ANESTHETIC MANAGEMENT OF A CHILD WITH ATAXIA TELANGIECTASIA

Şahin M 1*
1. Erzincan University Medical Faculty Department of Anesthesiology, Turkey

Correspondence
Dr. Murat Şahin. Erzincan University Medical Faculty Department of Anesthesiology, Turkey
Email: muratsahin@erzincan.edu.tr


ABSTRACT

Ataxia telangiectasia (AT) is a rare, autosomal recessive, progressive syndrome. The syndrome includes progressive cerebellar ataxia, oculocutaneous telangiectasia, elevated α-fetoprotein level. The prevalence of AT has estimated to be one case in 40,000 to one case in 100,000. Here we report a case of AT becoming dental problem. This patient had typical clinical appearance of AT, α-fetoprotein level was also increased. Because of rarity of AT, we report this case in anesthetic management.

Key words: Ataxia telangiectasia, general anesthesia

INTRODUCTION

Ataxia Telangiectasia (AT) is a rare, autosomal recessive syndrome characterized by progressive cerebellar ataxia, pathognomonic oculocutaneous telangiectasia, recurrent sinopulmonary infection, variable humoral and cellular immunodeficiency, a high incidence of malignancy and hypersensitivity to ionizing radiation. High serum α-fetoprotein levels, retardation of somatic growth, gonadal dysgenesis and defective cell cycle checkpoints are also found 1-2. This disorder occurs world-wide, with an incidence of 1:40,000 to 1:100,000 of live births 3. The syndrome subsequently received the name of Louis Bar. Males and females are equally affected and there are no racial or regional preferences 4. Telangiectasia is usually found at corners of eyes, or on the surface of the ears and cheeks exposed to sunlight 4.

Most patient with AT have laboratory evidence of immune dysfunction, but systemic bacterial and opportunistic infections are uncommon. The discordance between the frequent laboratory expression of significant immune system abnormalities and the relatively infrequent clinical expressions of enhanced vulnerability to infection remains unexplained 5.

Mental retardation has been reported frequently as part of the AT syndrome 6. The disease heterogeneous, both clinically and genetically as shown by existence of four complementation groups (A,C,D,E) 4.
Anesthetic management of ataxia telangiectasia

CASE REPORT

We report on a 14 year old, 19 kg, girl becoming dental problem. Because of this problem, it was planned dental repairing by the pedodontic clinic. She was born at term from non-consanguinity, healthy parents. On physical examination, mental and growth retardation, bilateral ocular telangiectasia, muscular atrophy, cervicothoracal kyphoscoliosis was noticed. Chest and cardiac auscultation was no signs. Echocardiography was normal. On neurologic examination, choreoathetotic movement and dysarthria was noted. On laboratory findings, α-fetoprotein was 33.40 IU/mL(normal range 0-5.8 IU/mL). Hematologic and biochemical laboratory findings were normal. She monitorized and pulse oximetry, blood pressure and electrocardiogram data observed. She was pre-medicated with intramuscular midazolam, 2 mg and 0.25 mg atropine. Anesthesia was induced with propofol 30 mg, given intravenously and 2% sevoflurane. Muscle relaxation was obtained with 1 mg/kg atracurium and No. 6 cuffed endotracheal intubation tube was inserted in trachea. Difficulty intubation score was grade II. After that anesthesia was maintained with 1.5% sevoflurane and 60% nitrous oxide in oxygen. Hemodynamic parameters were stable during anesthesia. It was not observed any complication preoperative and postoperative period. Then child was recovered from anesthesia and transferred to her bed. She was discharged from clinic the next day without complication.

DISCUSSION

Some features of AT are concern to anesthesiologist and surgeon. These features are variable degree of immunodeficiency, hypersensitivity to ionizing radiation and frequent predisposition to cancer, mental and growth retardation. Cancer is 1,000 times more in AT than in the general population. Lymphoma and leukemia are particularly common. AT heterozygote is at increased risk of breast cancer. The cause of death in AT is often pneumonia or chronic lung disease which might result from defects in chewing and swallowing owing to progressive neurological impairment. Therefore aspiration risk may occur. As a result we have to be careful in anesthetic management and surgical process for sterilization. Aspiration risk does not have to be forget for patients with AT in preoperative and postoperative period. Because sinopulmonary infections are common, we have to be careful. Ionizing radiation has to be avoided if possible.

COMPETING INTERESTS

The author declares that the author has no competing interests.

REFERENCES


