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CEREBRAL ABSCESS AND HEREDITARY TELANGIECTASIA — REPORT OF A CASE

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SUMMARY

One case of cerebral abscess complicating hereditary hemorrhagic telangiectasia (HHT) is described. A confirmed pulmonary arteriovenous fistula (PAVF) was connected with its primary etiology. The high incidence of cerebral abscess complicating PAVF in HHT found by some authors may suggest other possible mechanisms on the origin of this entity.

RESUMO

Abcesso cerebral e Telangiectasia Hereditária — A propósito de um caso.

Descreve-se um caso de abcesso cerebral num doente com Telangiectasia Hemorrágica Hereditária. Na sua origem estaria uma fístula arteriovenosa pulmonar. Uma incidência muito elevada de abcessos cerebrais na Telangiectasia Hemorrágica Hereditária, referida por alguns autores, parece sugerir a existência de outros mecanismos etiopatogénicos além dos dependentes da fístula arteriovenosa pulmonar.

INTRODUCTION

The hereditary hemorrhagic telangiectasia (HHT) is a familial disorder characterized by multiple mucocutaneous and/or visceral telangiectases, recurrent bleeding and autosomic dominant character. Neurologic manifestations have been uncommonly described in HHT. Most of these situations result from pulmonary arteriovenous fistulae (PAVF),[1-5] with cerebral abscess as the most serious complication.[1, 3-8] Our purpose is to report a typical case and to discuss the high frequency of cerebral abscess complicating PAVF in HHT.

CASE REPORT

A 33-year-old man was admitted to the Neurology and Neurosurgery Department on July 1980 because of headache, vomiting, convulsions and right hemiparesis. One year before admission epistaxis had occurred. One week before he complained of headache, followed few hours later by right-sided clonic movements of the face. Paresis of the right upper limb and face was then noted. Identical partial epileptic crisis repeated on the next day. Subsequently there was progressive worsening of right hemiparesis accompanied by headache, vomiting and mental confusion until admission.

Examination revealed a cooperative but drowsy man, afebrile, with a pulse of 66 and a blood pressure of 110/70 mm Hg. Multiple lip and tongue telangiectases, perioral and peripheral cyanosis and clubbing of the fingers and toes were observed. Ophthalmoscopy was normal. Right-hemiparesis was present. On the right side there was increased tendon reflexes, absent abdominal reflexes and a Babinsky sign. There was no sensory impairment. A continuous murmur with systolic intensification was heard over the right hemithorax at the mamilary level. Heart auscultation was normal. Hemoglobin level was 20.5 g/dl; hematocrit value 0.59 l/l; white blood cell count 12×10^6/l; arterial blood gases were pO_2 67 mm Hg, pCO_2 35 mm Hg, pH 7.43 and oxygen saturation 92.4%. Routine blood chemistries and urinalysis were within normal range. Chest roentgenogram revealed a dense shadow in the medium segment of the right lung with vascular marketings to the hilum thought to be consistent with a pulmonary arteriovenous fistula[9] (Fig. 1A and 1B). Skull roentgenogram was normal, and electroencephalogram showed irregular and slow waves on the left temporal and parietal areas.

Computed axial tomography revealed a large left temporal mass suggestive of brain abscess (Fig. 2A). Parenteral therapy with penicillin and cloramphenicol was iniciated. The abscess was drained through a left temporal burr hole, under local anesthesia, and the purulent material obtained grew Streptococcus on anaerobic conditions.

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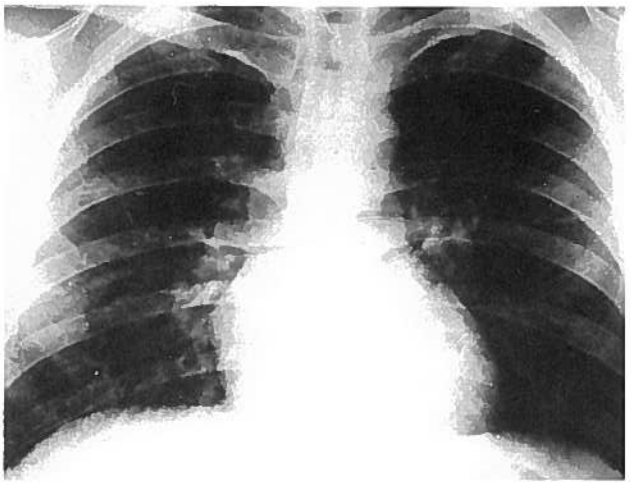

Fig. 1A: Pulmonary arteriovenous fistula in right lung seen on postero-anterior chest roentgenogram

Fig. 1B: Pulmonary arteriovenous fistula in right lung seen on postero-anterior lung tomogram

The patient had an uneventful recovery and was discharged on September 1980 with no neurological sequelae. Follow-up computed axial tomography two months later showed resolution of the abscess (Fig. 2B).

On January 1981 a segmental resection of the pulmonary arteriovenous fistula was successfully performed at Thoracic Surgery Department of this hospital (Prof. M. Rodrigues Gomes).

As the disease was not known before, a family study as complete as possible was done and available members were examinated (Fig. 3).

DISCUSSION

HHT is a common familial disorder with an estimated incidence of 1 or 2 per 100 000.[1] However, the diagnosed cases are below the true incidence of the disease because, as in the case above described, most often the diagnosis is only considered when a serious complication shows off. Among other reasons, the late onset and the great variability of the clinical manifestations of this entity may play a relevant role to explain that.[4, 8] HHT is a chronic and progressive disease whose manifestations usually begin in early adult life[4, 8, 10, 11] showing as a rule no symptoms during childhood, as is also illustrated in the family study of our patient (Fig. 3). On the other hand, the telangiectases usually predominate in the skin and mucous membranes but may have only a visceral localization.[11] In either instance, but mainly in the latter, hemorrhagic features may never occur and frequently the disease will only be recognized when secondary complications become obvious. Stressing this point is the somewhat surprising negative familial history associated with the demonstration of overlooked characteristic lesions in some documented cases like in our purpose and his relatives.

Neurological manifestations in HHT have not been frequently reported. Their overall frequency is estimated to be 8 to 12%, according to Román et al,[5] 61.4% of them representing secondary complications of pulmonary arteriovenous fistulae (PAVF), which are by far the most frequently seen; the remaining neurological presentations of the disease, are those resulting from cerebral or spinal cord vascular malformations and finally portal systemic encephalopathy.[2-5]

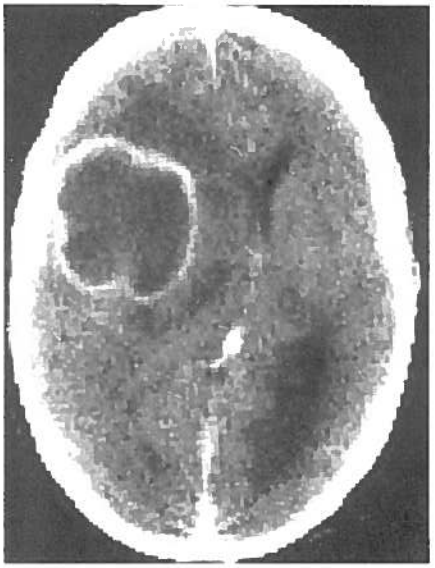

Fig. 2A: Computerized axial tomography at admission, showing cerebral abscess

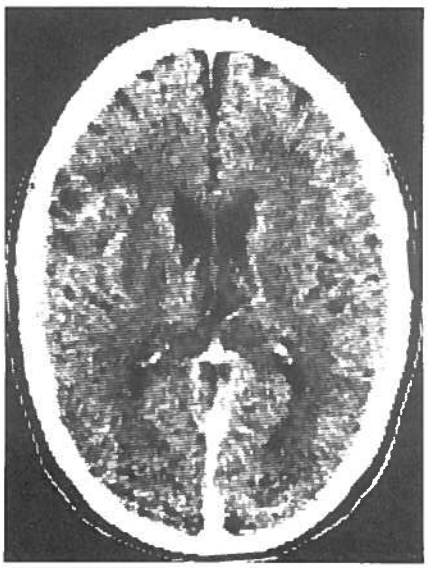

Fig. 2B: Follow-up scan two months after medical and surgical treatment

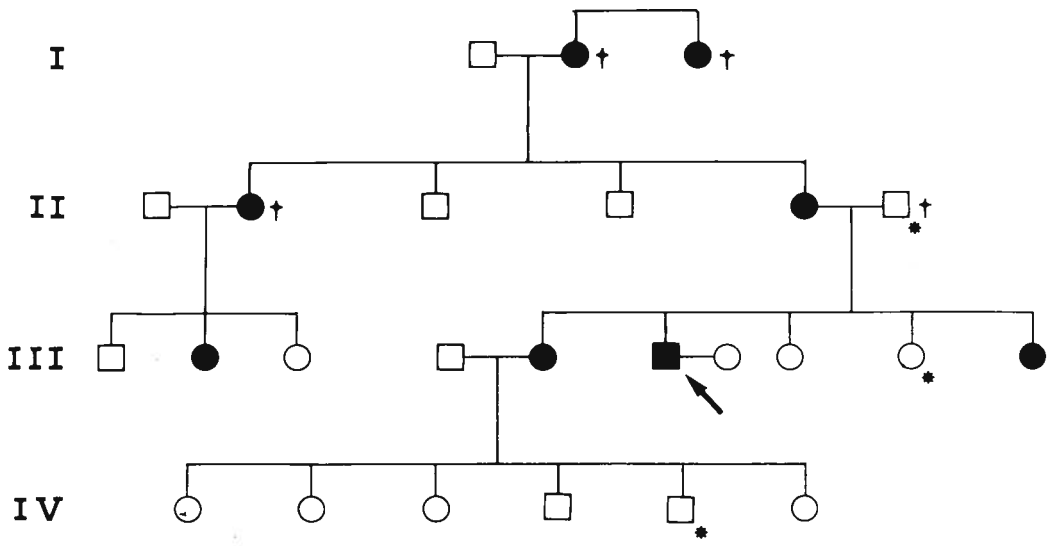

Fig. 3: Family trees of the propositus (/). Relatives with HHT are in black. (*) Associated epilepsy

Cerebral abscess is the most important of the neurological disorders arising from PAVF in HHT [1, 3-5, 7] and pathophysiologycally is similar to those complicating right-to-left shunts.[1, 6] Román et al.[5] found a frequency of 21.2% for cerebral abscess complicating PAVF in HHT, based on a review of the reported cases. This data are conflicting with the smaller number of 5% found by others for cerebral abscess complicating the whole PAVF.[1, 4, 6, 7, 11-13] The different criteria used for their assessment may in great part account for such a disagreement. At this regard it seems undoubtedly more reliable a calculated incidence on the basis of larger series of PAVF than based on few sporadic reported cases of cerebral abscesses complicating PAVF in HHT. Although considering exagerated the discrepancy found by Román et al.,[5] a higher incidence of cerebral abscess in HHT could really exist as a result of associated unrecognized disorders.

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