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Cumulative Radiation Dose in Patients With Hereditary Hemorrhagic Telangiectasia and Pulmonary Arteriovenous Malformations

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Abstract

Purpose: To determine the cumulative effective dose (CED) of radiation from medical imaging and intervention in patients with hereditary hemorrhagic telangiectasia (HHT) who have pulmonary arteriovenous malformations and to identify clinical factors associated with exposure to high levels of radiation.
Methods: All patients with at least 1 pulmonary arteriovenous malformation were identified from the dedicated patient database of a tertiary HHT referral center. Computerized imaging and electronic patient records were systematically examined to identify all imaging studies performed from 1989-2010. The effective dose was determined for each study, and CED was calculated retrospectively.
Results: Among 246 patients (mean age, 53 years; 62.2% women) with a total of 2065 patient-years, 3309 procedures that involved ionizing radiation were performed. CED ranged from 0.2-307.6 mSv, with a mean of 51.7 mSv. CED exceeded 100 mSv in 26 patients (11%). Interventional procedures and computed tomography (CT) were the greatest contributors, which accounted for 51% and 46% of the total CED, respectively. Factors associated with high cumulative exposure were epistaxis (odds ratio 2.7 [95% confidence interval, 1.1-6.3]; $P = .02$), HHT-related gastrointestinal bleeding (odds ratio 2.0 [95% confidence interval, 1.0-3.8]; $P = .04$) and number of patient-years ($P < .0001$).
Conclusions: Patients with HHT are exposed to a significant cumulative radiation dose from diagnostic and therapeutic interventions. Identifiable subsets of patients are at increased risk. A proportion of patients receive doses at levels that are associated with harm. Imaging indications and doses should be optimized to reduce radiation exposure in this population.

Résumé

Objet: Déterminer la dose efficace cumulative de radiation émanant des examens d'imagerie médicale et des interventions chez les patients atteints de télangiectasie hémorragique héréditaire et présentant des malformations pulmonaires artério-veineuses, et définir les facteurs cliniques associés à une forte radioexposition.
Méthodes: Tous les patients présentant au moins une malformation pulmonaire artério-veineuse qui figuraient dans la base de données sur les patients d'un centre de soins tertiaires spécialisé en télangiectasie hémorragique héréditaire ont été identifiés. Une revue systématique des images informatisées et des dossiers électroniques de ces patients a ensuite permis de relever tous les examens d'imagerie ayant été réalisés entre 1989 et 2010. Enfin, la dose efficace a été déterminée pour chaque examen, puis a mené au calcul rétrospectif de la dose efficace cumulative.
Résultats: 3 309 interventions comportant de la radiation ionisante ont été réalisées chez 246 patients (âge moyen de 53 ans; 62,2 % de sexe féminin) représentant un total de 2 065 années-patients. Les calculs ont révélé une dose efficace cumulative variant de 0,2 à 307,6 mSv et une moyenne de 51,7 mSv. Une dose efficace cumulative supérieure à 100 mSv a été constatée chez 26 patients (11 %). Par ailleurs, les interventions et les examens de tomodensitométrie (TDM) ont eu une incidence décisive sur les résultats, représentant respectivement 51 % et 46 % de la dose efficace cumulative. Enfin, les facteurs suivants ont été associés à une radioexposition cumulative élevée : l'épistaxis (rapport

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des cotes de 2,7 [intervalle de confiance de 95 %, de 1,1 à 6,3]; $P = 0,02$), les saignements gastro-intestinaux liés à la télangiectasie hémorragique héréditaire (rapport des cotes de 2,0 [intervalle de confiance de 95 %, de 1,0 à 3,8]; $P = 0,04$) et le nombre d'années-patients ($P < 0,0001$).

Conclusions: Les patients atteints de télangiectasie hémorragique héréditaire sont exposés à une dose cumulative significative émanant des interventions à visée diagnostique et thérapeutique. Certains sous-groupes de patients présentent un risque accru. Une certaine proportion de patients a reçu des doses dont le niveau est associé à des effets nocifs. Les indications et les doses en matière d'imagerie doivent être optimisées afin de réduire la radioexposition que subit ce type de patients.

Key Words: Telangiectasia; Hereditary hemorrhagic; Arteriovenous malformations; Radiation; Ionizing; Radiation dosage; Multidetector computed tomography

Hereditary hemorrhagic telangiectasia (HHT), historically known as Rendu-Osler-Weber syndrome, is a rare hereditary disorder that may be complicated by arteriovenous malformations (AVM) in the brain, lung, gastrointestinal (GI) tract, and liver. Pulmonary AVMs (PAVM) are frequently present in HHT, occurring in 15%-45% of patients (Figure 1) [1]. Adults with untreated PAVMs are at risk of life-threatening hemorrhage and neurologic complications [1]. Routine screening for PAVMs in all patients with HHT is recommended, given the serious nature of complications that may occur and the availability of preventative treatment with transcatheter embolotherapy [2].

Given the potential for multiorgan involvement, as well as the role of interventional radiology, standard care for patients with HHT results in repeated imaging procedures, and patients may be exposed to a cumulative dose of radiation much higher than with the average patient. Unenhanced thoracic computed tomography (CT) is recommended to confirm the presence of PAVMs after an initial positive screening with transthoracic contrast echocardiography. Small PAVMs may be followed with repeated serial CTs because growth is demonstrated in up to 18% of small PAVMs, and these may eventually warrant treatment [2−4]. Significant PAVMs are treated with transcatheter embolotherapy, then follow-up CT is recommended at 12 months after treatment, and then every 1-3 years (depending on the size of residual PAVMs). Long-term follow-up is recommended to detect growth of previously small, insignificant PAVMs but also to detect reperfusion of embolized PAVMs [2].

However, high radiation exposure is an established cause of cancer, and there is evidence from observational studies of excess risks from fractionated exposures in the dose range that would be received from repeated CTs [5−8]. The purpose of this study was to quantify the cumulative effective dose (CED) associated with diagnostic, interventional, and follow-up imaging in a cohort of patients with HHT and with at least 1 PAVM, and to identify factors associated with exposure to high levels of diagnostic radiation. The lifetime radiation dose received by patients with HHT has not been studied previously.

Materials and Methods

Overview

To characterize radiation exposure in this population, all patients with at least 1 PAVM were identified from the dedicated patient database of a tertiary HHT referral center. Demographic and clinical data were obtained from the

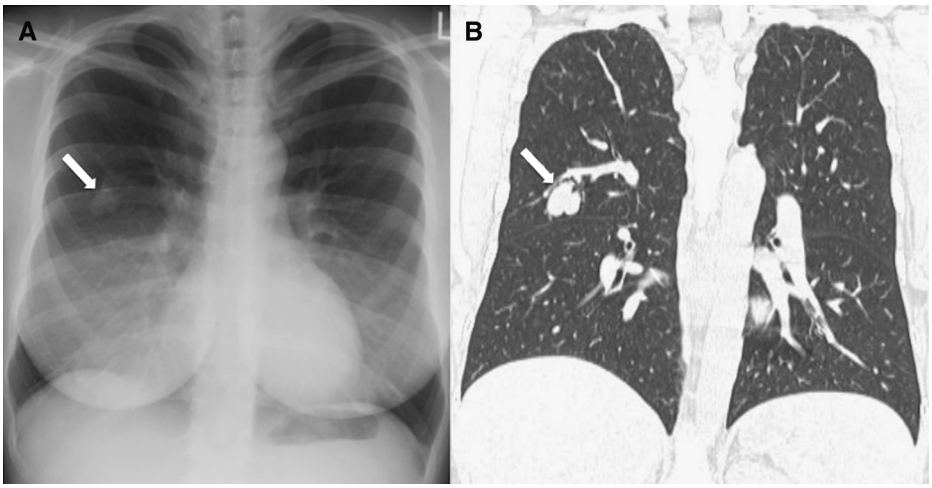

Figure 1. A 30-year-old woman with hereditary hemorrhagic telangiectasia. (A) Frontal chest radiograph and (B) coronal computed tomography image, demonstrating a right upper lobe pulmonary arteriovenous malformations (arrows).