Radiologically Indistinguishable Contiguous Meningioma and Schwannoma in the Same Cerebellopontine Angle in a Patient with NF2: Case Report and Literature Review

Sir,

Schwannomas are the most common tumors in the cerebellopontine (CP) angle (accounting for 80%–85%), followed by meningiomas (5%–10%).[1] Simultaneous occurrence of these tumors in the same CP angle is rarely reported in literature even in patients with neurofibromatosis-2 (NF2).[2,3] We report a case of 45-year-old female with a contiguous acoustic schwannoma and a meningioma in the same CP angle which were radiologically indistinguishable. We also discuss the possible hypotheses leading to such a condition.

A 45-year-old female patient who was a known case of NF2, presented to us with progressively increasing difficulty in hearing from both ears (right >left) for the last 2 years. She also complained of imbalance while walking for the last 1 month. She had undergone right frontal craniotomy and Simpson Grade 1 excision of right frontal parasagittal meningioma 2 months ago when she had presented to us with difficulty in walking and climbing stairs. On examination, she had right-sided Grade 2 facial palsy and bilateral (right >left) sensorineural hearing loss. Gag reflex was impaired, and her tongue was deviated toward the right side. On tandem gait, she had tendency to fall to the right side. Magnetic resonance imaging brain [Figure 1] done revealed a 4.1 cm × 4.3 cm × 3.2 cm intensely enhancing extra-axial lesion with central necrosis in the right CP angle cistern. Another similar lesion of size 1.9 cm × 2.1 cm × 1.9 cm was seen in left CP cistern. Both lesions were hypointense on T1 weighted images and hyperintense on T2 weighted images. The cisternal and intracanalicular portions of bilateral

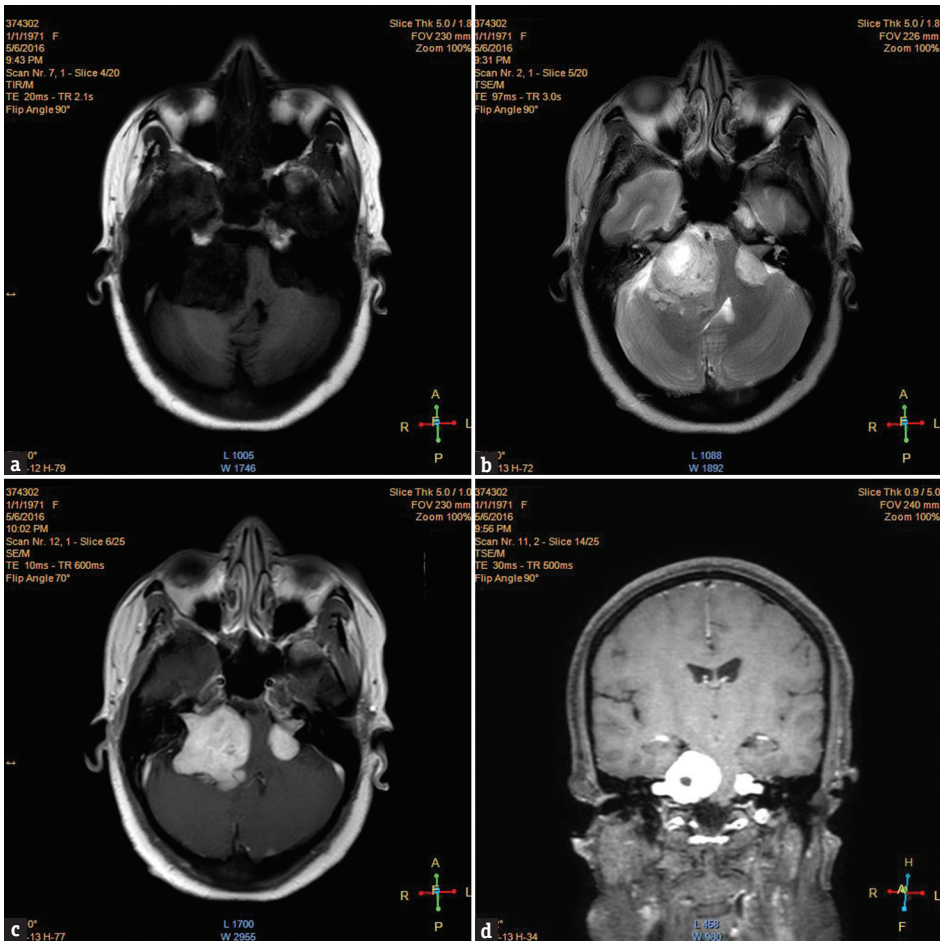

Figure 1: Magnetic resonance imaging brain (a) T1-weighted image showing hypointense tumor. (b) T2-weighted image showing hyperintense tumor. (c and d) Contrast image showing homogenously enhancing tumor in bilateral cerebellopontine angle

seventh and eighth nerves were not seen separately from the lesion suggestive of bilateral acoustic schwannoma (right >left). The patient underwent right-sided retromastoid suboccipital craniotomy in the lateral position. Two lesions were identified in the right CP angle after retraction of the cerebellum. The lateral part of the tumor was firm, vascular, fibrous, suckable, attached to posterior part of petrous bone suggestive of meningioma. The anterior part of the tumor was soft to firm, moderately vascular, and extending into the internal auditory canal (IAC) suggestive of schwannoma. Neurophysiological monitoring was used to identify VII nerve. IAC was then drilled and tumor extending into canal was removed. VII-VIII cranial nerves complex identified and preserved. Postoperative period was uneventful. Histopathology revealed that the anterior part of the tumor [Figure 2a and b] was composed of sheets and interlacing fascicles of cells along with Verocay bodies suggestive of a spindle cell tumor (schwannoma). The lateral part of the tumor [Figure 3a and b] was composed of a moderately cellular tumor with sheets and whorls of spindle to oval cells and few psammoma bodies suggestive of meningioma. On immunohistochemistry, focal positivity with epithelial membrane antigen (EMA) was seen.

Tumors of the CP angle comprise about 8% of all intracranial tumors.[4] Schwannomas form 80% to 85% while menigiomas form 5% to 10% of the CP angle tumors.[4] Simultaneous occurrence of these tumors in the same CP angle is a rare occurrence [Table 1].

NF2 is an autosomal dominant disorder affecting the central nervous system characterized by the occurrence of simultaneous bilateral acoustic schwannomas or family history of NF2 with either unilateral vestibular schwannoma or two other tumors such as glioma, schwannoma, or meningioma.[2,3] The disorder is caused by mutation of NF2 gene located on chromosome 22q12.2. Tumors in NF2 tend to present at an early age usually in the third decade[2,3] while in our case, the patient presented in the fifth decade.

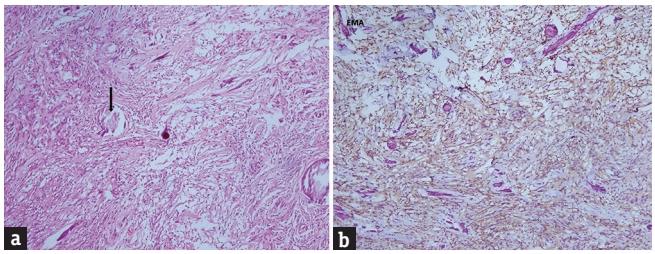

Figure 2: (a) Spindle cell tumor with hypo- and hypercellular areas suggestive of schwannoma. Verocay bodies (thin arrow) are seen (H and E, ×20). (b) Tumor is negative for epithelial membrane antigen

The radiological differentiation of schwannoma and meningioma has been well described in literature.[4,11] Schwannomas are usually rounded and are centered on the IAC. They cause expansion of the canal and are associated with heterogeneous enhancement. Meningiomas on the other hand are usually broad based, have a dural tail, and are located eccentrically outside the IAC. They are associated with hyperostosis in about 70% of cases and have a uniform contrast enhancement. In our case, the tumors were not radiologically distinguishable.

Schwannomas exhibit interlacing bundles of spindle cells arranged in compact or loose arrangement referred to as Antoni A or B, respectively. There may also be areas of dense concentration of cells with palisading nuclei (Verocay bodies).They are positive for S-100 and negative for EMA while meningiomas are EMA positive and S-100 negative. Both the tumors in our case were confirmed with immunohistochemistry [Figure 2b and 3b].

Simultaneous occurrence of intracranial tumors is a rare occurrence in the absence of phakomatoses or prior irradiation. Simultaneous or contiguous tumors must be differentiated from mixed tumors which are schwannomas associated with meningeal cell proliferation reported to occur in about 21% of NF2 patients.[2,3,12] Our patient is a case of "contiguous tumor" as the tumor was radiologically indistinguishable but histologically distinct.

The possible mechanism for such an occurrence seems debatable. The simultaneous occurrence of different tumors in the same location may occur due to metaplasia in the original tumor, collision of two different tumors, or differentiation into various cell lines from a common progenitor.[8,12] Till date, however, there is no evidence of a common progenitor cell for schwannoma and meningioma. Meningothelial reaction or hyperplasia adjacent to the main tumor (schwannoma) has been proposed as a plausible mechanism for such an occurrence and may have been responsible for contiguous tumors in our case.

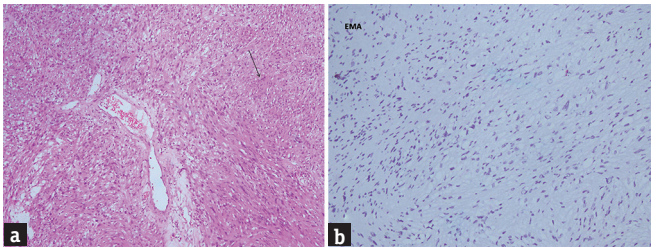

Figure 3: (a) Moderately cellular tumor with oval to spindle cells arranged in sheets and whorls suggestive of a meningioma. Psammoma bodies (bold arrow) are also seen (H and E, ×20). (b) Tumor is positive to epithelial membrane antigen

Table 1: Reported cases of meningioma and schwannoma in the same cerebellopontine angle

Serial number	Author	Year	Age/sex	Side	NF2	Radiologically distinguishable
1	Garder and Turner[5]	1939	48/female	Left	-	-
2	Thomassin *et al*.[6]	1991	64/female	Right	-	-
3	Wilms *et al*.[7]	1992	47/female	Right	-	+
4	Kim *et al*.[8]	1997	18/female	Left	+	+
5	Chandra and Hegde[9]	2000	35/female	Right	-	+
6	Elizabeth *et al*.[3]	2001	20/male	Right	+	-
7	Izci *et al*.[10]	2007	57/female	Left	-	+
8	Kutz *et al*.[11]	2009	43/female	Right	-	+
9	Grauvogel *et al*.[4]	2010	46/female	Left	-	+
10	Frassanito *et al*.[12]	2011	72/male	Left	-	+
11	Matyja *et al*.[2]	2012	18/male	Left	+	-
12	Matyja *et al*.[2]	2012	16/male	Left	+	+
13	Neto *et al*.[1]	2016	53/male	Right	+	-
14	Verma *et al*.[13]	2015	40/female	Left	-	+
15	Hoogar *et al*.[14]	2017	24/male	Left	-	-
16	Present case	2017	45/male	Right	+	-

NF2: Neurofibromatosis-2, +: Present/yes, -: Absent/no

Simultaneous occurrence of multiple tumors in the same location is a rare occurrence. It is more likely to occur in case of phakomatoses or postirradiation. In many cases, these tumors may not be distinguishable radiologically preoperatively. Histopathology leads to diagnosis in such cases which may be confirmed with immunohistochemistry.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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