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ANESTHETIC CHALENGES AND MANAGMENT IN PATIENT WITH ATAXIA- TELANGIECTASIA

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ABSTRACT

Louis-Bar Syndrome is a synonym for a very rare complex neurodegenerative disorder ataxia-telangiectasia (A-T). This is an autosomal recessive inherited disease that encompasses abnormalities in the cerebellum, multisystem degeneration, immunodeficiency, increased risk for malignancy and consecutive respiratory insufficiency. Most of the patients are radiosensitive and any exposing to ionization may lead to progression of the disease. Potential risks from anesthesia, mechanical ventilation, and postoperative complications in these patients have been insufficiently discussed in the literature. We present a successful anesthetic and respiratory management with one-lung ventilation in a patient with Louis-Bar Syndrome who underwent video assisted thoracoscopy (VATS) for recurrent pleural effusion.

Keywords: Ataxia-telangiectasia, Anesthesia, Management, One lung ventilation

INTRODUCTION

Ataxia-telangiectasia (A-T) is a rare (1:40,000 to 1:100,000 people worldwide), autosomal recessively inherited multi-system disorder [1,2]. The common clinical features of the disease include progressive neurological manifestations, telangiectasias (dominantly oculocutaneous, but also with possibility to be found in internal organs), recurrent sinus-pulmonary infections, a wide range of immunological abnormalities, an increased risk of malignancy, slow growth, incom-

plete pubertal development, diabetes, deformities of the lower limbs and scoliosis [2,3,4]. The median survival age is 25 years, and the majority of deaths are due to lung disease and cancer (22%) [3,4,5]. According to the clinical features of this disease it is hypothesized that surgery might be often needed for these patients, but due to the rarity of the disease, the exact incidence of surgery and anesthesia needed is questionable [3,4]. Therefore, no references or guidelines are present regarding the anesthesiologic management and approach to these patients.

Many aspects of the disease may interfere and increase the risk of perioperative and postoperative morbidity and mortality. Preoperative diagnostic assessments are usually limited [4,6]. Numerous anesthetics and anesthetic techniques directly influence the immunological and neurological level, as well as the muscles' strength. The deformities of the thorax may lead to difficult airway management. Immunodeficiency is a risk for postoperative severe infections [3,4]. Chronic respiratory insufficiency and common pulmonary comorbidities (pulmonal fibrosis) convey potential risks of intraoperative ventilation problems and increase the need for postoperative prolonged ventilator support [6,7]. Diabetes and glucose intolerance may lead to extreme variations of the glycemia with consecutive repercussions on the brain. Therefore, it is easily concluded that almost every feature of this disease is a potential anesthesiologic risk. Contrary to this, literature supporting any type of anesthesia techniques for these patients is very poor. Almost all data are based on a single case or a series of case reports [7,8,9].

Hereby, we report on successful anesthesia management of a patient with Louis-Bar Syndrome in Total Intravenous Anesthesia (TIVA), undergoing one-lung ventilation (OLV). Additionally, in this report we have tried to summarize the literature for this disease and discuss potential risks that could influence anesthesia and those which might help clinicians who are treating such challenging cases.

CASE REPORT

In publishing this case, written consent from the parents and consent from the internal ethical board of the Department for Anesthesia at the Medical Faculty in Skopje was given.

We present 20 years old male, height 157 cm and 50 kg body weight, presented for recurrent pleural effusion treatment with Video Assisted Thoracoscopy (VATS) under general anesthesia. At present, the patient was transferred to the surgery ward for treatment of pleural effusion that was diagnosed with ultrasound at the Clinic for Pulmonal Diseases and was suspicious for malignancy. Namely, the pleural effusion in the patient was several times aspirated but the condition had not been solved. Additionally, the patient had a constant cough and subfebrile temperatures during the past 3 months.

The patient's developmental history revealed a normal pregnancy, on-time birth, and normal growth during the first year of life. During the second year of life, the patient started having recurring lung infections and difficulties with motor development. By the age of 6, he was diagnosed with A-T. Since then, he began receiving immunoglobulins. To the knowledge of the parents, they had no consanguineous marriage, even though the family history was not fully investigated.

At presentation, the patient's physical status revealed a mask-like facial expression, facial and ocular telangiectasias, deformities of the feet, muscular atrophy, and kyphoscoliosis (which was not diagnosed by imaging technics and was never treated). A neurological examination showed dysarthria, difficult phonation, difficulty in swallowing, full range of extra ocular movements, slow up/fast phase nystagmus, difficulty with standing or walking by himself, intentional tremors with past pointing down going reflexes, and hypotonia on all extremities. The patient's nutritional status was satisfactory overall, but he was extremely precautious fed by the parents due to his inability to swallow.

The electrocardiogram was normal. Periphery oxygen saturation was 96% with ambient air. A dull sound was detected upon percussion of the right side. Auscultatory vesicular breathing was presented on the left side with shifting dullness, while high pitched, bronchial, and diminished breath sounds were present on the right middle and low thorax. Further diagnostical testing, x-ray and computer tomography, were contraindicated because of the primary disease, thus the diagnosis of effusion was made after additional ultrasound examinations and clinical signs. Arterial blood gases were not taken, as the patient was severely immunodeficient, so any additional invasive pro-

cedures might be a risk for opening new doors to infections.

Other than immunoglobulins and high doses of vitamin supplements, the patient had no anamnesis for chronic therapy.

From the laboratory findings, the patient had leukocytosis present WBC 17.8 x109/L; RBC 5.20 x109/L, HGB 140g/L and PLT 367 x109/L, with normal hepatic, degradation, electrolyte, glucose essays and secondary activated thrombocytosis on the hemostasis.

At this point in the consultation with the immunologist, prophylactic-therapeutic doses of ceftazidime (50mg/kg/iv q8hr) for 2 days and 10gr of immunoglobulins, as well as LMWH Clexane (1mg/kg sc) were prescribed. Two days later, WBC were down to normal values, and after careful evaluation and discussions between the immunologist, the pulmonary specialist, the anesthesiologist, the neurologist and the parents, the decision for a VATS evacuation of the effusion and biopsy was made.

Out of precaution, the patient fasted preoperatively for 12 hours. Upon entry of the theatre room, the patient was monitored with ECG, non-invasive blood pressure (NIBP), SpO2, and capnograph. Vital signs were as follows: NIBP (120/70mmHg), pulls rate of 93bpm, SpO2 93% on room air. The patient was preoxygenated with O2 (100%) for 5 minutes with a face mask. For premedicating, 1mg iv of midazolam was given. Remifentanil (Ultiva, GlaxoSmith Kline, UK), 2mg dissolved in 40ml saline, with a starting dose of 0.25-1 mcg/kg/min and slow increasing rates for every 10 minutes was initiated. After 15 minutes, infusion of propofol (Fresolol, Fresenius) was added until loss of reflex. The patient was successfully ventilated on a face mask and successfully intubated without muscle relaxant with a double lumen tube (Mallinckrodt 35 F-left). Placement of the tube was auscultatory confirmed. The maintenance of anesthesia was with remifentanil doses of 0.25mcg/kg/min and propofol. Ventilation was adjusted with 8ml/kg to have EtCO2 from 30-44mmHg at FiO2 50%.

After placement in a lateral decubital position, tube placement was once more confirmed. OLV was obtained via standard method and the respiratory rate was increased, and tidal volume lowered. After OLV was started, peak pressures went from 14mbar to 20mbar, EtCO2 was raised from 36 to 40 with SpO2 remaining from 91% to 95% during the surgery. Hemodynamics were stable during the interventions. Hypotension (90mmHg systolic pressure) occurred only for a period of 3 minutes and reacted to volume doses (100ml) of crystalloids.

A surgically small incision was introduced, the lung was visualized, a biopsy of the lung and culture of the pleural effusion was taken (for further investigation), 400ml of serosa hemorrhagic liquid was evacuated, and chest tube was placed. After the chest tube insertion, both lungs were ventilated with adequate re-expansion of the collapsed lung. Skin was closed, a dose of acetaminophen 1000mg (Apotel, Uni Pharma Leon Tsetis, Greece) and ondansetron (Setronon, Pliva, Croatia, Zagreb) 4mg were given. Surgery lasted for 30 minutes, shortly after stopping the infusions of propofol, the patient was fully awake, extubated with sufficient spontaneous ventilation.

Postoperatively, pain management was with acetaminophen 1000mg and ketoprofen (Lek-Skopje, Lek Ljubljana). Ceftazidime was continued, immunoglobulins were given during the first three postoperative days. The patient was sustained from any eating or drinking for the first 24 hours to elicit the risk of pulmonary aspirations, therefore amino acid infusion and glucose at 10% were given. Glycemia was controlled every 4 hours. After the third day, another ultrasound was done, and pleural effusion was not present. The patient was then transferred to the Clinic for Pulmonal Diseases without any complications. Further therapy and results from the biopsy showed chronic pulmonal fibrosis and cells with atypical mitotic potential. However, further treatment was conducted at other clinics and these are beyond our authority to evaluate them.

DISCUSSION

We present a case of successful anesthesia management in a patient with progressive signs of Louis-Bar Syndrome who underwent anesthesia for therapeutic and diagnostic VATS. To our knowledge, this is the only report of a patient with this disease who underwent OLV.

When we speak of A-T, we speak of a rare disease, but historically the records of the first recognition of the disease started in 1926 [10]. Further clinical pathological explanation of the disease can be found in from the 1940s to the 1950s, when it was considered a clinical

syndrome, named Louis-Bar Syndrome [11-14]. Genetic studies done in the 1960s have confirmed a high family incidence of the disease which is followed by multi-degenerative, multi-systemic affection. However, no gender or race predilection was found [15].

As an autosomal recessively inherited multi-system disorder, the disease includes ataxia, progressive neurological manifestations, telangiectasias, recurrent sinus-pulmonary infections, different immunological abnormalities, high risk of malignancy, slow growth, intellectual disability, incomplete pubertal development and different endocranial disorders [2-5].

According to the current knowledge of this disease, ataxia is considered generally as the first symptom for startup diagnosis. When ataxia is present, changes in the cerebellum should be considered. However, there are contradictory statements which argue that vasculature changes are not sufficient to account for the cerebellar degeneration. Thus it is postulated that metabolic changes account for the vascular and cerebral alterations in patients with A-T [16]. Children with A-T have delayed walking and standing onset, which may look like cerebral palsy. Therefore, ataxia as a single clinical feature is not enough to confirm the diagnosis [2]. On the other hand, progressive neurological deficit, especially in motor neurological functions, may be noticed at the age of three and may give a dough for the disease. As for our patient, development milestones were delayed at the age of three, after which abnormalities of the central nervous system occurred (dull face mask facial expression) dysarthria and prominent cerebral signs occurred (tremor, nystagmus, dysphagia) [15]. In this case report we discuss the clinical features especially because confirming the diagnosis is expansive (genetical testing for A-T) and available only in several centers around the world. We found it necessary to contribute to the global knowledge for this disease, especially when it is not confirmed.

Other features of this disease include different stages of immunodeficiency. Immunodeficiency may be seen in different lowered levels of immunoglobulins and their subclasses. The literature points out that mainly classes of IgA are affected, but not restricted to them. What is interesting is the fact that the immunological deterioration is constant and worsens as time passes [3]. Sedwick P. and Border E. [12] postulated that there is aplasia or hypoplasia of the thymic gland in A-T patients, but this is not always confirmed. As in our patient, immunodeficient replacement of immunoglobulins were often needed and given. The patient was treated preoperatively and three consecutive days postoperatively with immunoglobulins, as prescribed by immunologist.

Patients with A-T are prone to recurrent sinus-pulmonary infections. This may be attributed to several factors [3,7]. Firstly, immunodeficiency; secondly, low levels of immunoglobulins IgA that line the respiratory mucosa and act as a film for protection from pathogens; and thirdly, difficulty in swallowing and aspiration pneumonias as well as inability to have effective coughs. In our case report, the frequent pulmonal infections started in the second year of his life and even pulmonal fibrosis might be present. He was treated with prophylactic antibiotics even before the surgery. Chronical changes were confirmed on the histopathological findings.

Other signs for the disease are telangiectasias, mainly ocular or on sun-exposed skin [2]. Our patient had ocular telangiectasias, but as he was presented with serosa-hemorrhagically effusion, we even considered the possibility of telangiectasias found in the lungs. Unfortunately, during a literature search we did not find any strong evidence or cases of pulmonal telangiectasias in these patients. The lack of evidence might be due to another common feature in these patients. Namely, patients with Louis-Bar Syndrome have extreme sensitivity to radio diagnostic and imaging methods, thereby limiting the exposure to diagnostic tools. Knowledge of the disease implies that these patients have a presence of a specific gene that has an enzymatic prophylactic role in stopping the damaged cells from dividing and spreading. Therefore, in these patients, not just cellular but also humoral immunity is affected, and in the absence of protection for the damaged cells, they are highly radiosensitive. This is also one of the reasons why these patients are prone to malignancies [3-6]. Any imagining technique may contribute to the worsening of the condition, so relative contraindication is present. Since radiosensitivity was confirmed in our patient, we agreed in our multidisciplinary approach that noninvasive imaging techniques should be used, such as ultrasound and multiple aspirations of the pleural effusion and when no improvement was seen in the patient's state. The patient underwent anesthesia and diagnostic and therapeutical VATS. Therefore, we must state that in finding and se-

questering the gene as well as radiosensitivity are very valuable diagnostic tools, but the procedure is expensive and unavailable in all laboratories.

As for anesthesia in patients with A-T, very little data is present. In this manuscript we shall discuss anesthesia relaying and comparing the data of two studies done on the series of cases by Lockman JL and collaborates [8]. In their study, retrospective analyses were done, for both the clinical interventional and anesthetic management in a total of 21 patients who underwent 34 anesthesia for 41 surgical interventions. The authors have found that most of the cases had neurological, pulmonal, and immunological issues, like those in our patient. However, in their study age difference variations of the patients are from 6 to 33 years old. They report on intravenous induction of anesthesia in most cases and different volatile techniques of anesthesia maintenance (sevoflurane, isoflurane, halothane, desflurane, N2O) [8].

For our patient, not a single muscle relaxant was given, contrary to the aforementioned study, where non depolarizing muscle relaxants (NDMR) were given in 59% of the patients. Even though muscular dystrophy was noted in their study, patients had no residual block or need for prolonged mechanical ventilation after anesthesia. This simply means or suggests that the muscle plate is not affected with the disease.

What the literature proposes is that when a genetic mutation on some gene is present, the consequential mutation of other genes may be present. Hence, the interaction of genes responsible for malignant hyperthermia (MH) and A-T might be an issue. Only one genetic study is present that notices that image radiation interpolates with hyperthermia and A-T genes, even though this has not yet been confirmed in clinical practice [17]. Therefore, we cannot be sure whether other agents, like anesthetics, can introduce MH gene expressions, even though there are cases presented and anesthetized with agents that can trigger MH. Luckily, no case with MH due to anesthetic has yet to be reported on, so the assumption is that direct interpolating is not present. This may be suggesting that the choice of TIVA in patients with A-T is a more secure method of anesthesia, compared to others. This was confirmed in our case.

As for intubation in patients with musculoskeletal disorders and facial atony, it is proposed that airway management might be a problem, and this also translates to A-T patients. Since these patients have hypersalivation and dysphagia [14], intubation and pulmonal complications can be real. In our case, airway management and intubation went uneventful with the double lumen tube (DLT). Even though we cannot confirm any report and intubation in such a patient with DLT, there is confirmation that airway management and intubation is not a real problem for A-T. Furthermore, the respiratory profile in the study of Lockman JL et al. [8] shows similar changes that were noted in our patient (rises in peak inspiratory pressure, lowering of the mean end tidal volume and rise of EtCO2, although insignificant). Hypoxia was reported in one patient in PACU for their study which was not the case for our patient. According to PACU, they concluded that the ventilatory course in these patients is stabile despite pulmonary conditions. However, McGrath MS et al. [7] report that respiratory functional testing in these patients is good follow-up for pulmonary condition, so preoperative precondition of pulmonary function may be an imperative in these patients. Pulmonary functional testing represents another challenge. In a different study done by the same authors in seven patients with A-T, it was shown that admitting to ICU was due to respiratory insufficiency not after anesthesia [18].

When talking about prognosis in this patient and all patients with A-T, one must consider the severity of the disease, prophylaxis of a recurrent pulmonary infections, stage of the pulmonal fibrosis, as well as early diagnosis of malignancies. Proper surveillance and early diagnosis are essential for care approach. Malignancy treatment has additional limitation in correspondence with the immunodeficiency and radiosensitivity.

Most available treatments concentrate on immunoglobulins, prevention of recurrent respiratory treatments and restrictive pulmonary diseases. Novel treatments are suggested in relation to the tumorigenesis [19] like novel antioxidants, aminoglycoside antibiotics and some studies try the efficiency dexamethasone [20]. Besides this treatment, no novel studies have yet arisen in relation to surgery and anesthesia. Patient survival is dependent on the screening, attentive care, form of the disease, and complications. The median survival age is 25 years, but some patients have survived up to 45 years [4].

Even though no specific therapy available for patients with A-T is present, knowledge of the diagnostical and clinical features must be reconsidered with a multidisciplinary approach when there is need for anesthesia and surgery.

The limitation of this manuscript is that it is a single case report. It would be more consistent if several cases were presented, but as this is a very rare disease, the likelihood of having this patient in for anesthesia and surgery is also rare. Despite this, we must be aware that the knowledge of an anesthesia approach in these patients is limited; every single case improves the knowledge for an overall strategy. As we presented in our discussion, these patients have many clinical aspects where different types of surgery and anesthesia might be needed, thus every experience should be documented.

CONCLUSION

Ataxia Telangiectasia is a rear disease, and the challenges of surgery and anesthesia are numerous. The literature is scattered on types of anesthesia, we present here a challenging case of a one lung ventilation in total intravenous anesthesia in a patient with Louis-Bar syndrome. The OLV was uneventful. Our case presentation may lead to the conclusion that different anesthesia techniques, especially TIVA, can be used in these patients.

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Резиме

АНЕСТЕЗИОЛОШКИ ПРЕДИЗВИЦИ И МЕНАЏМЕНТ НА ПАЦИЕНТ СО АТАКСИЈА-ТЕЛАНГИЕКТАЗИЈА

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Синдромот Луј Бар е синоним за ретко комплексно невродегенеративно нарушување познато како атаксија-телангиектазија (А-Т). Заболувањето се наследува автосомно рецесивно и кај пациентите со ова заболување доминираат промени во церебелумот, мултисистемска дегенерација, имунодефициенција, зголемен ризик од малигнитет и, последователно на овие промени, и респираторна инсуфициенција. Најголем број од пациентите се радиосензитивни и какво било изложување на јонизација може да доведе до прогресија на болеста, со што дијагностичките методи за верификација на одредени промени кај овие пациенти се ограничени. Потенцијалните ризици од анестезија, механичка вентилација и постоперативни компликации кај овие пациенти не се доволно дискутирани во литературата. Претставуваме случај на успешен анестезиолошки и респираторен менаџмент со вентилација на едно белодробно крило кај пациент со синдромот Луј Бар, кој беше подлегнат на видеоасистирана торакоскопија (ВАТС) поради повторувани плеврални ефузии.

Клучни зборови: атаксија-телангиектазија, анестезија, менаџмент, вентилација на едно бело дробно крило