

Two Cases of Subfrontal Schwannoma, Including a Rare Case Located between the Endosteal and Meningeal Layers of the Dura

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Abstract

Subfrontal schwannomas arising from the olfactory groove are rare and their origin remains uncertain because olfactory bulbs do not possess Schwann cells. We present two cases of subfrontal schwannomas treated with surgical resection. In one case, the tumor was located between the endosteal and meningeal layers of the dura mater. This rare case suggests that subfrontal schwannomas may originate from the fila olfactoria.

Key words: subfrontal schwannoma, anterior cranial fossa, Schwann cell, fila olfactoria

Introduction

Schwannomas of the anterior cranial fossa, termed subfrontal schwannomas, are extremely rare tumors of the central nervous system with clinical and radiological features similar to those of neuroblastomas and meningiomas in the same region. Subfrontal schwannomas are usually observed in younger patients compared to schwannomas associated with cranial nerves and are predominantly observed in males.¹⁾ Schwannomas usually arise from Schwann cells of peripheral or cranial nerves, whereas the origin of subfrontal schwannomas remains debatable because olfactory nerves lack Schwann cells.^{1,2)}

We report two cases of subfrontal schwannomas without neurofibromatosis. The tumor in one of our cases appeared to be located between the outer endosteal layer and the inner meningeal layer of the dura mater, suggesting that subfrontal schwannomas arise from the Schwann cells of nerves piercing the dura mater.

Case Reports

I. Case 1

A 44-year-old man presented with a 2-year history of anosmia and headache. Neurological examination

revealed no abnormalities except for anosmia. Head magnetic resonance (MR) imaging with gadolinium revealed a tumor measuring, 8 × 8 × 7 cm in the midline of the anterior cranial fossa (Fig. 1A–C), comprising multiple cysts and exhibiting heterogeneous enhancement. Computed tomography (CT) showed thinning of the bone surrounding the anterior cranial fossa (Fig. 1D). Cerebral angiography revealed no apparent vascularization. An interhemispheric approach was used for surgical resection and the tumor was found to be an encapsulated, solid, intra-dural tumor, comprising yellow fluid-filled cysts (Fig. 1E). The tumor was strongly adherent to both olfactory nerves, particularly on the left side, such that the left olfactory nerve could not be recognized. In addition, it was adherent to the cribriform plate and the meningeal layer of the dura mater, but we were able to detach it from the dura following gross total resection. The right olfactory nerve was preserved, but anosmia remained following surgery.

II. Case 2

A 40-year-old man presented with a 10-year history of headache, which recently showed a gradual increase in their severity. On examination, he was alert and well-oriented without any focal neurological deficits. Head MR imaging with contrast media revealed a 3 cm midline tumor in the anterior cranial fossa (Fig. 2A–C),

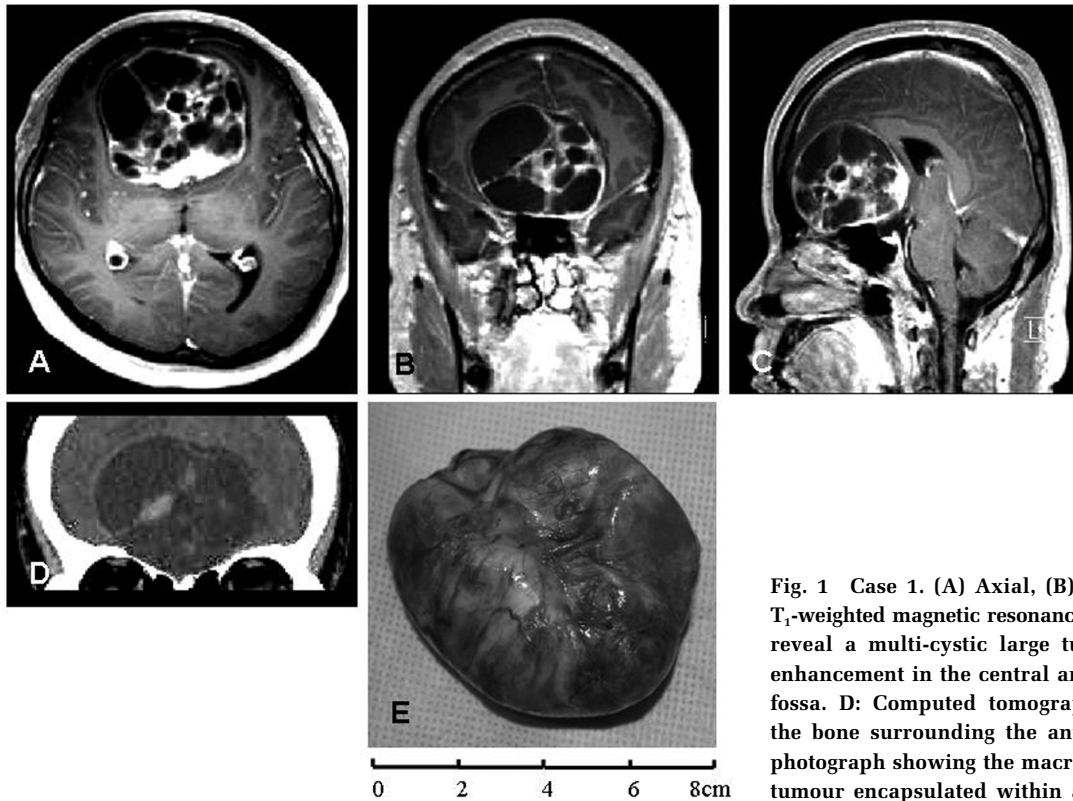


Fig. 1 Case 1. (A) Axial, (B) coronal, and (C) sagittal T₁-weighted magnetic resonance images using gadolinium reveal a multi-cystic large tumor with heterogeneous enhancement in the central area of the anterior cranial fossa. D: Computed tomography showing thinning of the bone surrounding the anterior cranial fossa. E: A photograph showing the macroscopic appearance of the tumour encapsulated within a solid membrane.

and CT showed thinning of the bone (Fig. 2D). As in Case 1, cerebral angiography showed no vascular staining. We used the interhemispheric approach to the anterior cranial fossa and observed that the dura was swollen at its convexity. After dissecting the dural enlargement, soft tumor tissue was found between the outer endosteal layer and the inner meningeal layer of the dura mater (Fig. 3). Membranes of the tumor were also recognized and gross total resection of the tumour with the membranes was performed. After resection, it was confirmed that olfactory tracts were compressed and stretched by the dura covering the tumor; however, no neurological deficits appeared after surgery.

III. Histopathological examinations

In both cases, histopathological examination revealed a biphasic pattern characterized by dense areas of spindle cells with elongated nuclei forming palisades (Antoni A) and less dense (hypocellular) areas (Antoni B) (Fig. 4). The tumor cells in both cases expressed S-100 but were negative for the epithelial membrane antigen (data not shown).

Discussion

Schwannomas only rarely arise within the parenchyma, ventricles, and other areas of the brain and spinal cord, which are not related to the cranial nerves. Because

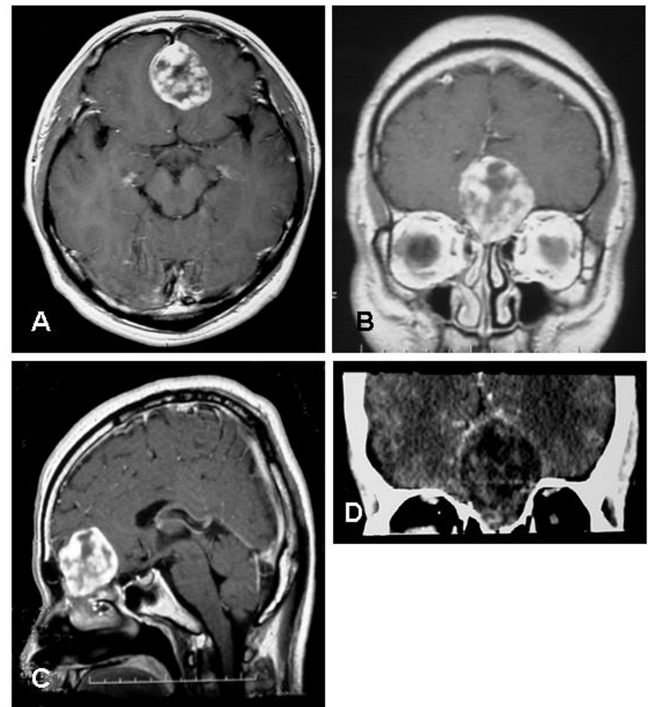


Fig. 2 Case 2. (A) Axial, (B) coronal, and (C) sagittal T₁-weighted magnetic resonance images using gadolinium reveal a tumor with heterogeneous enhancement in the central area of the anterior cranial fossa. D: Computed tomography showing thinning of the bone in the anterior cranial fossa.

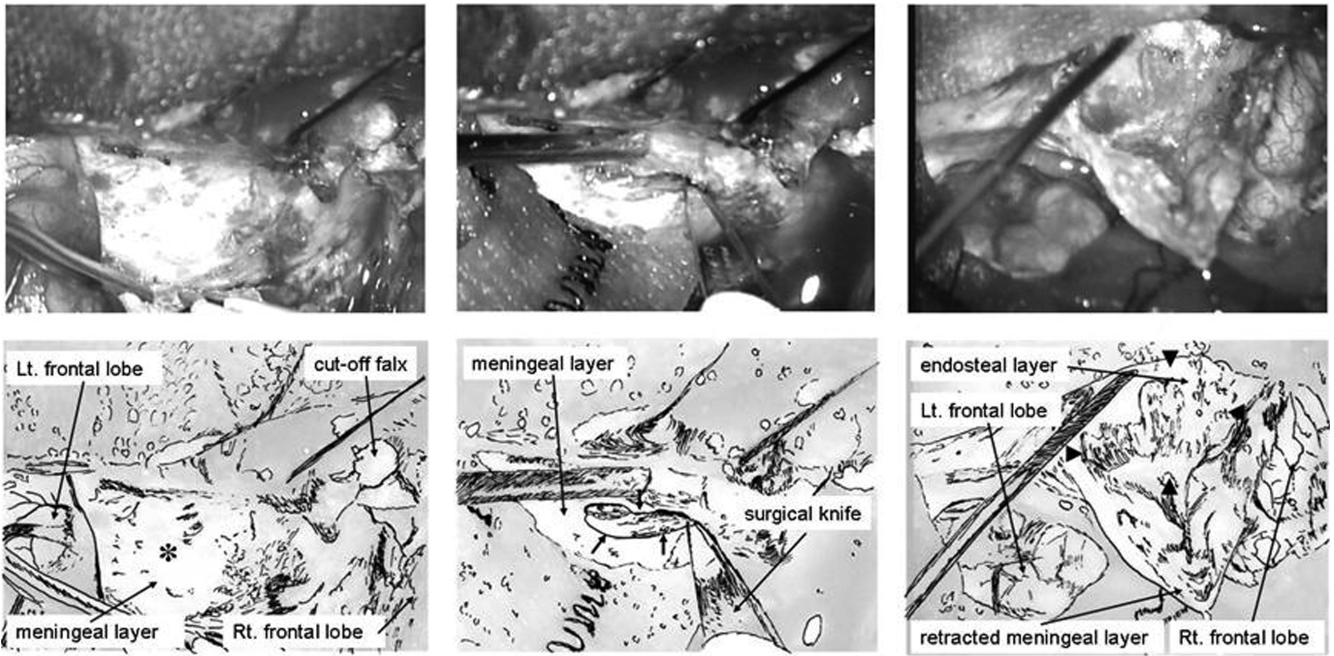


Fig. 3 Intraoperative photographs (*upper*) and illustrations (*lower*) in Case 2. Left: The eminence (*) of the tumor was covered by the meningeal layer of the dura mater in the anterior cranial fossa. Middle: The tumor (*arrows*) is exposed partly by cutting the meningeal layer of the dura using a surgical knife. Right: the cavity (*arrowheads*) after tumor resection has been shown. Also, the endosteal layer of the dura is exposed through the cavity.

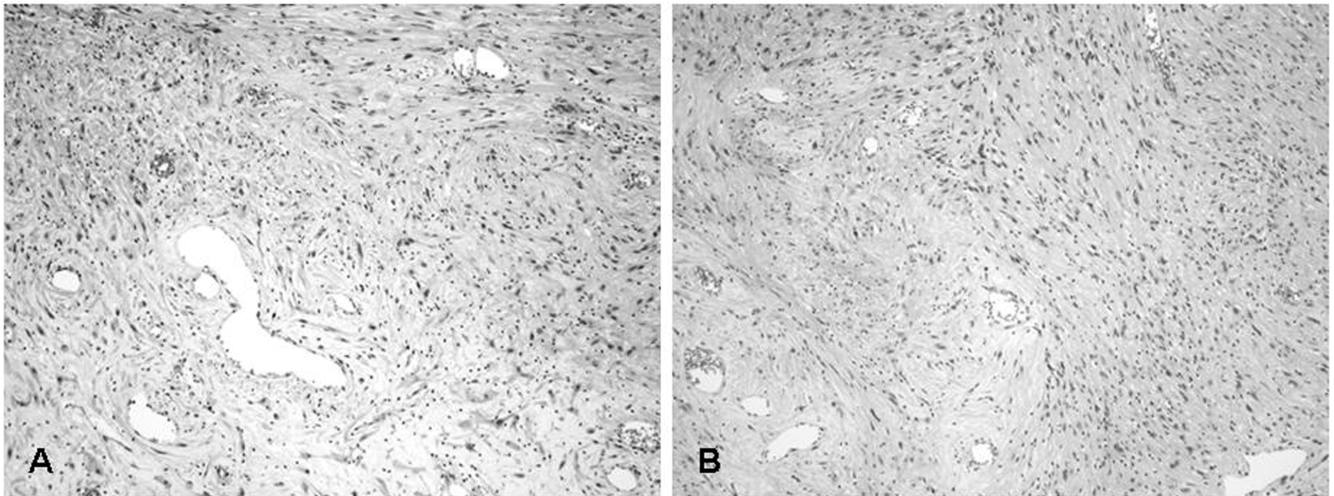


Fig. 4 Haematoxylin and eosin (HE)-stained tumor sections from Case 1 (A) and Case 2 (B) showing a biphasic pattern comprising compact Antoni A and hypocellular Antoni B areas (original magnification 100 \times).

schwannomas arise from the Schwann cells of the cranial nerves in most cases, the origin of such “ectopic” schwannomas is controversial.^{1,2} Approximately 40 cases of extra-axial schwannomas in the anterior cranial fossa (termed subfrontal schwannomas, olfactory schwannomas, or olfactory groove schwannomas) have been reported.^{1,2} Histopathologically, subfrontal schwannomas show no features that are distinct from other common schwannomas

such as vestibular schwannomas.

The origin of subfrontal schwannomas remains uncertain because olfactory bulbs do not possess Schwann cells. Several developmental and non-developmental hypotheses have been discussed to reveal their origins. The developmental theories include that mesenchymal pial cells transform into ectodermal Schwann cells or displacement of neural stem cells forms aberrant Schwann

cells, followed by giving rise to schwannomas in the neural “parenchyma,” but it remains uncertain whether subfrontal schwannomas in the “extra-axial” lesion can arise from those cells.¹⁾ Meanwhile, the non-developmental theories explain that Schwann cells that exist normally on adjacent structures such as the branches of trigeminal nerves, anterior ethmoidal nerve, perivascular nerve plexus, or the fila olfactoria.

Subfrontal schwannomas are usually intra-dural and extra-axial tumors in the anterior cranial fossa and frequently adhere to the cribriform plate and olfactory groove.^{1,2)} However, in one of our cases, the tumor was found between the outer endosteal layer and the inner meningeal layer of the dura mater. No previous report has described such a case and the location suggests that such schwannomas arise from the Schwann cells of nerves that penetrate through the dura mater.

Several investigators have speculated that subfrontal schwannomas may arise from meningeal branches of the trigeminal nerve and anterior ethmoidal nerves or the nerve plexus of dural vessels.^{1,2)} However, dural nerves and blood vessels run mainly between the dural endosteal layer and the calvaria^{3,4)} and such tumors originated from these nerves are not expected to be located in ‘intra-dural’ region which most of subfrontal schwannomas have been reported to be found in. Schwann cells associated with fila olfactoria are another possible origin of subfrontal schwannomas.^{1,2)} Several axons from the basal olfactory epithelial surface are enveloped by the cytoplasm of Schwann cells to form fine nerve fiber.⁵⁾ Some of these fibers join to form fascicles that are collectively termed the fila olfactoria. These fascicles pass the cribriform plate and pierce the endosteal and meningeal layers of the dura mater to reach the olfactory bulb (Fig. 5A).⁵⁾

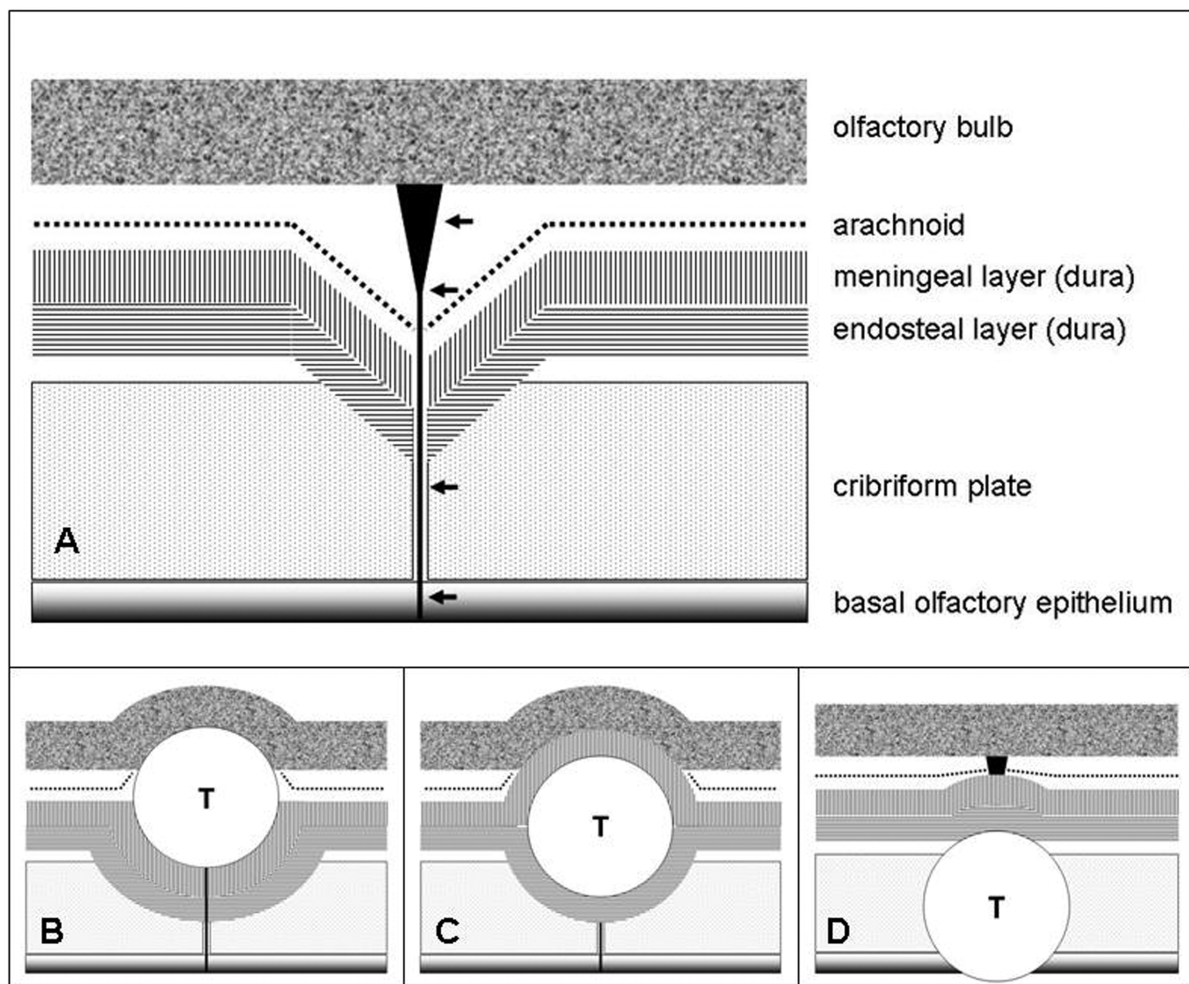


Fig. 5 Schematic illustrations showing the anatomy surrounding fila olfactoria (arrows) (A) and three types of schwannomas which may arise from the Schwann cells of fila olfactoria (B, C, and D). B: “Typical” subfrontal schwannomas arise from the Schwann cells of “intradural” fila olfactoria. C: The Schwann cells from fila olfactoria between the meningeal layer and endosteal layer of the dura can give rise to the rare subfrontal schwannomas as in case 2. D: The Schwann cells from “extradural” fila olfactoria may give rise to sinonasal schwannomas. T: tumor.

Therefore, Schwann cells of the fila olfactoria may give rise to schwannomas both beneath the dura and between the endosteal and meningeal layers (Fig. 5B, C). This theory can also explain why some subfrontal schwannomas, including one of our cases, lack anosmia. The fila olfactoria might be preserved partly if these tumors originate from the fila olfactoria and stretch out the olfactory nerve.²⁾ Furthermore, it is noteworthy that fila olfactoria has been suspected to give rise to rare sinonasal schwannomas which origin remains unclear.⁶⁾ Subfrontal schwannomas are associated with a younger age but the affected age in sinonasal schwannomas is various from childhood to the elderly. However, some sinonasal schwannomas extend into the cranial vault through the cribriform plate^{6,7)} and such cases have been reported to be observed in younger ages, the second to fourth decade,⁷⁾ that are similar to the ages in subfrontal schwannomas. We suggest that some of these schwannomas may also arise from a common source, the Schwann cells of fila olfactoria (Fig. 5D). However, a detailed description of Schwann cell markers in different regions and stages of development or transformation is necessary to determine the origin of specific Schwannoma cases.

Acknowledgment

The authors are thankful to Ms. Yumiko Ohishi for her technical assistance.

Conflicts of Interest Disclosure

The authors have no personal, financial, or institutional

interest in any of the drugs, materials, or devices in the article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms through the website for JNS members.

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