Mixed vestibular schwannoma and meningioma without neurofibromatosis

Abdulrahman H. Al-Anazi, MD, KFUFNS, *Ibrahim M. Al-Luwimi,* MD, KFUFNS, *Mohamed A. Shawarby,* MD, PhD, *Tansu Mertol,* MD.

ABSTRACT

تم وصف توافق التعايش بين الورم السحائي والورم الشفاني كعنصرين مختلفين نسيجيا داخل الورم نفسه في الورم الليفي العصبي من النوع الثاني (NF2) . ولكن التوافق بين هذين الورمين بدون وجود الورم الليفي العصبي من النوع الثاني NF2 نادر جداً. وندون هنا حالة حدوث ورم مختلط نسيجياً بدون وجود دليل سريري لورم ليفي عصبي من النوع الثاني NF2. فقد لوحظ عند سيدة سعودية فقدان سمع تدريجي من الجهة اليسرى وتبين عندها بواسطة الرنين المغناطيسي MRI ورم مخيخي جسري في الزاوية اليسرى، وقد تبين بالتشخيص النسيجي أن الورم هو ورم سمعي شفاني مختلط نسيجيا مع ورم سحائي. نوقشت هذه الحالة بالأدب الاستعراضي

The co-existence of meningioma and schwannoma as 2 distinct histologic components within the same tumor has been described in neurofibromatosis 2 (NF2), but the co-existence of both tumors without evidence of NF2 is much rarer. Here, we are reporting a case of mixed schwannoma with meningioma without clinical evidence of NF2. In an adult Saudi lady with progressive left-sided hearing loss, left cerebellopontine tumor was diagnosed by MRI, and the histopathological diagnosis revealed that this tumor was composed of vestibular schwannoma and meningioma. This case is discussed with literature review.

Neurosciences 2009; Vol. 14 (4): 371-373

From the Department of Neurosurgery and Pathology, King Fahd Hospital of the University, King Faisal University, Al-Khobar, Kingdom of Saudi Arabia.

Received 24th February 2009. Accepted 7th June 2009.

Address correspondence and reprint request to: Dr. Abdulrahman H. Al-Anazi, Department of Neurosurgery, King Fahad Hospital of the University, King Faisal University, PO Box 40010, Al-Khobar 31952, Kingdom of Saudi Arabia. Tel/Fax. +966 (3) 8966743. E-mail: abdulalanazi@yahoo.com

Mixed tumors of schwannoma and meningioma component in patients with neurofibromatosis type 2 (NF2) were originally described by Cushing and Eisenhardt in 1922.[1] This kind of tumor has been documented in 12 subsequent case reports.[2] In this report, we present an unusual case of unilateral cerebellopontine angle tumor of mixed vestibular schwannoma and meningioma components without clinical evidence of NF2. This is the second reported case of tumor with mixed vestibular schwannoma and meningioma components. The first case was reported by Lüdemann from Hannover Medical Center.[3] The hypothesis on the pathogenesis of this kind of mixed tumor are discussed in light of literature.

Case Report. A 55-year-old Saudi lady was admitted to our hospital with a history of left-sided progressive hearing loss for 2 years, and tinnitus for few months in May 2007. A few weeks before presentation, she started to have left facial asymmetry with headache. Examination revealed horizontal nystagmus and left peripheral facial palsy with mild left cerebellar signs. Hyperkeratosis was present in the palms and feet with no clinical evidence of NF2. Audiometry showed profound left sensorineural hearing loss. An MRI showed left cerebellopontine angle (CPA) tumor of 3.5 cm x 3 cm diameters with intense contrast enhancement. The initial radiological diagnosis of this tumor was schwannoma (**Figures 1a & 1b**). She underwent gross total excision of the tumor via left retromastoid suboccipital craniectomy (**Figure 2**). The histopathology of the tumor showed schwannoma mixed with meningioma (**Figures 3a - 3d**).

Discussion. Mixed vestibular schwannoma associated with meningioma is an extremely rare tumor. They are thought to be exclusively associated with neurofibromatosis. Cushing and Eisenhardt were the first authors to report this entity in 1922.[1] Twelve more cases have been reported until this time. These reports described this mixed tumor as a primary tumor of vestibular Schwannoma interspersed with meningotheliomatous cells.[2] In our report, we discuss a second case without neurofibromatosis. The age of our patient is similar to the first case, but was different from the patient with NF-2, where presentation is typically earlier (in the second or third decade).[4] In

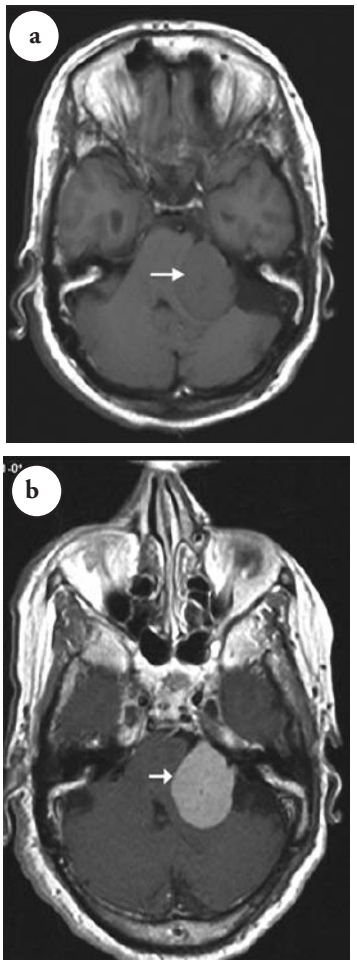

Figure 1 - An MRI of the brain **a)** revealing left cerebellopontine angle (extra axial) mass lesion, isointense signal in T1. **b)** Same with brisk contrast enhancement post gadolinium injection picture implying left vestibular schwannoma.

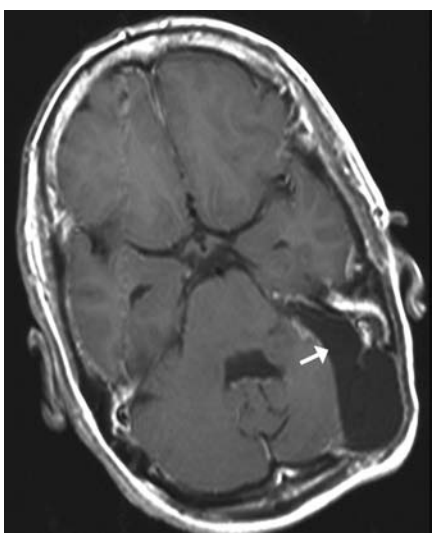

Figure 2 - An MRI of the brain TI image with contrast showing complete macroscopic excision of the tumor.

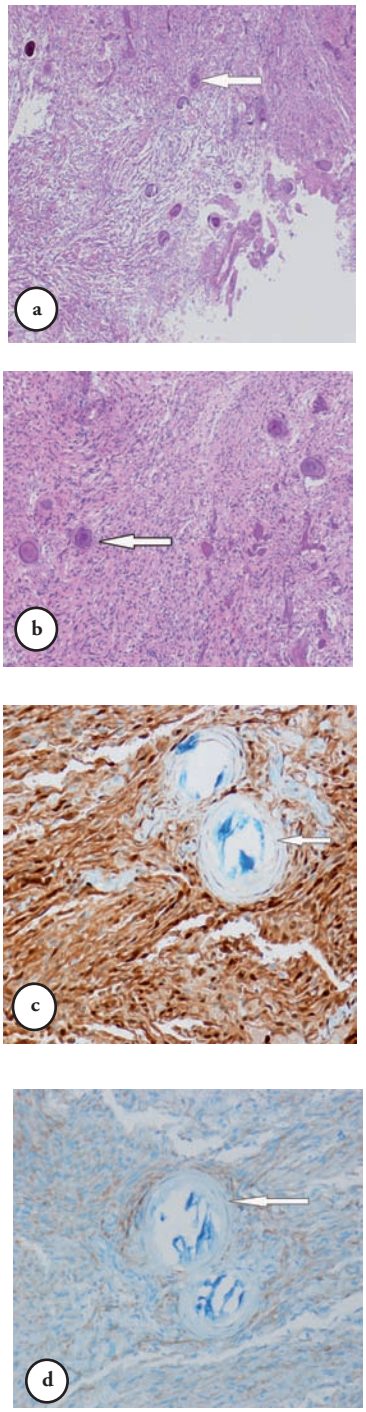

Figure 3 - Fascicles of **a)** spindly cells interspersed by numerous psammoma bodies. H & E x100, **b)** sheets of oval to spindly cells interspersed by numerous psammoma bodies. H & E x 200. **c)** tumor cells showing strong cytoplasmic and nuclear immunoreactivity for S-100 protein (brown color). Note also 2 psammoma bodies (non-reactive). Immunohistochemistry with DAB as chromagen x 200. **d)** section stained immunohistochemically for epithelial membrane antigen (EMA) with DAB as chromagen. Positive staining (brown) is restricted to only few cells, mainly around 2 psammoma bodies. Immunohistochemistry x 200. H & E - hematoxylin and eosin (stain).

the literature, there has been no explanation for the genesis of such mixed tumors. It was described simply as an unusual histological combination. Potential explanations for this phenomenon include tumor collision in which 2 tissue types emerge independently in the same location. A collision process theory may be supported by a clear "dividing plane between the 2 tumor components".[5] Additional mechanisms include differentiation from a common cell line, entrapment of hyperplastic arachnoid cells, or metaplasia within an acoustic neuroma. Vimentin staining as demonstrated in both the meningothelial and schwannoma elements of mixed tumors.[6] Schwannoma also express a growth factor known as Schwannoma-derived growth factor (SDGF), a member of the epidermal growth factor (EGF) family. This protein promotes neurite outgrowth and induction of neural differentiation markers such as GAP-43 and transin in initiating meningotheliomatous cell proliferation. A recent report[7] of an "EGF-like molecule" in the cerebrospinal fluid of a patient with NF and rapid growth of a vestibular schwannoma adjacent to a subsequently diagnosed meningioma potentially supports the possible role of paracrine growth factors in the development of meningioma in NF2. The same hypothesis might still be involved since no explanation for this association was found in the literature review, further studies are needed to explain this association.[2] There has been no typical MRI finding to differentiate these mixed tumors preoperatively.[8] In our case, the MRI appearance gave no indication of the mixed tumor. Histopathological examination is the best valuable tool for diagnosis of this kind of mixed tumors.

References

1. Cushing H, Eisenhardt L, editors. Meningiomas: Their Classification, Regional Behavior, Life History, and Surgical End Results. Springfield (IL): Charles C. Thomas; 1938.
2. Hoffman CE, Ament JW, Black P. Meningioma and NF2: A Review. ***Neurosurgery Quarterly Journal*** 2007; 17: 128-131.
3. Lüdemann W, Stan AC, Tatagiba M, Samii M. Sporadic unilateral vestibular schwannoma with islets of meningioma: case report. ***Neurosurgery*** 2000; 47: 451-454.
4. Eldridge R. Central neurofibromatosis with bilateral acoustic neuroma. ***Adv Neurol*** 1981; 29: 57-65.
5. Elizabeth J, Menon G, Nair S. Radhakrishnan VV. Mixed tumor of schwannoma and meningioma in a patient with neurofibromatosis-2: a case report. ***Neurol India*** 2001; 49: 398-400.
6. Kim DG, Paek SH, Chi GJ, Chun YK, Han DH. Mixed tumour of schwannoma and meningioma components in a patient with NF-2. ***Acta Neurochir (Wien)*** 1997; 139: 1061-1065.
7. Gelal F, Rezanko T, Uyaroglu MA, Tunakan M, Bezircioglu H. Islets of meningioma in an acoustic schwannoma in a patient with neurofibromatosis-2: pathology and magnetic resonance imaging findings. ***Acta Radiol*** 2005; 46: 519-522.
8. Pallini R, Tancredi A, Casalbore P, Mercanti D, Larocca LM, Consales A, et al. Neurofibromatosis type 2: growth stimulation of mixed acoustic schwannoma by concurrent adjacent meningioma: possible role of growth factors. Case report. ***J Neurosurg*** 1998; 89: 149-154.

Related topics

Saberi H, Khashayar P. Olfactory groove schwannoma masquerading as an orbital mass. ***Neurosciences*** 2008; 13: 73-76.

Kanno T. Surgery for petroclival meningioma and large sized acoustic schwannoma. ***Neurosciences*** 2008; 13: 27.

Deleu D, Salim K, El-Siddig A, Al-Hail H. Schwannoma of the vestibulocochlear nerve presenting as isolated paroxysmal vertigo. ***Neurosciences*** 2005; 10: 307-308.