

Cystic Schwannoma of the Brainstem

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ABSTRACT: A rare case of cystic schwannoma occurring in the brainstem in the absence of von Recklinghausen's disease is reported. This appears to be the first case in the literature of a cystic schwannoma ever described in this location. While the exact origin of this tumor in this unusual location remains uncertain, different hypotheses are discussed and the possible origin of this tumor from perivascular elements in the brainstem is seriously considered.

RÉSUMÉ: Schwannome kystique du tronc cérébral Nous rapportons un cas rare de schwannome kystique du tronc cérébral en l'absence de maladie de von Recklinghausen. Il semble que ce soit le premier cas rapporté dans la littérature de schwannome kystique ayant une telle localisation. Bien que l'origine exacte de cette tumeur dans ce site inhabituel demeure incertaine, nous discutons de différentes hypothèses et nous envisageons sérieusement la possibilité que cette tumeur soit issue d'éléments périvasculaires du tronc cérébral.

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Solitary schwannoma comprises approximately 8% of all the primary intracranial tumors and the vast majority originate from the acoustic nerve.¹ This tumor has been previously reported to occur in the neuraxis, being more frequent in an intramedullary rather than an intracerebral location.²⁻¹⁵ A review of the literature has revealed only one case of brainstem schwannoma,¹⁶ which was solid, but no case of cystic schwannoma appears to have been previously described in this location. In this report, a case of cystic schwannoma of the brainstem is described along with the findings on computed axial tomography scan of the head, angiography of the posterior fossa and pathological examination.

CASE REPORT

This 46-year-old right-handed woman was admitted on July 14th, 1987 complaining of progressive blurred vision, dysarthria, dysphagia, left-side weakness and gait unsteadiness evolving for the last 6 months. The week before admission, she felt some transitory paresthesiae over the left side of her face.

Examination

Physical examination revealed no sign or stigmata of von Recklinghausen's disease. Visual acuity was 20/25 on both sides. There was no papilledema. On neurological examination, there was bilateral horizontal nystagmus. The patient exhibited right-sided cerebellar signs with ataxic gait. There was a proportional right-sided hemiparesis with cerebellar dysarthria. She also had marked hearing loss on the left side.

Laboratory

Studies revealed a normal CBC with the hemoglobin at 12.5 gm%. Serum electrolytes and the sedimentation rate were also normal. The E.E.G. showed mild diffuse nonspecific abnormalities. Computerized tomography scan of the posterior fossa demonstrated a large cystic lesion 3 cm. in diameter, localized ventrally to the fourth ventricle and

displacing it posteriorly. The third and lateral ventricles were slightly dilated. With contrast-medium infusion, there was enhancement of the solid portion of the lesion that was localized ventrally to the cystic portion and extended into the left cerebellopontine angle (Figure 1).

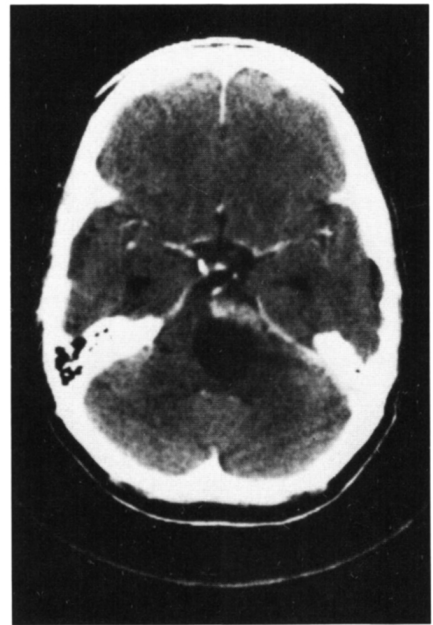


Figure 1 — Axial postcontrast CT scan shows a complex mostly cystic mass 3 cm in diameter, localized anteriorly to the fourth ventricle. Enhancement by contrast medium is noted in the ventral part of the lesion. The fourth ventricle is compressed and displaced posteriorly. A slight dilatation of the temporal horns of the lateral ventricles is noted.

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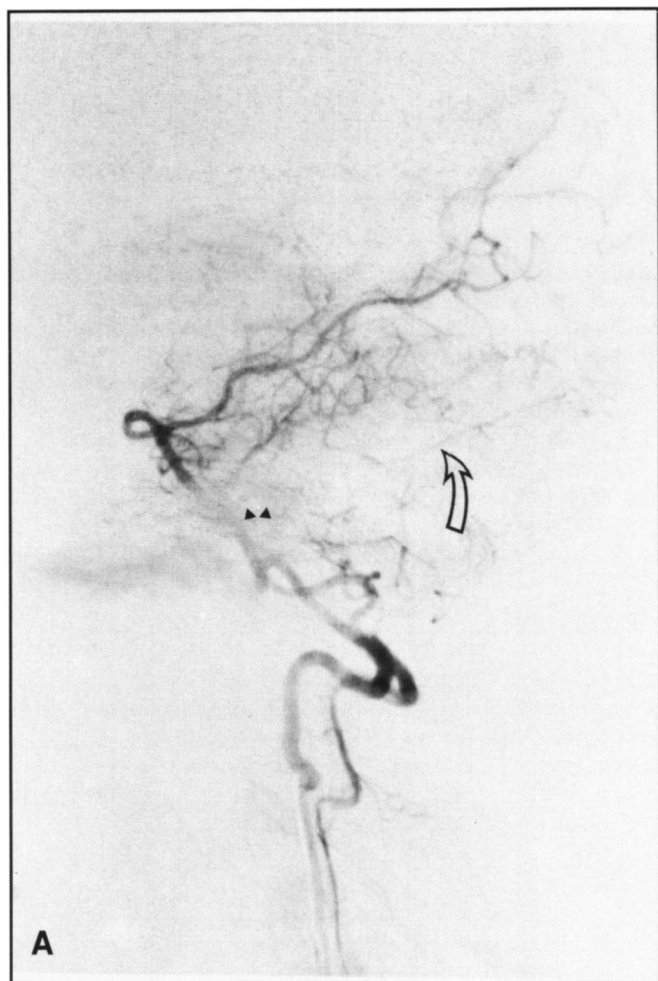


Figure 2A — Vertebral angiogram in lateral projection. Anterosuperior displacement of the first segment of the left anterior inferior cerebellar artery (AICA) (double arrowhead) and normal position of the marginal artery (curved arrow).

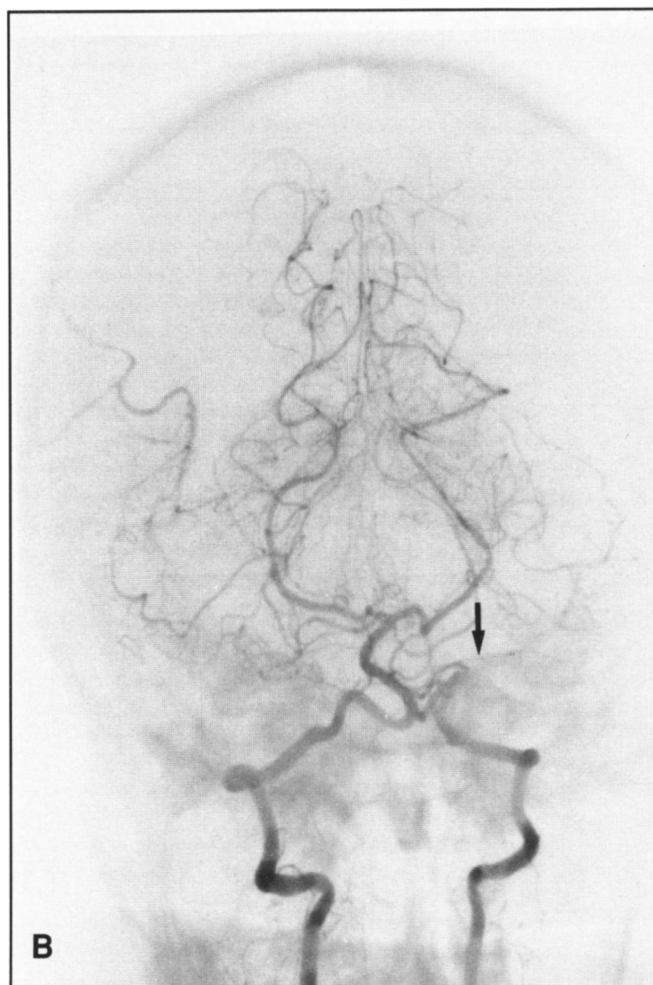


Figure 2B — Vertebral angiogram in antero-posterior projection. Stretching of the main trunk of the AICA, without significant displacement on the supero-inferior axis (straight arrow).

Vertebral angiography showed in lateral projection an anterosuperior displacement of the first segment of the left anterior inferior cerebellar artery (AICA) (double arrowhead) and normal position of the marginal artery (curved arrow) (Figure 2A). On the antero-posterior projection, there was a stretching of the main trunk of the AICA, without significant displacement on the supero-inferior axis (Figure 2B).

Operation

On July 28, 1987, the patient underwent a left paramedian suboccipital craniectomy with intraoperative monitoring of the short-latency somatosensory evoked potentials (SSEPs) obtained upon right and left median nerve stimulation. The cystic part of the tumor was localized by ultrasound and an attempt to puncture the cyst under ultrasound supervision was unsuccessful. On exposing the left cerebellopontine angle through a retromastoid approach, the extra-axial extension of the solid part of the lesion was encountered. Under microdissection, it was easily removed and there was no evidence of attachment to the fifth, the eighth or the lower cranial nerves. The solid intra-axial part of the lesion was just above the left fifth nerve and extended between the cerebral peduncles superiorly. Posteriorly, a cyst was indentified and opened. An oily yellowish fluid was drained. There was no plane of cleavage between the brainstem and the solid portion of the tumor preventing total removal of the mass without risking further brainstem damage and neurological deficit. This solid portion was yellowish and well vascularized. Its center was necrotic and easy to suction.

Post-operative course

Post-operatively, there was a significant improvement of the left-sided hemiparesis and the patient denied any diplopia. Dysarthria disappeared after 48 hours and positional vertigo after 10 days. She also complained of a decrease in hearing on the left side and an audiogram confirmed that only the high frequency sounds were preserved on that side. At the time of discharge, after one month of physiotherapy, she was autonomous in her daily living activities, walking with a cane. On follow-up, four months later, she was not using a cane anymore but complained of occasional transitory dizziness.

Pathological examination

The hematoxylin and eosin stain revealed a mixture of the two usual distinct patterns, Antoni A and B (Figure 3). In some areas, the tumor was composed of elongated bipolar fibrillated cells with small darkly staining spindle nuclei arranged in compact interlacing bundles. Formation of vague palisades was seen. In other fields, the pattern was much looser and less cellular with stellate pleomorphic tumor cells and a large number of foam cells. Reticulum stain demonstrated the presence of numerous strongly argyrophilic fibers running parallel to the long axis of the cells. Immunohistochemical localization of GFAP was negative but the protein S-100 reactivity was strongly positive for the tumors cells. The histological diagnosis was that of a classical schwannoma.

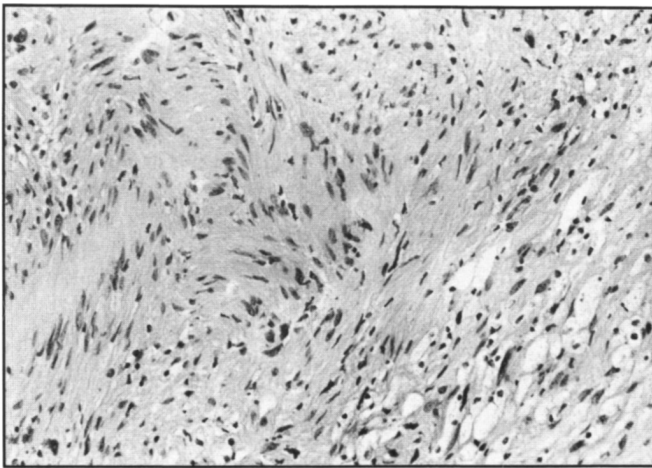


Figure 3 — Section of tumor showing palisading of the Schwann cell nuclei. Looser areas containing lipid-laden macrophages and pleomorphic nuclei are also seen (H & E, X 212).

DISCUSSION

Solitary schwannoma accounts for only 8% of all primary intracranial tumors, most of which arise from the acoustic nerves.¹ Very few cases of intracranial parenchymatous schwannomas have been previously reported in the literature. 1-10, 13, 14, 16 None of them was associated with neurofibromatosis.

All these tumors occurred in the cerebral or cerebellar hemisphere except one which was solid and localized in the brainstem.¹⁶ Most of them became manifest in childhood between 6 and 14 years of age and males were affected predominantly. In most cases, the evolution was uneventful after 6 months. This is in contrast to acoustic schwannoma which occurs mainly in women,^{1, 17} during the fourth and fifth decades.^{17, 18} These tumors have a tendency to grow slowly and there is an interval of a few years between the beginning of the symptoms and the diagnosis.

Although rare, spinal intramedullary schwannomas are more common than those in the intracranial location.^{8, 11, 15} They usually occur in the third and fourth decades of life and the interval between the beginning of the symptoms and the diagnosis is shorter as compared to acoustic schwannoma.

This case brings up many interesting points. First, despite the presence of an extra-axial extension of this tumor, there was no macroscopic evidence that the extra-parenchymal part arose from one of the cranial nerves, suggesting that the tumor grew from the brainstem parenchyma. Second, the intra-axial solid part of the lesion located ventrally to the intra-axial cystic component, was poorly circumscribed, had no plane of cleavage, and was not totally removeable. This is in contrast to previous reports recommending that parenchymatous schwannomas should be removed as completely as possible because they are circumscribed and potentially enucleable lesions¹⁶ like the encapsulated peripheral nerve tumors. Third, the radiographic findings by angiography and computerized tomography of the posterior fossa were suggestive of an intra-axial lesion pre-operatively and the differential diagnosis at that time was limited to cystic astrocytoma, hemangioblastoma, metastasis or a parasitic infectious disease like cysticercosis.

A valid question is whether this lesion was really in an extra-axial instead of an intra-axial location. Unfortunately, an NMR was not done before the surgery for technical reasons but the vertebral angiography was strongly suggestive, both on the lateral and antero-posterior projection of an intra-axial tumor.^{19,20}

The histogenesis of intracerebral and intraspinal schwannoma has been the subject of many controversies. Several theories concerning the origin of intramedullary schwannoma of the spinal cord have been previously put forward. The most attractive hypothesis suggests that parenchymatous schwannoma arises from the perivascular plexus of the spinal cord. According to Riggs and Clary, hyperplasia of the perivascular nerve plexus could be developmental or due to a chronic disease.^{12, 21} Russell and Rubinstein, observing the resemblance of pial cells to Schwann cells, have suggested that the pial cells might be converted to Schwann cells.¹ They even proposed the term "schwannosis" to indicate a hamartomatous lesion of the CNS where foci of Schwann cells and related reticulin fibers are embedded within the substance of the spinal cord. DeMyer has suggested the possibility that myelinated fibers with Schwann cells may be incorporated within the substance of the brainstem as a neuronal hamartomatous malformation and could form a nidus for the development of an intramedullary schwannoma.²²

The present case emphasizes the importance of tissue diagnosis in tumors of the brainstem before considering any specific treatment.

SUMMARY

A case of cystic schwannoma of the brainstem in a 44-year-old woman is described. The findings on cerebral angiographic and computerized tomography scan of the posterior fossa are reported. The diagnosis was confirmed on pathological examination. The importance of histological diagnosis of brainstem lesions is emphasized.

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