Key Role of Otolaryngology in a Hereditary Hemorrhagic Telangiectasia Center of Excellence

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ABSTRACT

OBJECTIVES: Review the initial experiences of an HHT Center of Excellence and understand which HHT patients require otolaryngologic evaluation and interventions.

METHODS: Retrospective review of all patients referred to our HHT Center of Excellence from its inception in May 2010 through June 2012.

RESULTS: In our cohort of 144 patients, 98 have HHT based on Curacao clinical criteria, and 16 have possible or unlikely HHT based on the Curacao clinical criteria. 30 patients met 2 of the 4 criteria, or are undergoing further work-up, and possibly have the disorder. 30 patients met less than one of the four criteria and do not have HHT.

CONCLUSIONS: An HHT Center of Excellence is important in providing comprehensive care for patients with this rare disease with significant clinical sequelae. Otolaryngologists are critical members of this multidisciplinary team. Additional, 47 patients have been treated for symptomatic AVMs in other locations or are being followed closely with radiologic imaging to rule out enlargement of previously treated AVMs. Several of these patients have had interventions for multiple AVMs at different locations in the body for a total of 53 interventions. These interventions have included embolization, neurosurgical clipping, or radiation for AVMs in the lung, brain, abdomen, and pelvis.

DISCUSSION

HHT Centers of Excellence allow early diagnosis, screening and treatment. Severe complications of HHT, such as stroke, lung and brain hemorrhages can be prevented. In addition, early intervention and close monitoring can improve HHT patients’ quality of life and minimize the morbidity associated with severe epistaxis and GI bleeds.

Otolaryngology plays a key role in managing these patients, especially considering that most common symptom of HHT is epistaxis which occurs in >90% of patients1. In just 16 months, 20% of the patients with HHT at our center have been referred to otolaryngology, including 11 patients who have needed interventions in the OR. In addition to these 20 patients, 10 additional patients were seen by outside otolaryngologists before the opening of UCLA HHT Center.

METHODS

We performed a retrospective review of all patients from the Southwestern United States referred to our HHT Center of Excellence from its inception in May 2010 through June 2012. Clinical presentation, radiographic imaging, genetic testing, otolaryngologic treatments, and other operative interventions were analyzed.

RESULTS

- Of the 98 patients with confirmed HHT, 20 (20%) have been referred to otolaryngology for further evaluation for recurrent epistaxis or persistent oral bleeding. Of these 20 patients, 11 (55%) have had KTP laser treatments for symptomatic oral, opharyngeal, and nasal mucosal telangiectasias. Two of these nine patients have had multiple treatments over the last two years. The 11th patient did not have KTP treatments, but had excision of an enlarged telangiectasia from the hard palate for repeated acute hemorrhages requiring transfusion; she has had significant improvement since surgery.

- Additionally, 47 patients have been treated for symptomatic AVMs in other locations or are being followed closely with radiologic imaging to rule out enlargement of previously treated AVMs. Several of these patients have had interventions for multiple AVMs at different locations in the body for a total of 53 interventions. These interventions have included embolization, neurosurgical clipping, or radiation for AVMs in the lung, brain, abdomen, and pelvis.

REFERENCES