HEMORRHAGIC ACOUSTIC SCHWANNOMA: RADIOLOGICAL AND HISTOPATHOLOGICAL FINDINGS

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SUMMARY

A 49-year-old man on anticoagulation treatment with phenprocoumon presented with acute right sided 7th and 8th cranial nerve palsy, acute hearing loss, headache, vertigo, and vomiting. CT and MRI revealed a cerebellopontine angle tumor 15mm in diameter and acute intratumoral hematoma. A cellular schwannoma composed predominantly of Antoni A tissue with dilated thin-walled vessels, surrounded by old hemorrhage with hemosiderin-laden macrophages was found histologically.

Key words: schwannoma, cerebellopontine angle, neoplasms, magnetic resonance (MR), computed-tomography (CT).

INTRODUCTION

Massive hemorrhage in acoustic schwannomas is rare, especially in tumors smaller than 25mm. We report a case of massive hemorrhage into an acoustic schwannoma, 15mm in diameter, in a patient on oral anticoagulation treatment presenting with acute 7th and 8th cranial nerve palsy.

CASE REPORT

A 49-year-old man presented with acute right side palsy of the 7th and 8th cranial nerves, acute hearing loss, headache, vertigo, and vomiting. Coagulation time was increased (international normalized ratio > 7) secondary to oral anticoagulation treatment with phenprocoumon. Audiographic examination revealed perceptive deafness in the right ear, pathological vestibular response to caloric stimulation, and pathological brainstem auditory-evoked responses in the right ear. Cranial computed tomography (CT) showed a hyperdense mass in the right cerebellopontine angle extending about 15mm (figure 1a). At the dorsolateral aspect, contrast enhancement was detected after intravenous administration of contrast material (figure 1b). Findings from CT angiography and catheter angiography performed to exclude an aneurysm of the verteobasilar circulation were normal. The MRI signal in the center of the mass was hyperintense on T2-weighted and isointense on T1-weighted images relative to cerebellar white matter, suggesting acute hematoma at the oxyhemoglobin stage (figures 2a and 2b). On T2-weighted images the center of the mass was surrounded by a hypointense rim which was thought to represent old hemorrhage at the hemosiderin stage. After administration of Gd-DTPA, contrast enhancement was evident within the internal auditory canal as well as along the periphery of the CP angle compartment of the tumor. The tumor was neurosurgically resected. Histological examination revealed a cellular schwannoma composed predominantly of Antoni A tissue. Loosely textured Antoni B tissue was hardly detectable. A mild degree of nuclear atypia was noted and only few mitotic figures were present. The cellular areas were composed of closely packed spindle-shaped neoplastic Schwann cells. In some areas, numerous dilated thin-walled vessels, often surrounded by old hemorrhage with hemosiderin-laden macrophages, were conspicuous. Immunohistochemically, the majority of tumor cells were strongly reactive for S-100 protein. After neurosurgical resection of the tumor, the 7th cranial nerve dysfunction improved slightly. However, perceptive deafness in the right ear remained and hearing loss persisted.

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DISCUSSION

Acoustic schwannoma is the most common cerebellopontine angle mass. Radiologically, an acoustic schwannoma is typically seen as an extraaxial, well-defined, noncalcified mass with mild to marked increased signal on T2-weighted MR images [11]. Cystic degeneration is often detected. The solid components of acoustic schwannomas typically show intense enhancement after contrast administration. Clinical symptoms include slowly progressive sensorineural hearing loss over months and tinnitus [4]. Histologically, schwannomas are composed of spindle-shaped neoplastic Schwann cells with alternating areas of compact, elongated cells showing occasional nuclear palisading (Antoni type A) and less cellular, loosely textured, often lipidized tumor areas (Antoni type B) [1]. Intracranial tumors causing hemorrhage are mainly glioblastomas, metastatic chorioncarcinomas, melanomas, pituitary adenomas, meningiomas, oligodendrogliomas, and choroid plexus papillomas [5]. Massive, i.e., macroscopically or radiologically detectable, intratumoral hemorrhage in acoustic schwannomas is very rare: There are only about 40 reports of this phenomenon in the medical literature [9]. Occult, i.e., only microscopically detectable,

Fig. 1. – Axial CT at the level of the internal auditory canal before (A) and after (B) contrast administration shows a high-density, right cerebellopontine angle mass 15mm in diameter with contrast enhancement at the dorsolateral aspect.

Fig. 2. – (a) Axial T2-weighted MR images (3000/80/1/3mm [TR/TE/NEX/section thickness]) confirmed the right cerebellopontine angle mass, with high signal in the center and low signal at the periphery of the tumor. On axial T1-weighted MR images (672/20/1/3mm) before contrast administration (b) the central component of the mass is isointense whereas the dorsolateral aspect of the periphery is slightly hypointense relative to cerebellar white matter. The center and the periphery of the mass seem to be separated by a hypointense rim. On axial (672/20/1/3mm) and coronal (672/20/1/3mm) T1-weighted images strong enhancement of the IAC component and periphery of the CP angle component is evident after contrast administration (c, d). There is moderate mass effect upon the middle cerebellar peduncle.

Fig. 2. – (a) Coupe axiale, séquence pondérée en T2. Lésion de l’angle pontocérébelleux droit, hypersignal central, hyposignal périphérique. Coupes axiales. Séquence pondérée en T1. Avant contraste, (b) la partie centrale est en isosignal alors que la partie dorsolatérale est légèrement en hyposignal par rapport à la substance blanche. Le centre et la périphérie de la masse semble séparée par un anneau hypointense. Il existe un rehaussement net des portions intraméatale et extraméatale périphérique (c, d) et un effet de masse modéré sur le pédoncule cérébelleux moyen.
hemorrhage is reported to occur in five of 57 cases (8.8%), but only in tumors larger than 25mm in diameter [6]. Risk factors for bleeding in acoustic schwannomas are anticoagulation therapy as in our patient [3], trauma [7], pregnancy [9], drug abuse [13], high vascularization of tumors [10], and tumors more than 25mm in diameter [2]. According to Misra [10], mixed histological Antoni type seems to be an additional risk factor for bleeding. In our patient, radiological findings revealed a cerebellopontine angle tumor with acute intratumoral hematoma in the oxyhemoglobin stage. Before surgery, arterial aneurysm, the most important differential diagnosis apart from petroclival meningioma, was excluded angiographically. Histologically, a cellular schwannoma composed predominantly of Antoni A tissue with dilated thin-walled vessels, surrounded by old hemorrhage with hemosiderin-laden macrophages was found. We believe that the sudden onset of vertigo, vomiting, and headache as well as the 7th cranial nerve palsy in our patient were the result of the rapid increase in tumor volume due to massive intratumoral bleeding.

In most of the case reports of hemorrhage into acoustic schwannomas, hypervascularity and dilated vessels were the main pathological changes [5, 8]. According to Russel and Rubinstein [12], these pathological changes in acoustic schwannomas are not different from vascular changes in gliomas. This is consistent with the fact that, in vitro, neoplastic Schwann cells also have angiogenic properties, like other tumor cells. The mechanism of hemorrhage in such tumors is not fully understood. It is postulated that ectatic hyalinized vessels may undergo spontaneous thrombosis, followed by tumor necrosis and hemorrhage. Alternatively, vascular tumors may be obliterated by endothelial proliferation. Recanalization by collateral meningeal vessels may occur, with hemorrhage into the necrotic tumor [12].

In conclusion, although quite rare, hemorrhagic acoustic schwannoma should be included in the differential diagnosis of cerebellopontine angle masses, especially when the onset of symptoms is acute and the patient is on anticoagulation treatment.

REFERENCES