Journal of Clinical Neuroscience 21 (2014) 1205–1208

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Clinical Study

# Hemangioblastomas in the elderly: Epidemiology and clinical characteristics

Charles D. Kassardjian [a], R. Loch Macdonald [c,d,e,f], David G. Munoz [b,e,*]

[a] Division of Neurology, University of Toronto, Toronto, ON, Canada
[b] Division of Pathology, St. Michael's Hospital, 30 Bond Street, Toronto, ON M5B 1W8, Canada
[c] Division of Neurosurgery, St. Michael's Hospital, Toronto, ON, Canada
[d] Labatt Family Centre of Excellence in Brain Injury and Trauma Research of the Li Ka Shing Knowledge Institute of St. Michael's Hospital, Toronto, ON, Canada
[e] Keenan Research Centre of the Li Ka Shing Knowledge Institute of St. Michael's Hospital, Toronto, ON, Canada
[f] Department of Surgery, University of Toronto, Toronto, ON, Canada

ARTICLE INFO

*Article history:*
Received 30 July 2013
Accepted 13 October 2013

*Keywords:*
Cerebellar tumor
Elderly
Hemangioblastoma
von-Hippel Lindau

ABSTRACT

Intracranial hemangioblastomas are benign vascular tumors. The peak age of incidence is between 20 to 50 years. Hemangioblastomas rarely occur in patients over the age of 65. To our knowledge there is no review of the prevalence and clinical features in an elderly population. We reviewed our 12 year experience with intracranial hemangioblastomas, and characterized the clinical features of hemangioblastomas in patients over the age of 65. We present a 72-year-old man with a cerebellar mass initially thought to be a metastasis as an illustrative case. We reviewed our pathology database and identified all patients with a histopathologically confirmed diagnosis of hemangioblastoma over the last 12 years in a large tertiary adult hospital; all patients were over the age of 18. Of all cases of hemangioblastoma in the last 12 years, six of 77 (7.7%) occurred in patients over the age of 65. All were cerebellar in location, and none were associated with von-Hippel Lindau disease. Hemangioblastomas are uncommon, but not rare, in patients over the age of 65, and should be included in the differential diagnosis of patients presenting with gait ataxia and a cerebellar lesion in this age group.



## 1. Introduction

Hemangioblastomas are benign World Health Organization (WHO) grade I vascular tumors of the central nervous system (CNS) that represent about 7% of posterior fossa neoplasms [1,2]. Although they can occur in any part of the nervous system, they most commonly arise in the posterior fossa (94%), and less commonly in the spinal cord or supratentorially [1,3]. Hemangioblastomas can be either sporadic and isolated, or associated with tumors in multiple organs as part of von Hippel-Lindau disease (VHL, 20–30%) [3–5]. The most common tumors associated with VHL are retinal angiomatosis, pheochromocytoma, and pancreatic and renal cysts [1,3,5,6].

Hemangioblastomas tend to be tumors of adults, with a peak age incidence between 20 and 49 years [3,7]. Although cases are reported in pediatric and elderly patients, these are relatively less common, especially in patients over the age of 65 [3]. The diagnosis is challenging in the elderly, as alternate tumors are usually considered first when an elderly patient is found to have an enhancing posterior fossa mass on imaging. Malignant neoplasms, particularly metastases, are the most common tumor.

We are not aware of any articles systematically reviewing the incidence and features of hemangioblastoma in patients over 65 years. We were prompted to review our 12 year experience with hemangioblastomas in elderly patients after providing a histopathologic diagnosis in a 72-year-old man presenting with a contrast-enhancing posterior fossa mass who was being investigated for brain metastasis.

## 2. Illustrative patient

The patient presented with a 1 week history of occipital headache, associated with nausea and vomiting. His physical examination was notable for a wide-based ataxic gait, without appendicular ataxia. There was no nystagmus, and the remainder of the neurological examination was unremarkable. He had no previous history of neurological complaints. He had a past medical history of diabetes and hypertension, and his medications included metformin, sitagliptin, candesartan, and acetylsalicylic acid. He

* Corresponding author. Tel.: +1 416 864 5858; fax: +1 416 864 5648.
*E-mail address:* munozd@smh.ca (D.G. Munoz).

http://dx.doi.org/10.1016/j.jocn.2013.10.023
0967-5868/

was a previous smoker, but had quit over 20 years prior to this presentation. There was no known family history of neoplasm or neurological illness.

MRI of the brain with and without gadolinium contrast demonstrated a solitary enhancing mass in the right posterior fossa, measuring 2.4 × 2.2 × 2.7 cm (Fig. 1). The mass was hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences, with a small non-enhancing cystic component. It was centered superficially within the cerebellum and extended to the dura. There was no enhancement of the dura, but the possibility of an extra-axial mass could not be excluded. There was some effacement of the fourth ventricle, but no hydrocephalus. A metastatic lesion was felt to be the most likely diagnosis. CT scan of the thorax, abdomen and pelvis with contrast revealed no evidence of primary or metastatic neoplasm. An incidental right adrenal adenoma was noted.

The patient underwent a suboccipital craniotomy for resection of the mass, with no complications. He recovered fully and returned to all of his premorbid activities 3 months later, and had a modified Rankin Scale score of 0. Histopathological examination of the tissue revealed a tumor composed of a dense network of small blood vessels lined by a single layer of endothelial cells, among which were numerous stromal cells, characterized by large dark nuclei and abundant cytoplasm containing lipid vacuoles (Fig. 2). There was no mitotic activity, the Ki-67 proliferation index was 1%, and there were no areas of necrosis. The stromal cells showed immunoreactivity for cytoplasmic inhibin, membranous D2-40, and cytoplasmic and nuclear neuron-specific enolase. There was no immunoreactivity for cytokeratin. The diagnosis was hemangioblastoma (WHO grade I).

## 3. Methods

We identified cases diagnosed histopathologically as hemangioblastoma from 2000 to 2012 at our institution in patients over the age of 65 (our definition of "elderly"). Patients under the age of 18 are not seen at our hospital. We extracted available clinical information regarding presentation, tumor location, radiological features, management, survival, and association with VHL.

## 4. Results

A total of 77 patients with hemangioblastoma were found in the last 12 years. Of these, seven patients were over the age of 65 at the time of pathological diagnosis. One patient represented a recurrence of a tumor previously identified 15 years earlier (when the patient was 54 years old), so this case was excluded. Thus, there were six cases of hemangioblastoma first diagnosed in patients over the age of 65, out of a total of 77 hemangioblastomas seen over the last 12 years (8%). Four patients were male. A summary of the clinical data from these six patients can be found in Table 1. The clinical presentation was ataxia or headache in all patients. The lesion location in all six elderly patients was cerebellar, and all patients had a single CNS hemangioblastoma. No patients had a known family history of VHL disease, and all were considered sporadic hemangioblastomas. One patient was noted to have bilateral renal cysts, but this was insufficient to diagnose VHL disease [1,5]. All six patients survived postoperatively and were discharged home. One patient was lost to follow-up. Three patients have shown no evidence of tumor recurrence (follow-up between 2 and 10 years). One patient had a recurrence of the cerebellar hemangioblastoma 3 years after initial resection, and another had recurrences at 7 and 9 years after initial resection.

## 5. Discussion

Hemangioblastomas are uncommon tumors of young to middle-aged adults, and are often a consideration when symptoms or imaging demonstrate a posterior fossa lesion in this age group. Although the most common location for hemangioblastoma (both sporadic and VHL-associated) is in the cerebellum, many other locations have been reported, including the medulla, pituitary stalk, spinal cord, in addition to other rarer locations [3,8–11]. Several case reports of unusual locations of hemangioblastoma (for example, cranial nerves, lateral ventricle) include patients over the age of 65, but these are rare cases of hemangioblastoma, rather than the more common cerebellar location.

The prevalence of hemangioblastomas in elderly patients, particularly those over the age of 65, is not well characterized. In one review of 47 patients with CNS hemangioblastoma over 10 years, the reported age range was 12 to 71 years, with a mean age of 42.4 years [3]. Fewer than 5% of patients were over 65 years of age, and there were no patients with hemangioblastoma associated with VHL disease over the age of 50 [3]. In another review of surgical outcomes in 52 patients with posterior fossa hemangioblastoma, the reported age range was between 20 and 73 years (mean 43.4 years) [2]. A more recent retrospective study of 49 patients with CNS hemangioblastoma reported a mean age of

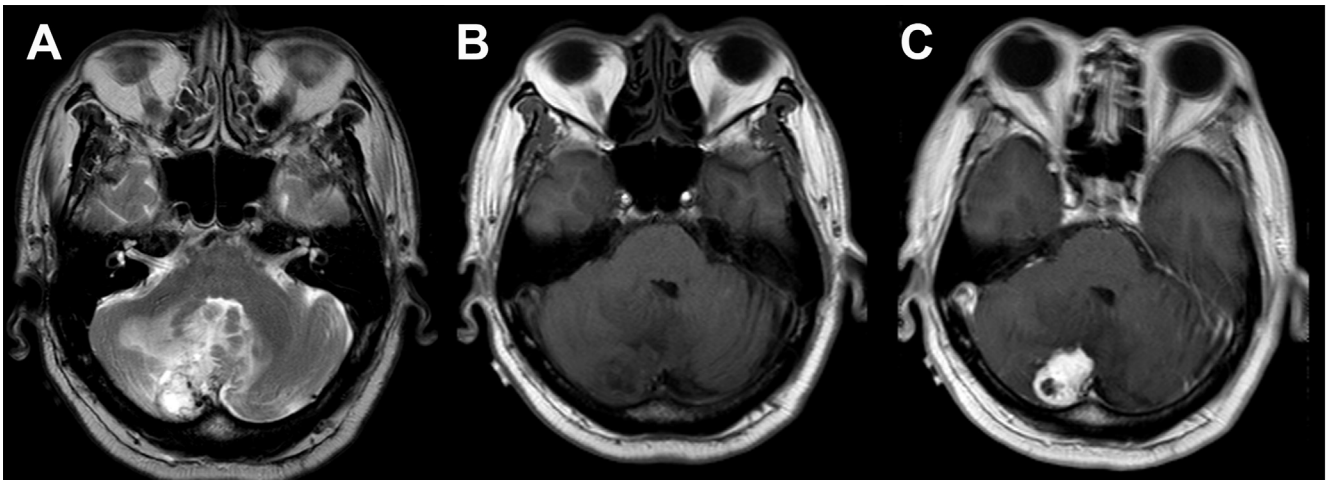


**Fig. 1.** Axial T2-weighted (A) and T1-weighted MRI without (B) and with (C) gadolinium contrast. There is a mass in the superficial medial part of the right cerebellar hemisphere that is hypointense on T2- and T1-weighted images and that enhances with gadolinium. It is well defined, with flow-voids seen adjacent to it on T2-weighted imaging, suggesting that it is highly vascular. There is surrounding hypointensity on T2-weighted imaging and some mass effect on the fourth ventricle.