Case Report

Cystic cervical intramedullary schwannoma with syringomyelia

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We report a case of cervical intramedullary cystic schwannoma associated with segmental syrinx in a young adult without evidence of neurofibromatosis. The relevant literature is reviewed.

Key Words: Intramedullary tumor, schwannoma, spinal cord, syringomyelia

Neurilemmomas or schwannomas originating from the Schwann cells account for approximately 30% of primary intraspinal tumors.1 Spinal neurinomas are usually extramedullary intradural tumors. Because Schwann cells are not normally found within the parenchyma of the spinal cord, intramedullary schwannomas are rare.2 We report a case of cervical intramedullary cystic schwannomas associated with segmental syrinx in a young patient without evidence of neurofibromatosis.

Case Report

A 29-year-old man presented with a 3-year history of progressively worsening interscapular pain that radiated to the upper limbs. The pain was continuous, nondermatomal, and burning in nature. For about 1.5 months, he had progressive numbness and paresthesias of both upper limbs, and weakness of the left hand. Later, he developed stiffness in both lower limbs, and difficulty in walking and sphincter dysfunction. Examination revealed no evidence of neurocutaneous markers. Neurological evaluation revealed bilateral grip weakness with wasting of small muscles of the hand, causing claw-hand deformity of the left hand. Muscle tone and deep tendon reflexes were markedly increased in both lower limbs. Sensory examination was normal except for decreased pain sensation in the C6 to T12 dermatomes.

Magnetic resonance imaging (MRI) of the cervical spine revealed diffuse enlargement of the cervical spinal cord. The tumor was isointense to hypointense on a T1-weighted image and hyperintense on T2-weighted images. It showed a ring-like peripheral enhancement on Gadolinium injection. The tumor was extending into the root, causing its thickening [Figures 1 and 2]. There was a small segmental syrinx extending from the lower end of the tumor. At operation, the spinal cord was found diffusely enlarged from the C4 to C7 segment. A median myelotomy was done and the syrinx cavity was entered. At the upper end of the syrinx, the lower pole of the tumor was reached.

Figure 1: Magnetic resonance imaging of the cervical spine (sagittal and coronal section) shows diffuse enlargement of the cervical spinal cord. The tumor was iso- to hypointense in a T1-weighted image. It showed a ring-like peripheral enhancement on Gadolinium injection. There is also a small segmental syrinx extending from the lower end of the tumor

Figure 2: Magnetic resonance imaging of the cervical spine (coronal section) shows that the tumor was iso- to hypointense in a T1-weighted image and hyperintense in T2-weighted image (A, B). It showed a ring-like peripheral enhancement on Gadolinium injection (C). The tumor was extending into the root, causing its thickening (D)
The tumor was well defined and contained xanthochromic fluid. It was dissected all around and excised totally. Postoperatively the patient improved progressively. The histopathological examination revealed features typical of schwannoma. Immunostaining of the tumor cells with glial fibrillary acidic protein (GFAP) was negative but showed strong positivity with S-100 [Figures 3A-C).

**Discussion**

Schwannomas most commonly arise from nerve sheaths in the spinal canal and are usually located in the intradural extramedullary region. Intramedullary schwannomas are rare, representing 0.3-1% of spinal cord neoplasms.[1-3] Since the first case of intramedullary schwannoma reported by Kernohan in 1931, about 60 cases of solitary schwannomas of the spinal cord have been reported in the literature.[3] Rarely, intramedullary neurinomas can be multiple.[4] The probable origin of these tumors could be from (1) Schwann cells along the intramedullary perivascular nerve plexus, (2) Schwann cells ensheathing aberrant intramedullary nerve fibers, (3) transformation of pial cells of neuroectodermal origin into Schwann cells (4) central displacement of Schwann cells during embryonic development, and (5) from the dorsal nerve root entry zone and centripetal growth.[1,5]

Men are affected more commonly than women in a ratio of 2:1.[2] The age of the patients ranged from 9 to 75 years with a mean age of 40 years.[6] In nearly two-third of the patients, the cervical cord was affected followed by the thoracic cord in about 25% of cases.[2] In about 12% of reported patients, there was evidence of von Recklinghausen’s disease.[4]

The MRI signal intensity depends not only on macroscopic features such as solid or cystic components and degenerative processes but also on histological composition such as the presence of Antoni A or Antoni B tissue and melanotic variant.[3] A correct preoperative diagnosis is possible only when a predominant extramedullary component is present[5] or when the intramedullary spinal cord tumor is in continuity with a thickened spinal root.[3] The association of syringomyelia with intramedullary schwannomas is rare.[7]

Intramedullary schwannomas are benign tumors posteriorly located in the spinal cord. Hence, a complete resection is the treatment of choice.[2] Because of the rarity of Schwann cell tumors in the intramedullary region, diagnosis should be made with caution, and tumors that resemble schwannoma should be excluded using immunostaining with GFAP and S-100 protein.[8]

**References**