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Original

Management of Hemangioblastomas: our experience

Tratamento de hemangioblastomas: nossa experiência

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ABSTRACT

Objective: *The aim of this paper is to present our experience in the management of hemangioblastomas.* ***Material and Method:*** *ten patients with sporadic hemangioblastomas (6 male and 4 female; range 4-75 years) were analyzed. Central ventriculography, cerebral arteriogram, cranial CT scan and head MRI (MR spectroscopy and diffusion MRI) were performed. Location was at cerebellum in 8 patients, left occipital lobe in 1 and brain stem in 1. Nine patients were surgically treated and 1 patient underwent stereotactic radiosurgery due to tumour location in the brainstem and surgery was clinically contraindicated.* ***Results:*** *Only one recurrence occurred in a 4-year-old patient who had undergone subtotal resection and had to be operated 10 years later due to tumor growth. Two patients died; one subject during the immediate post-operative period due to massive gastrointestinal bleeding and cardiac arrest and the other due to myocardial infarction 25 years after surgery. The 8 surviving patients currently present with normal neurological status and have perfectly normal lives, including the subject who underwent stereotactic radiosurgery of brainstem. The survival rate of these patients is 1-16 years.*

Key words: *Sporadic hemangioblastoma; Spectroscopy; Diffusion; Surgery; Stereotactic radiosurgery.*

RESUMO

Objetivo: *Apresentar nossa experiência no tratamento de hemangioblastomas.* ***Material e Método:*** *Foram analisados dez pacientes (6 homens e 4 mulheres, na faixa entre 4-75 anos) com hemangioblastoma esporádico localizado no cerebelo (8 pacientes), lobo occipital esquerdo (1 paciente) e tronco encefálico (1 paciente). Nove pacientes receberam tratamento cirúrgico e em um paciente foi realizada a radiocirurgia estereotáxica, devido à sua localização no tronco encefálico, sendo clinicamente contra-indicando a cirurgia.* ***Resultados:*** *Um paciente de 4 anos apresentou recorrência pós-ressecção subtotal, sendo operado 10 anos depois devido a crescimento tumoral. Dois pacientes evoluíram para óbito: um durante o período pós-operatório imediato por intensa hemorragia gastrointestinal e infarto do miocárdio 25 anos após a cirurgia. No acompanhamento final, os demais 8 pacientes apresentavam condições neurológicas normais e com "vida normal", inclusive aquele sujeito à radiocirurgia estereotáxica de tronco encefálico. A taxa de sobrevida destes pacientes foi de 1-16 anos.*

Palavras-chave: *Hemangioblastoma esporádica; Espectroscopia; Difusão; Cirurgia estereotáxica.*

Introduction

Hemangioblastomas are slow-growing and very much vascularized tumors located in the brain, cerebellum, brainstem and spinal cord mostly in adult patients[1]. They represent 2% of all intracranial tumors. Nearly 10% are located in the posterior fossa and represent between 2% and 3% of intramedullary tumors[3,10].

About 25% of this type of tumors is linked to Von Hippel-Linau syndrome (VHL), that is an autosomal dominant genetic condition whose inclusion criteria are: the presence of one or more hemangioblastomas of the central nervous system (CNS), the presence of hemangioblastomas of the CNS linked to tumor diseases of VHL, or family history of this disease[2].

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The tumor diseases of VHL are: retinal hemangioblastoma, renal cell carcinoma, feocromocitoma, pancreatic tumor and visceral cysts. This is why patients under 50 years of age must be studied not only with MRI of the whole nervous system but also with contrast-enhanced CT scans of the orbit and abdomen[1,2,3,10].

Material and Method

Through January 1972 to December 2013, a total of 1,411 Central Nervous System (CNS) tumors were operated on our Department, from which 13 (0.92%) were hemangioblastomas. Only 10 cases were selected because they were the only subjects who achieved a good follow-up.

From these 10 patients with hemangioblastomas, 6 were male and 4 female. The age range was 4-75 years, with a mean age was 51 years and a median age of 58 years.

Early symptoms are described in Table 1, and the clinical signs at the time of admission are shown in Table 2. Until diagnosis development of the symptoms had a range of 1-24 months, a mean of 4.7 months and a median of 2 months. These tumors were located in: cerebellar vermis: 4, cerebellar hemisphere: 4, left occipital lobe: 1, and brainstem: 1.

The diagnostic procedures used in this series are described in Table 3.

The MR Spectroscopy (MRS) and Diffusion MRI (dMRI) were of great value in the identification of this type of tumor (Figure 1). In addition, all the patients under 50 years of age were studied with CT scans of the orbit and abdomen to rule out Von Hippel-Lindau syndrome (VHL).

Nine patients were surgically treated and the other one underwent stereotactic radiosurgery because of the age (75 years-old), the location of the tumor (brain stem) and clinical contraindications to surgery. From the 9 operated patients, total resection was achieved in 8 (Figure 2) and subtotal resection in 1, for which this patient was reoperated 10 years later due to tumor growth. The stereotactic surgery was performed with Novalis Tx, which includes image-guided system and robotic alignment that allows high accuracy in a short period of time.

Table 1. Early Symptoms

Symptom	n
Abnormal gait	8
Cephalea	8
Vomit	5
Humming	1
Vision problems	1
Brachial-crural paresis	1
Swallowing Difficulty	1
Breathing problems	1

Table 2. Symptoms at admissions

Symptom	n
Ataxia	7
Papillary Edema	7
Unilateral Dysmetria	2
Hemianopsia	1
Dysarthria	1
Brachial-crural paresis	1
Spatial and Temporal Disorientation	1

Table 3. Diagnostic Procedures

Procedure	n
Cerebral Arteriogram	1
Ventriculography	1
CT	2
MRI	8

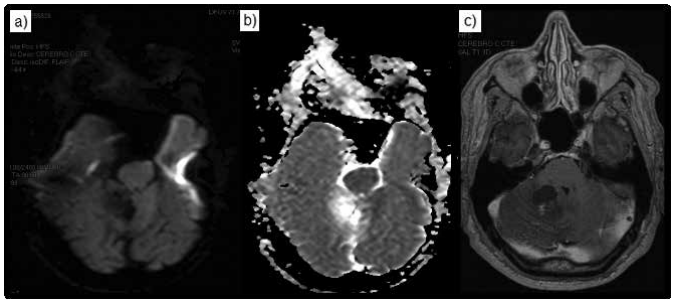

Figure 1: MRI of cerebellar hemangioblastoma: a) Diffusion; b) ADC; c) T1 with gadolinium.

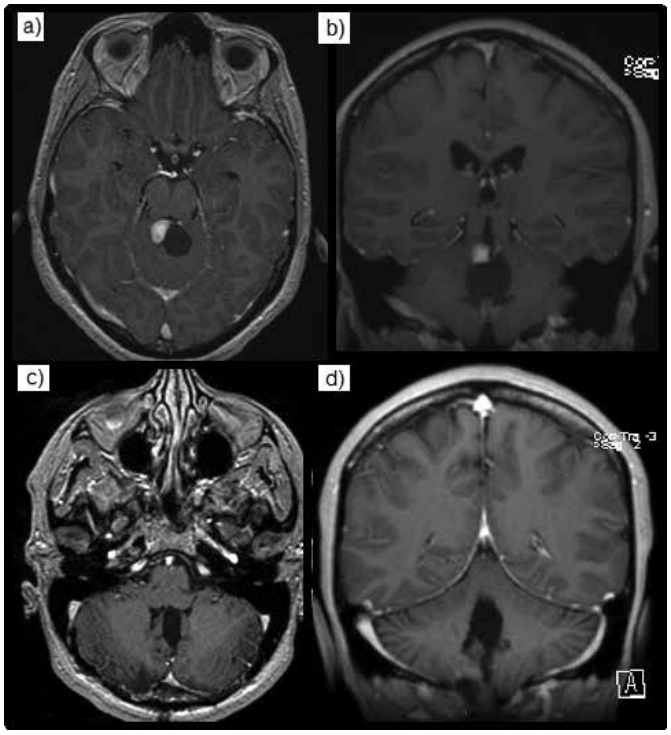

Figure 2: MRI of cerebellar hemangioblastoma: a-b: pre-op; c- d: post-op.

Results

Only one recurrence occurred in a 4-year-old girl with subtotal resection, who was reoperated 10 years later after observing a progressive increase of the tumor size in follow-up MRIs. Currently, and 16 years after the second surgery, the patient shows with a normal neurological status and has a perfectly normal life.

Two patients died; one subject during the immediate post-operative period due to massive gastrointestinal bleeding followed by cardiac arrest. The other subject died due to myocardial infarction 25 years after total resection.

It is important to emphasize that, before treatment, the patient who underwent stereotactic radiosurgery of the brainstem presented with severe neurological deficit, ataxic gait, brachial-crural hemiparesis, swallowing difficulty and breathing problems. After treatment with Novalis Tx radiosurgery a total neurological recovery was achieved.

The surviving patients[8] presented a normal neurological status, and have perfectly normal lives with a survival rate between 10 months and 16 years, a mean of 39.5 months and a median of 24 months.

Discussion

No patient had Von Hippel-Lindau syndrome (VHL) in our series; all were sporadic cases[2,3].

The cerebellum was the most frequent location, 8 out of 10 patients[1,3,4,10].

MRI plays a very important role in the diagnostic procedures, especially MR spectroscopy (MRS), that detects the proteomic profile of the cyst content as a consequence of bleeding in the cyst fluid. Diffusion MRI (diffusion-weighted imaging DWI and Apparent Diffusion Coefficient ADC) also plays an important role. These tumors have a similar behavior to cerebrospinal fluid (CSF), with a facilitated diffusion and a high ADC, a typical characteristic[7].

Because of tumor size and endocranial hypertension, surgery was the treatment of choice in 8 patients. One of these patients was reoperated 10 years after achieving subtotal resection in the first surgery.

In spite of tumor size, one case had no surgery indication due to clinical conditions. This patient underwent stereotactic radiosurgery at a dose of 54 Gy and the procedure was performed with Novalis Tx, which includes image-guided system and robotic alignment[5,6,8,9].

Conclusion

Hemangioblastomas of CNS despite their location, must be totally resected in order to avoid recurrences and to guarantee a very good survival rate.

In those cases where surgery is not indicated, stereotactic radiosurgery is an excellent choice, since it is a non-invasive procedure with very good results.

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