Title: Anesthetic management and Peri-operative implications of Ataxia-Telangectasia

Authors: Andrew J. Iskander, MD
Justin Lockman, MD
Melania Bembea, MD, MPH
Thomas Crawford, MD
Howard Lederman, MD, PhD
Sharon McGrath-Morrow, MD
R. Blaine Easley, MD

Affiliations: The Johns Hopkins Hospital, Baltimore, MD

ABSTRACT BODY:

Introduction: Ataxia Telangiectasia (AT) is a rare autosomal recessive genetic disease resulting in progressive multi-system degeneration and characteristic findings of cerebellar ataxia, immunodeficiency, sino-pulmonary infections, interstitial lung disease and malignancy. Anesthetic management and risk remains relatively unknown in this medically complex group. We hypothesize that a systematic review of our patients with AT can characterize the anesthetic management, procedures, and potential complications related to AT.

Methods: The Ataxia-Telangiectasia Clinical Center (ATCC) database was cross-referenced with operative records (1995 to present) to identify patients with AT who have undergone anesthesia at our hospital. Records were reviewed for each operative procedure to obtain pre-operative assessment (age, ASA status, vital signs, co-morbid conditions), intra-operative issues (vent settings, ASA monitors, agents used, blood loss, surgical/diagnostic procedure, duration of procedure, duration of anesthetic), post-operative issues (complications – GI, respiratory, bleeding; duration of PACU/PICU stay, duration of hospital stay) and overall morbidity and/or mortality. Descriptive statistics were performed.

Results: Evaluation of the ATCC database (350 subjects) identified 21 patients (13 female/8 male) with AT who underwent anesthesia for 38 procedures. Mean age was 11.9 years (median 9.5; range 3 to 34). Median number of procedures requiring anesthesia per patient was 1 (mean 1.7; range 1 to 6). Preoperative evaluation occurred in 20/38 procedures. The most common “in-patient” procedure was line insertion/removal (7/38). The most common “out-patient” procedures were 10/38 orthopedic and 8/38 ophthalmologic. All “out-patients” 10/10 were recovered in the PACU and were discharged home within 24 hours of procedure. All “in-patients”, 11/11, returned to their pre-procedure location. There was only one death (>48 hours after procedure), all others survived to hospital discharge.

Conclusions: Though medically complex, patients with AT appear to undergo anesthesia at our center with minimal complications. The majority of “in-patient” procedures are related to co-morbid conditions of infection and malignancy. Future analysis will compare this cohort with a retrospective cohort of non-AT patients matched on age, sex, ASA status, procedure, procedural physician and anesthesiologist, to determine AT related perioperative risks.