Supratentorial convexity schwannoma unrelated to cranial nerves: Case report and review of the literature

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INTRODUCTION

The majority of intracranial schwannomas arises from the cranial nerves and usually occurs in the middle decades of life. Intracranial schwannoma not associated with cranial nerves is infrequent and usually involves in neurofibromatosis, and occurs among children or young adults. \(^{14}\) However, schwannoma, not related to cranial nerves nor neurofibromatosis Type I, is very rare entity. We describe a middle-age man with a supratentorial convexity schwannoma, and we review the previous reports about such cases, and then discuss the characteristics of these cases and their magnetic resonance image (MRI) findings.

CASE REPORT

A 48-year-old man experienced a sudden onset of seizures in his left lower limb. He had never previously experienced such an episode. Gadolinium-enhanced MRI (Gd-MRI) revealed a small, homogenous-enhancing mass in the peripheral aspect of the right parieto-frontal lobe. The
patient was referred to our hospital for further evaluation. The results of the physical examination performed at the time of admission were unremarkable. No cutaneous stigmata of neurofibromatosis Type I was observed. A neurological examination revealed no significant findings.

The lesion appeared as a well-defined, isosignal intensity area on T1-weighted MRI [Figure 1a], was hyperintense on T2-weighted MRI [Figure 1b], and also hyperintense on fluid attenuation inversion recovery MRI [Figure 1c]. The lesion demonstrated homogenous enhancement on Gd-T1-weighted MRI [Figure 1d-f]. Curvilinear enhancement was observed beneath the parietal bone but was unlikely to represent a “dural tail sign;” however, these findings are similar to convexity meningioma. T2-weighted MRI showed definite evidence of perilesional edema. Digital subtraction angiography showed some feeding arteries (right middle cerebral artery branch) [Figure 2a and b], right anterior cerebral artery branch [Figure 2c and d], and right middle meningeal artery (MMA) branch [Figure 2e and f]. Preoperative feeder embolization targeted to the right MMA branch was performed [Figure 2g and h].

Our preoperative diagnosis was an extra-axial tumor, such as convexity meningioma or other tumors. A whole-body evaluation revealed no evidence of malignancy. The patient underwent a parietal craniotomy, with total resection of the tumor. During surgery, tumoral invasion beyond the dura mater was observed [Figure 3a]. When the dura was opened, a well-demarcated, firm, spherical tumor was observed beneath the arachnoid membrane. The tumor was elastic, hard, grayish in color, and loosely adherent to but separable from the dura mater [Figure 3b]. The tumor was totally removed piece by piece, with the adjacent brain tissue and dura [Figure 3c]. Postoperative MRI showed the total removal of the tumor [Figure 3d-f]. The postoperative course was uneventful, and the patient was discharged without neurological deficits.

In light-microscopic assessments, the tumor was composed predominantly of Antoni Type A, and intermingled with Antoni Type B [Figure 4a]. Immunohistochemical staining was positive for S-100 protein [Figure 4b] but negative for glial fibrillary acidic protein, cluster of differentiation 34, epithelial membrane antigen [Figure 4c], signal transducer and activator of transcription 6 [Figure 4d], progesterone receptor, and synaptophysin. The histopathological diagnosis was schwannoma. The tumor was observed to be in the subarachnoid space extending to outer space of dura mater, intimately attached to the pia mater [Figure 4e and f]. Electron-microscopic studies were not performed.

**DISCUSSION**

Intracranial schwannomas are benign tumors that typically arise from the Schwann cells en-sheathing the axons beginning at the point of their exit from the pia mater. They usually develop among middle-age adults. Intracranial schwannomas comprise 6 to 8% of intracranial tumors. Intracranial schwannomas not related to cranial nerves have

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**Figure 1:** Preoperative magnetic resonance images (MRI). Preoperative MRI showed right frontal mass lesion. The lesion was isointense in T1-weighted MRI (a), hyperintense in T2-weighted MRI (b), hyperintense in fluid attenuation inversion recovery image (c), and homogenous enhanced with gadolinium intravenous injection (d: Axial image, e: Coronal image, and f: Sagittal image).
been reported to exhibit a predilection for occurrence among children and young adults.\textsuperscript{[14]}

Intracranial schwannomas not related to cranial nerves are relatively rare; 74 cases have been reported,\textsuperscript{[1-32,34-41,43,45-60]} in the English literature since the first article was published by Gibson et al. in 1966.\textsuperscript{[3]} The previous reports are reviewed in [Table 1]. There are 74 cases (Age: 0.5–84 years [average 31.7 years], male 42 cases/female 32 cases) reported previously. Cyst formation was observed in more than half of cases. Lesion location is dominant in intraparenchymal region (Convexity schwannoma (Left frontal: one case, Left temporal: two cases, Right frontal: three cases, Right parietal: one case, and Right occipital: one case), intraparenchymal schwannoma (Left frontal: four cases, Left temporal: eight cases, Left parietal: three cases, Left occipital: four cases, Right frontal: three cases, Right temporal: six cases, Right parietal: five cases, Right occipital: three cases, and Cerebellum: ten cases), Brain stem: two cases, Pons: four cases, Medulla:
**Table 1:** Clinical features of intracranial schwannomas (convexity) unrelated to the cranial nerves.

| Authors           | Year | Age | Gender | Symptoms      | Cystic | Size  | Location                  | Extent of Resection | Follow-up  | Postoperative state |
|-------------------|------|-----|--------|---------------|--------|-------|---------------------------|---------------------|------------|---------------------|
| Ghosh et al.      | 1992 | 27  | Male   | Seizure       | No     | 3.0 cm| Right frontal convexity   | Total               | N/A        | N/A                 |
| Frim et al.       | 1992 | 11  | Female | Seizure       | Yes    | 4.0 cm| Left temporal convexity   | Total               | 15 Months  | No symptoms          |
| Nakayama et al.   | 2002 | 53  | Male   | Seizure       | Yes    | 1.5 cm| Left frontal convexity    | Total               | N/A        | No symptoms          |
| Paredes et al.    | 2012 | 19  | Male   | Seizure       | No     | 1.0 cm| Right occipital convexity | Total               | 2 Years    | No symptoms          |
| Ma et al.         | 2013 | 24  | Female | Seizure       | Yes    | 8.0 cm| Right frontal convexity   | Subtotal            | N/A        | N/A                 |
| Wilson et al.     | 2016 | 34  | Male   | Incidental   | No     | 2.4 cm| Left temporal convexity   | Total               | 2.5 Years  | No symptoms          |
| Raswan et al.     | 2017 | 70  | Male   | Seizure       | No     | 8.0 cm| Right frontal convexity   | Total               | N/A        | No symptoms          |
| Present case      | 2019 | 48  | Male   | Seizure       | Yes    | 3.0 cm| Right parieto-frontal convexity | Total               | N/A        | No symptoms          |

It is reasonable to consider the possibility that intracranial schwannomas not related to cranial nerves may develop from ectopic Schwann cells, because there are no Schwann cells in the central nervous system. Proposed hypotheses include: (1) the presence of Schwann cells in the autonomic nerves of the subarachnoid vessels, (2) the conversion of pial cells into

three cases, intraventricle (Lateral ventricle: six cases, Third ventricle: one case, and Fourth ventricle: five cases)). Total removal was performed in 57 cases, and in 33 cases out of these 57 cases, good outcome was reported postoperatively. Especially, postoperative course in all cases of the convexity schwannomas is good (data shown in Table 2).
Schwann cells, and (3) the presence of sensory nerves in the meninges of the convexity.

Although several hypotheses have been proposed to explain the origin of ectopic Schwann cells within the central nervous system, the histological origin of these tumors remains controversial. Proposed hypotheses include:

**Schwann cell hyperplasia**

Riggs and Clary proposed Schwann cell hyperplasia as the origin of intracranial neurinomas; nonneoplastic Schwann cells accompanied by the perivascular nerve plexus, especially autonomic nerves, proliferate in certain chronic diseases of the spinal cord and may give rise to schwannomas.

**Schwannosis**

The term “Schwannosis” was introduced by Russell and Rubinstein, to indicate a hamartomatous lesion consisting of Schwann cells and related reticulin fibers in the substance of the spinal cord and pons.

**Reactive changes after injury**

Nelson and Rennels stated that nonneoplastic Schwann cells are sometimes observed in certain pathological conditions, such as multiple sclerosis, old infarctions, and certain experimental conditions, and they suggested that differentiated Schwann cells might arise as a result of tissue injury.

**The possible differentiation of multipotent mesenchymal cell into Schwann cells (Neural crest origin)**

Feigin and Ogata postulated that Schwann cells might be derived from multipotential mesenchymal cells of the neural crest.

In our case, the tumor was present not within the brain parenchyma but in the subarachnoid space, extending to epidural space. It is possible that schwannomas could arise from small nerves in the subarachnoid space or the leptomeninges. Although several cases of schwannomas attached to the dura have been reported, including our case, no authors confirmed that the tumors arose from dural nerves.

The imaging findings in our case were similar to convexity meningioma; however, there were no evident findings of “dural tail sign,” but the enhanced lesion extending along the meningeal or subarachnoid space; this location for schwannomas is exceedingly rare.

The differential diagnosis includes subdural, epidural, or leptomeningeal nodular brain metastasis, meningioma, glioma, and some granulomatous lesions (such as tuberculoma, fungal infections, and parasitic infections) when a well-demarcated spherical lesion associated with ring enhancement is observed adjacent to the cerebral convexity.

| Sites     | Cases | Average ages | Male/Female ratio (%) | Symptoms (%) | Cyst formation (%) | Average size (cm) | Detailed sites (%) | Total resection (%) | Postoperative ratio of no symptom or improved (%) |
|-----------|-------|--------------|------------------------|--------------|-------------------|-------------------|-------------------|-------------------|-----------------------------------------------|
| Convexity | 8     | 35.75 years old | 75                     | Seizure 87.5 | 50                | 3.86              | Frontal 50        | 87.50             | 100                                           |
|           |       |              |                        |              |                   |                   | Temporal 25       |                   |                                               |
|           |       |              |                        |              |                   |                   | Occipital 12.5    |                   |                                               |
|           |       |              |                        |              |                   |                   | Parietal 12.5     |                   |                                               |
|           |       |              |                        |              |                   |                   | Frontal 22.2      |                   |                                               |
|           |       |              |                        |              |                   |                   | Temporal 36.1     |                   |                                               |
|           |       |              |                        |              |                   |                   | Occipital 13.9    |                   |                                               |
|           |       |              |                        |              |                   |                   | Parietal 27.8     |                   |                                               |
|           |       |              |                        |              |                   |                   | Cerebellar hemisphere 66.7 |                   |                                               |
|           |       |              |                        |              |                   |                   | Vermis 33.3       |                   |                                               |
| Parenchymal | 36   | 23.63 years old | 55.60                 | Seizure 55.6 | Headache 27.8     | 61.10 | 4.18 | Frontal 22.2        | 88.90                             | 87.50                          |
|           |       |              |                        |              |                   |                   | Temporal 36.1     |                   |                                               |
|           |       |              |                        |              |                   |                   | Occipital 13.9    |                   |                                               |
| Cerebellar | 9    | 56 years old | 44.40                 | Gait disturbance 55.6 | 58.30 | 3.18 | Cerebellar hemisphere 66.7 | 66.70                             | 88.90                          |
| Brain stem | 9    | 34 years old | 66.70                 | Gait disturbance 88.9 | 55.60 | 1.79 | Vermis 33.3       | 66.70                             | 88.90                          |
| Ventricle  | 12   | 31.1 years old | 58.30                 | Headache 33.3 | Visual disturbance 50 | 83.30 | 3.19 | Pons 44.4          | 44.40                             | 77.80                          |
|           |       |              |                        |              |                   |                   | Medulla 44.4      |                   |                                               |
|           |       |              |                        |              |                   |                   | Midbrain 11.1     |                   |                                               |
|           |       |              |                        |              |                   |                   | Lateral ventricle 50 |                   |                                               |
|           |       |              |                        |              |                   |                   | Third ventricle 8  |                   |                                               |
|           |       |              |                        |              |                   |                   | Fourth ventricle 42 |                   |                                               |

Table 2: Clinical features of intracranial schwannomas unrelated to the cranial nerves.
These lesions are uncommon, except for meningiomas, gliomas, and metastases.

CONCLUSION

We have reported on a middle-age man with a schwannoma located in the subarachnoid space and have presented computed tomography and MRI findings. Although there was no diagnostic clue indicating a convexity schwannoma (because of the rarity and nonspecific imaging features of such tumors), it should be recognized that schwannomas may arise in such rare locations as the subarachnoid space.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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