Extraspinal meningioma

Case report


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A case of ectopic extraspinal meningioma is reported which presented as a tumor at the back of the neck of a 51-year-old woman. This neoplasm was of considerable size and arose extradurally in the region of the C-2 nerve root, with its main bulk lying almost totally outside the spinal canal in the right suboccipital region. Total resection was accomplished in two stages, with no evidence of recurrence after 1 year. The rarity of such cases prompted this report.

KEY WORDS • spinal cord tumor • meningioma • extraspinal tumor

Spinal meningiomas are encountered mostly in the thoracic region in middle-aged women, but occur elsewhere along the spinal canal. They usually present clinically with signs of cord or root compression and are intradural extramedullary neoplasms; however, extradural meningiomas have been described, and all reported cases were reviewed recently. With the exception of one case, all tumors were contained within the spinal canal. We report a case of an ectopic extraspinal meningioma located at the C-2 level, which lay mostly outside the spinal canal.

Case Report

This 51-year-old woman presented for evaluation of a painless swelling in the right side of her neck behind and below the mastoid process. She became aware of this mass 4 months before seeking help. The swelling was ill-defined, firm, and deeply attached, occupying both the suboccipital region and the apex of the posterior cervical triangle. No evidence was found of a lesion in the ear, scalp, or pharynx, and the skull and cervical x-ray films were normal. The mass was assumed to be a benign lesion and, on this basis, the patient declined further investigations at that time. She was examined regularly thereafter in the surgical outpatient department. Two years later, she began to complain of neck pain, especially on looking to the right, and the swelling had enlarged sufficiently to prompt her admission to the hospital.

Examination. Biopsy showed a cellular spindle-cell tumor with psammoma bodies which involved a lymph node peripherally, apparently by direct spread. The patient was then referred to the Department of Neurosurgery for further treatment. Neurological examination was entirely normal. Faint calcification posterior to the spinous process of C-1 had now appeared on the lateral cervical x-ray film. Selective catheterization of the right vertebral artery clearly demonstrated a vascular tumor, gaining most of its blood supply from the meningeal branches of the right vertebral artery in the region of C-1 and C-2 (Fig. 1).

Operations. The tumor was exposed at the right side of the neck and subtotally resected. The deeper portion encircled the right vertebral artery, which was sacrificed in an attempt to achieve a radical excision. The patient recovered well with no neurological deficit, but it seemed likely that more tumor was left behind than was originally thought. Six weeks later, she developed a mild spastic tetraparesis, worse on the right side. A postoperative myelogram clearly revealed the extent of the remaining tumor compressing the upper cervical cord (Fig. 2).

At a second operation the meningioma was completely removed through an upper cervical laminectomy. The tumor was attached to the dura in the region of the right C-2 nerve root, and lay extradurally with no intradural extension. The dural attachment gave rise to a narrow neck extending posterolaterally into the
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Fig. 1. Vertebral angiogram showing a vascular extraspinal tumor supplied by meningeal branches of the right vertebral artery.

Fig. 2. Cervical myelograms showing extradural compression of the upper cervical cord on the right side.

Fig. 3. Photomicrograph of the excised tumor showing whorls of spindle cells with multiple psammoma bodies. H & E, × 145.

Fig. 4. Electron micrograph showing a tumor cell with an abundance of fine fibrils in its cytoplasm, interdigitating plasma membranes with desmosomes, finely granular nuclear chromatin, and sharply defined nuclear membrane. Uranyl acetate-lead citrate, × 410.

atlantoaxial ligament, which was thickened and vascular. The tumor expanded into a larger mass which had invaded and occupied the muscles of the right suboccipital region and the apex of the right posterior cervical triangle. Histologically, the tumor was composed of whorls of spindle cells with multiple psammoma bodies (Fig. 3). Electron microscopy studies of the tumor showed the characteristic features of a meningioma (Fig. 4).

Postoperative Course. The patient made a full satisfactory recovery from her second operation and was discharged home. She resumed her job as a shop assistant and there was no evidence of recurrence at 12 months postoperatively.

Discussion

If cutaneous and orbital meningiomas are excluded, ectopic meningiomas are rare tumors. Hoye, et al., grouped the published cases into the following four categories: 1) extracranial extensions of a primary intracranial meningioma eroding through the cribiform plate or the temporal bone; 2) meningiomas arising from cranial nerve sheaths with extracranial extension

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related mostly to the optic nerve but also to the trigeminal nerve; 3) extracranial meningiomas not connected to either the foramina or the cranial nerves, found in the infratemporal, subgaleal, and maxillary antrum and assumed to arise from embryonic rests of arachnoid cells; and 4) meningiomas reported in the cervical lymph nodes, lungs, and liver as metastases from a benign-appearing intracranial meningioma. The tumor in our patient, unlike the previously reported cases, did not fit into any of these four categories; it was an ectopic meningioma related to a cervical nerve root.

The tumor in this case probably originated from the arachnoid cell rests of the C-2 nerve root at its exit from the spinal canal[1,2] and occupied an extraspinal position. The extradural component of the tumor was far smaller than the extraspinal portion, yet it caused compressive symptoms relatively late, despite the total size of the meningioma.

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References


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