รายงานผู้ป่วย

Giant frontal skull base schwannoma: a case report

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Abstract:

A 20-year-old woman presented with a 2-day history of seizure. The physical examination revealed no neurological deficit. Radiological imagings showed a large tumor on the right frontal skull base. The tumor was surgically removed. Histopathology confirmed the diagnosis of schwannoma. Immunohisto-

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chemical studies revealed that the tumor cells were positive for S-100 protein and negative for both epithelial membrane antigen (EMA) and Leu7. One year later magnetic resonance imaging (MRI) showed no recurrent tumor. Solitary schwannoma arising from the anterior skull base is rare and less than 35 cases have been reported in the literature.

Key words: *frontal skull base, olfactory groove, schwannoma*

บทคัดย่อ:

คณะผู้รายงานได้นำเสนอรายงานผู้ป่วยหญิงไทยอายุ 20 ปี มาพบแพทย์ด้วยอาการชัก การตรวจร่างกายไม่พบความผิดปกติทางระบบประสาท ส่งตรวจทางรังสีวิทยาวินิจฉัย พบลักษณะเนื้องอกก้อนโตบริเวณ frontal skull base ด้านขวา ได้ทำการผ่าตัดนำเนื้องอกออก ส่งตรวจทางพยาธิวิทยา ผลการวินิจฉัยเป็น schwannoma โดยเซลล์เนื้องอกให้ผลบวกต่อการย้อมด้วย S-100 protein แต่ให้ผลลบต่อการย้อม epithelial membrane antigen (EMA) และ Leu7 หลังจากนั้นอีก 1 ปีได้ส่งตรวจคลื่นแม่เหล็กไฟฟ้าซ้ำ ไม่พบเนื้องอกโตขึ้นมาใหม่ solitary schwannoma ที่เกิดบริเวณ anterior skull base พบได้น้อยมาก มีในรายงานไม่ถึง 35 ราย จนถึงปัจจุบัน

คำสำคัญ: *ฐานกะโหลกศีรษะส่วนหน้า, เนื้องอก schwannoma, ร่อง olfactory,*

Introduction

Schwannomas are benign well-encapsulated tumors arising from the Schwann cells surrounding the peripheral nerves. Schwannomas arising from the anterior skull base are very uncommon. We report a case of a large schwannoma arising from the right anterior skull base. The purpose of this report is to share all the characteristic radiological findings and discuss of the tumor origin and immunohistochemical studies.

Case report

A 20-year-old woman presented with a 2-day history of seizure. The patient had no significant past medical history, including no family history of von Recklinghausen disease. The physical examination revealed no neurological deficit. A computed tomography (CT) scan of her brain showed an inhomogeneous enhancing mass in the frontal skull base. A non-contrast computed tomography (NCCT) scan revealed some parts of intratumoral and rim calcifications but no sclerotic change of the adjacent bone.

Magnetic Resonance Imaging (MRI) showed a large round mass arising in the right frontal skull base with inhomogeneous hyposignal intensity on a T1-weighted image (T1WI) and hypersignal intensity on a T2-weighted image (T2WI), measuring 8x8x7.4 cm. Heterogeneous enhancement of this tumor was noted on contrast enhanced T1-weighted images (Figure 1). Surrounding brain edema with secondary mass effect upon the adjacent brain was also

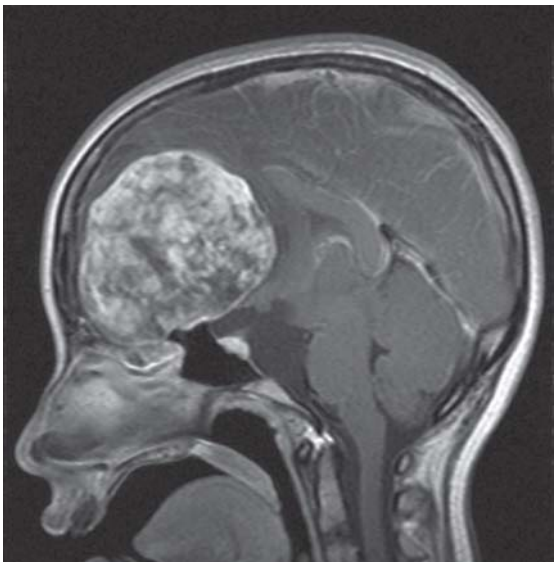

Figure 1 A large frontal skull base mass with heterogeneous enhancement on contrast enhanced T1-weighted images

observed, and the preoperative radiological diagnosis was frontal skull base meningioma.

The patient underwent total tumor removal via a bilateral frontal craniotomy. Intraoperative inspection revealed the tumor was attached to the tuberculum sellae and to the dura in the region of the cribiform plate. The right olfactory bulb could not be identified. After the tumor was totally removed, the dura was closed with a pericranial graft. The tumor was kept for pathological analysis. Serial sections of the gross specimen showed solid gray-white cut surfaces with hemorrhagic areas. Light microscopy revealed spindle-shaped cells in a biphasic pattern alternating between areas of highly cellularity, compact areas and loose, spongy areas of low cellularity (Figure 2A). Areas of nuclear palisading were also noted. Immunohistochemical studies were positive for S-100 protein (Figure 2B) and negative for both epithelial membrane antigen (EMA) and CD57 (Leu7). Electron microscopy demonstrated groups of cells with irregular nuclei with large nucleoli, and inter- cellular junctions were not observed. The cytoplasm was composed of the usual organelles with poor preservation. Cytoplasmic processes were also observed. Presence of basal lamina along the cell membrane was observed, however in some areas the discontinuities of the basal lamina were noted (Figure 2C).

The postoperative course was uneventful and the patient recovered quickly without any neurological deficits. One year later MRI showed no recurrent tumor but a right frontal subdural hygroma had developed. The patient had no clinically significant problems and the physical examination revealed no neurological deficit.

Discussion

We present a case of a giant frontal skull base schwannoma that shares all the imaging characteristics with other cases reported previously: an inhomogeneous enhancing mass in the frontal skull base with some intratumoral and rim calcification visible on CT, a decreased signal in T1WI and increased signal in T2WI on MRI and heterogeneous enhancement on contrast enhanced T1WI.

A solitary schwannoma arising from the anterior skull base is rare and less than 35 cases have been reported in the literature.[1-8] A frontal skull base schwannoma has been reported as originating from the olfactory bulb and nerve, the anterior ethmoidal

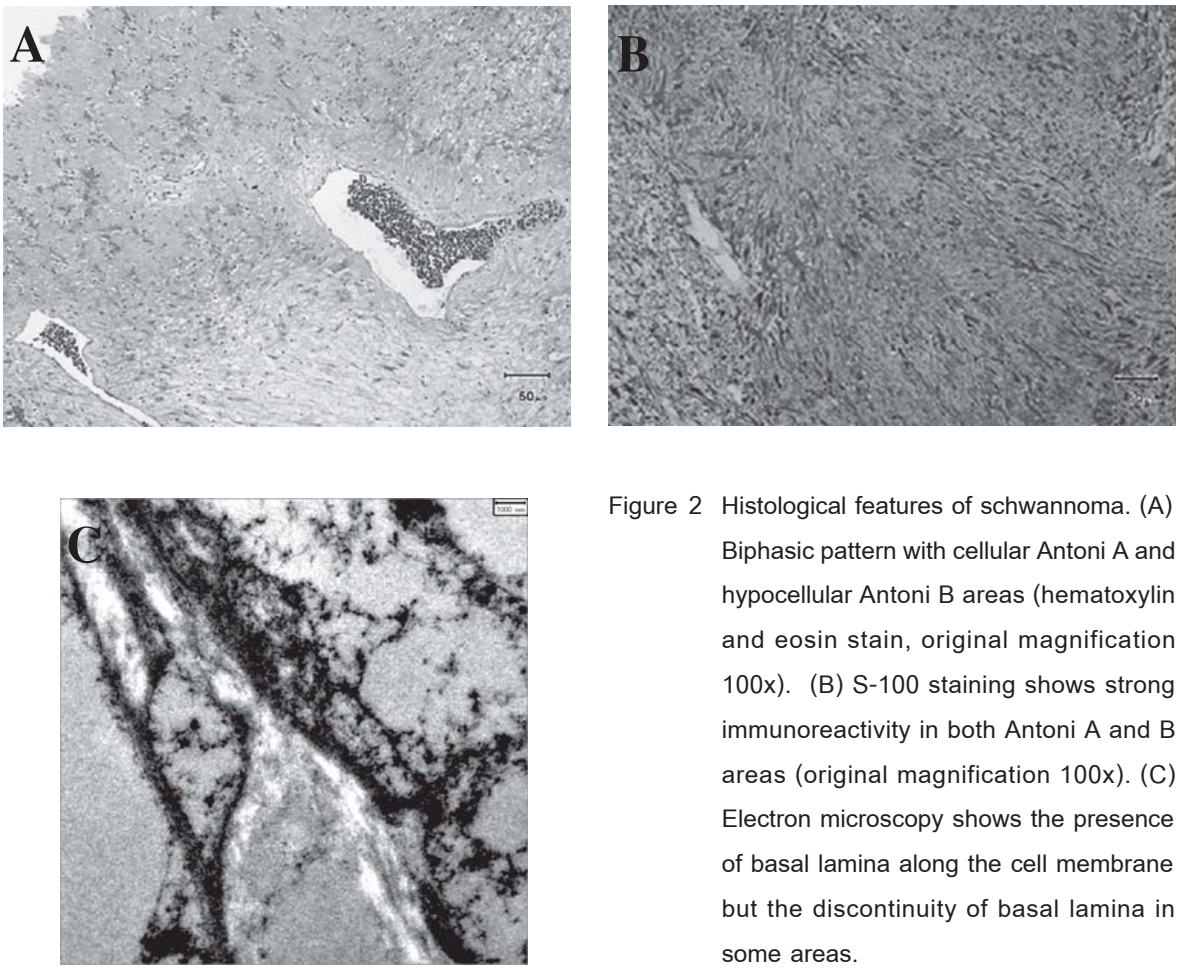

Figure 2 Histological features of schwannoma. (A) Biphasic pattern with cellular Antoni A and hypocellular Antoni B areas (hematoxylin and eosin stain, original magnification 100x). (B) S-100 staining shows strong immunoreactivity in both Antoni A and B areas (original magnification 100x). (C) Electron microscopy shows the presence of basal lamina along the cell membrane but the discontinuity of basal lamina in some areas.

nerve, the meningeal branch of the trigeminal nerve, an intracranial peripheral nerve such as cranial nerve zero (CN0), and the fila olfactoria.[1,3,7,9-10]

Frontal skull base schwannomas arising from the olfactory bulb and nerve have always been debatable on their origins, because the olfactory tract is a part of the central nervous system, which is devoid of a Schwann's cell layer.[1] Cranial nerve zero (CN0), also known as the terminal nerve or nervus terminalis, is present in humans as a plexus of small peripheral nerve fascicles found in the subarachnoid space that covers the gyri recti that lie between the olfactory bulbs and tracts.[10] The fila olfactoria is known to acquire a Schwann cell sheath approximately 0.5 mm beyond the olfactory bulb.[1,9] In our case, the right olfactory bulb could not be identified, indicating that the possible origin of the tumor might have been the olfactory bulb or nerve, the small peripheral nerves innervating the anterior cranial fossa, or the fila olfactoria. These are one potential source

of frontal skull base schwannomas with involvement of the adjacent olfactory bulb or nerve. Histological examination demonstrated a typical palisading pattern and immunohistochemical studies revealed that the tumor cells were positive for S-100 protein and negative for both epithelial membrane antigen (EMA) and Leu7. The presence of basal lamina along the cell membrane was observed through electron micro-scopy. All the findings were compatible with schwannoma, with the exception that the tumor was negative for Leu7, for which Schwann cells have previously been found to be positive.[11-12]

Our literature review indicated that frontal skull base schwannomas are more common in young males, but affect all age groups. There have been, including our case, less than 40% reported female cases. Common presenting symptoms and signs are headache, convulsion, and impaired vision without neurological deficits that are similar to the finding of our case.

Imaging features were nonspecific but there was frequently heterogeneous enhancement on contrast enhanced MR images, which we noted in our case. The most likely alternative diagnosis of an extra-axial frontal skull base mass is meningioma. Sclerotic changes of the adjacent bone are usually seen in meningioma that are not seen in our case and therefore raise the possibility of a schwannoma. However, signal intensity in MR images does not help the differential diagnostic distinction between frontal skull base schwannoma and meningioma.

Conclusion

We report a case of a giant frontal skull base schwannoma with discussion of the imaging characteristics and the likely origin. Preoperative diagnosis was difficult in this case and meningioma was a presumed radiological diagnosis. Although the origin of frontal skull base schwannoma is not certainly known, it should be recognized as the differential diagnostic possibility of a schwannoma in unusual location.

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