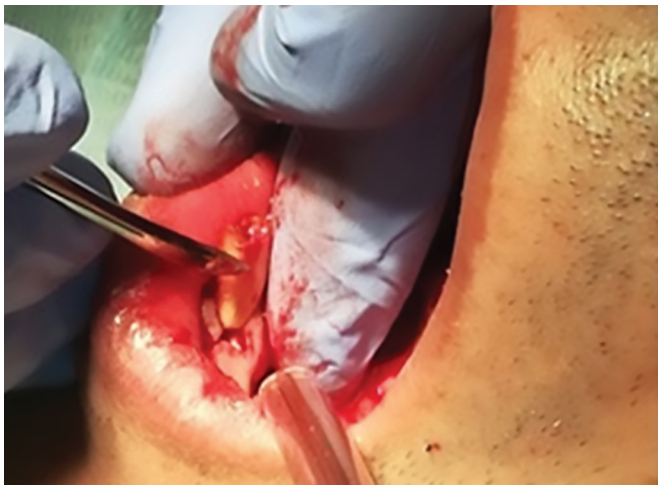

Fig. 1: Surgical phases

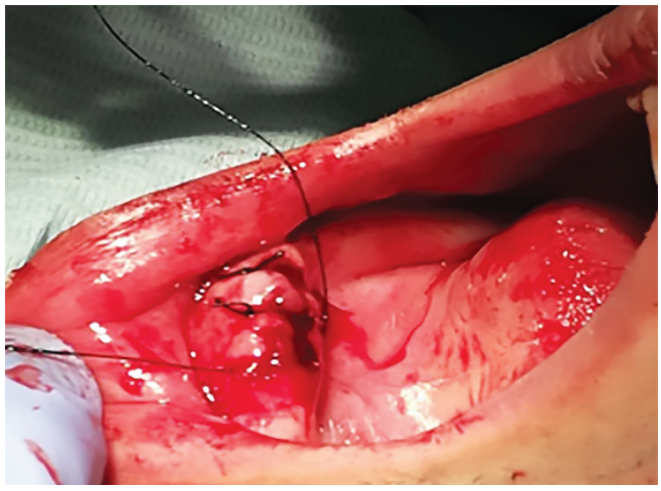

Fig. 2: Sutures of the socket

oxygen (5–6 L/min) for 1 to 2 minutes. Nasal breathing was encouraged, and the nasal hood was checked for leaks. The reservoir bag was monitored such that it remained uniform during breathing and did not expand or shrink. The percentage of N_2O was started initially at 10%. Then, it was titrated in approximately 10% increment rise every 60 seconds. The N_2O was titrated up to 40%. Constant communication with patient including physical, visual, and verbal contact was maintained. The dental operating room was previously held at an ambient temperature of about 18°C as guidelines. After performing anesthesia, the syndesmotomy of the elements 3.1, 4.1, and 4.2 was performed (Fig. 1). The elements were dislocated through a straight lever and avulsed through an appropriate clamp. The socket was courted, washing with saline solution, inserting a fibrin sponge, and applying sutures (silk 3-0) (Fig. 2). Postoperative indications of oral surgery were given to the patient's parents. The patient was monitored for about 4 hours postintervention in the ward, meanwhile intravenous paracetamol infusion was performed. The discharge took place in the afternoon of the same day. The patient did not present any problems and after 7 days the sutures were removed.

DISCUSSION

Malignant hyperthermia is a rare and potentially lethal complication for the patient with Steinert's disease. Therefore, during all minor surgical interventions, the use of locoregional anesthesia (oral surgery, gynecology, and ophthalmology) instead of general anesthesia is always recommended,. Surgical stress, according to various studies, can cause massive release of calcium and then can trigger a malignant hyperthermia crisis.[7] As regards the odontostomatological field is concerned, the patient must be treated in a protected hospital environment using local anesthesia without vasoconstrictor, anxiolysis in order to reduce stress and preventive postoperative analgesia; it is also recommended to have an operating room with low temperature.[8] Furthermore, dantrolene sodium, ready to use, must be available throughout the patient's admission. All of these preventive measures allow us to treat the patient with Steinert dystrophy, avoiding any possible complications.

CONCLUSION

The proposed treatment shows that patients suffering from Steinert's dystrophy must be treated in a protected hospital. The maneuvers are fundamental to reduce the stress of the surgical procedure as it could trigger the malignant hyperthermia crisis. Therefore, a possible sedation with protoxide is recommended, and all psychological maneuvers necessary to reduce the stress of the dental session as much as possible must be implemented. In conclusion, sedation with nitrous oxide is recommended to reduce surgical stress and decrease the possibility of the onset of malignant hyperthermia.

CLINICAL SIGNIFICANCE

This case report describes the precautions to perform oral surgery in patients with Steinert's disease and emphasizes the role of anxiolysis to avoid episodes of malignant hyperthermia.

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