INTRODUCTION

Hereditary Hemorrhagic telangiectasia (HHT), or Rendu-Osler-Weber disease, is a vascular anomaly characterized by multiple dilations of skin and mucosa capillaries and venules. It is an autosomal dominant disease, equally distributed between both genders and its incidence is of 1-2/100,000 inhabitants.1

Bleeding may occur in numerous places; however, epistaxis is the most common, present in 90% of the cases2. The basic lesion is found on the vessels’ walls, with defects in the elastic and muscular layers that make them more prone to bleeding3,4.

Many treatment modalities have been used to control epistaxis; none of them have rendered entirely satisfactory results. Options are nasal packing, hormone-therapy, vascular embolizations, fascioplasty and septodermoplasty. Young’s surgery is based on nasal occlusion, thus preventing the friction between air and the telangiectasia, precluding epistaxis episodes.5

CASE REPORT

A.B.L., 56 years old, history of recurrent epistaxis, mainly affecting the left side, since he was 15 years of age. He had been submitted to many blood transfusions, as well as cauterizations and fascioplasty, and had excessively pale skin (+++/4+). His hematocrit count was 24%. The patient had no respiratory disorder and the cosmetic aspect was very satisfactory.

Repetition epistaxis is the most common HHT symptom, present in 90% of the cases. This was the only symptom our patient had. The condition started on his second decade of life, becoming increasingly more intense, in agreement with data presented in the literature.

The effectiveness of Young’s technique in controlling epistaxis in patients with HHT is due to terminating with the air turbulence in a frail mucosa bearing diseased vessels6. This procedure is not much reported in the literature, and it is extremely efficient in controlling recurrent epistaxis.

Our patient did not have any epistaxis episode after two years and four months of surgery, and such time is in agreement with data presented in the literature.

DISCUSSION

Efficient occlusion was carried out on the skin-mucous joint (Figure 1), preserving the vibrissae on the side of the nostril, and suturing up in two planes, a more internal mucous layer and another external, skin layer. After two years and three months of follow up there was no other epistaxis episode and he also had hematologic normalization. There was no respiratory disorder and the cosmetic aspect was very satisfactory.

REFERENCES