Unusual Massive Neurinoma in the Suboccipital Region

—Case Report—

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Abstract

An 8-year-old boy with no evidence of von Recklinghausen's disease presented with an unusual neurinoma manifesting as a gradually progressive swelling in the suboccipital region over 2 years. The lesion was massive and had widely eroded the posterior aspects of the atlas, axis, and suboccipital bone. The tumor had involved the dura of the sigmoid and transverse sinuses, was highly vascular, and had encased the ipsilateral vertebral artery. The tumor was almost completely resected although with considerable loss of blood through a large rent in the right sigmoid sinus. This unusual benign neurinoma most probably arose from the second cervical ganglion.

Key words: cervical root ganglion, craniovertebral region, schwannoma, neurofibromatosis

Introduction

Neurinomas may arise and grow in various unusual sites and result in a variety of clinical symptoms.^{2,6,7)} Peripheral and plexiform pattern of growth of a neurinoma is relatively common in patients with von Recklinghausen's disease.¹⁻³⁾ Large extradural components of the tumor are frequently encountered and commonly located in the mediastinum or prevertebral areas and presumably grow in the direction of the course of the nerve.⁹⁾ Neurinomas arising from the spinal root are usually dumbbell shaped with both intradural and extradural extensions.⁷⁾ Lesions with predominantly intradural or extradural extensions are not uncommon.^{2,7)}

We describe a case of massive neurinoma located predominantly along the midline in the suboccipital region.

Case Report

An 8-year-old school boy with no family history or stigmata of von Recklinghausen's disease presented with a swelling in the suboccipital region slowly progressive for 2 years. The swelling increased in size rapidly over the last year to achieve a massive size. On admission, the swelling measured approxi-

mately $9 \times 9 \times 7$ cm, extending from the external occipital protuberance to the mid-cervical region in the superoinferior direction and to both mastoid processes in width. The extension was slightly more towards the right side. The swelling was firm, solid, multinodular, and non-tender. The skin over the swelling was smooth and shining but normal and free from the underlying mass. There was no neurological symptom or deficit.

Magnetic resonance (MR) imaging showed the extensions of the lesion vividly (Fig. 1). The tumor was extradural, and isointense on T₁-weighted images and hyperintense on T₂-weighted images with homogeneous enhancement after contrast administration. The tumor had widely destroyed the suboccipital bone and posterior elements of the atlas and axis. The vertebral artery was encased by the tumor along its course adjoining the arch of atlas on the right side. Both sigmoid and transverse sinuses were in direct contact with the tumor. Angiography showed extensive tumor vascularity arising from the external carotid artery and right vertebral artery. The right occipital artery was greatly enlarged. The lesion had compressed the brainstem, upper cervical cord, and the cerebellum.

The relatively young age of the child, huge tumor size, extensive vascularity, encasement of the vertebral artery, and relationship with sigmoid and transverse sinuses were all factors indicating the necessity for surgery. The external carotid artery

Fig. 1 left: Sagittal T₁-weighted magnetic resonance (MR) image showing the large extradural isointense suboccipital tumor eroding the suboccipital bone and the posterior elements of the atlas and axis. center: Axial T₁-weighted MR image showing the huge dilated right occipital artery skirting the lateral aspect of the tumor. right: Coronal T₁-weighted MR image showing the medial and lateral extensions of the tumor.

Fig. 2 Immediate postoperative sagittal T₁-weighted magnetic resonance image showing excision of the tumor.

Fig. 3 Photomicrograph showing a moderately cellular neoplasm with Antoni A features consisting of spindle cells arranged in compact bundles and interlacing whorls. HE stain, $\times 400$.

feeders were embolized. At surgery, the tumor was found to be entirely extradural, significantly firm, and only moderately vascular. The tumor was radically resected piecemeal. The dura adjoining the venous sinuses was markedly thinned and a large rent in the right sigmoid sinus resulted in significant blood loss. The bleeding was controlled with difficulty with the help of gelfoam and wide pressure. Care was taken not to occlude the sinus. A

small part of the tumor encasing the vertebral artery was left behind as the patient had already lost a significant amount of blood and dissection around the vertebral artery was not considered safe.

Postoperative MR imaging showed almost complete excision of the tumor and the residual tumor around the vertebral artery (Fig. 2). Postoperative MR imaging venography did not show occlusion of the sigmoid sinus. The patient made an uneventful recovery. Histological examination confirmed the diagnosis of a benign nerve sheath tumor (Fig. 3). The specimen contained a moderately cellular ne-

oplasm consisting of spindle cells arranged in a fascicular pattern. There was no evidence of nuclear pleomorphism or mitosis. The patient was asymptomatic at follow-up examination after 8 months.

Discussion

Our patient presented with an extremely unusual neurinoma causing a predominantly midline and huge mass in the nape of the neck. The tumor had widely eroded the adjoining bones of the craniovertebral region. Such widespread bony erosion is also unusual for these benign and slow growing tumors. The dura was markedly thinned, particularly in the region of the venous sinuses, but was preserved in its entirety despite the wide bony destruction. The vertebral artery was encased by the tumor in its course adjoining the axis, suggesting that the tumor probably arose from the second cervical ganglion. Neurinomas arising from the second cervical ganglion are relatively common, but are usually dumbbell shaped, seldom achieve such massive size, and have never been reported to encase the vertebral artery. Neurinomas are usually firm and relatively avascular tumor masses. The extensive tumor vascularity was another unusual feature of our case. Histological distinction between spinal nerve root neurofibroma and neurinoma (schwannoma) has potential clinical implications. The histological patterns in neurofibromatosis types 1 and 2 also have different tumorigenic mechanisms.^{2,12} Although the tumor had achieved a massive size, histological examination revealed no evidence of malignancy. Radical resection is usually advocated and the long-term outcome after such treatment is excellent.^{4,5,8,10,11}

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