Cervical spinal intramedullary myxoma in childhood

Case report

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A unique case is reported of a cervical intramedullary myxoma in an 18-month-old boy who presented with torticollis and monoparesis. Radical removal of the tumor by a planned two-stage procedure was curative.

KEY WORDS • myxoma • intramedullary tumor • spinal cord neoplasm • children

M yxomas are uncommon neoplasms that are rarely encountered in neurosurgical practice. A unique case of a cervical intramedullary myxoma presenting in a child is reported.

Case Report

This 18-month-old boy was referred in August, 1985, for management of torticollis following an obscure history of cervical trauma. His symptoms started 2 months before with intermittent neck tilt to the left side and neck pain precipitated by movement, associated with progressive clumsiness of the right arm.

Examination. The patient was in distress, exhibiting spasm of the left sternocleidomastoid muscle (torticollis). Neurological examination revealed monoparesis involving the lower part of the right arm and the hand. There was weakness of the flexor muscles of the wrist and fingers and of the intrinsic muscle of the hand. The deep-tendon reflexes were absent. The remainder of the neurological examination, studies of the general systems, and the laboratory analysis were normal.

Plain radiographs of the cervical spine revealed widening of the interpedicular distances throughout the entire cervical canal with accompanying spina bifida occulta at the C-5 and C-6 levels. Omnipaque myelography demonstrated findings characteristic of an intramedullary tumor with expansion of the cord from C-3 to T-1 (Fig. 1).

First Operation. A laminectomy from C-3 to T-1 was performed. On opening the dura mater, the spinal cord was found to be expanded. Through a small midline myelotomy, a gelatinous tumor with poor vascularization was encountered. The lesion could not be dissected free of the surrounding cord. A biopsy was taken and frozen-section examination revealed intramedullary myxoma. Therefore, a midline dorsal incision was made in the cord over the entire lesion, but further removal of the mass was deferred. The dura was left open and a layer of Surgicel was placed over the tumor. The postoperative course was uneventful and there was no change in the patient's neurological status.

Second Operation. The wound was reopened 10 days later. The dark red-colored tumor was found to be almost entirely extruded and demarcated from the surrounding cord (Fig. 2). By microsurgical techniques the tumor was easily separated laterally but there was no clear plane of cleavage inferiorly. Nevertheless, it could be dissected from the cord tissue and almost totally excised.

Pathological Examination. The tumor consisted of a 4 × 1.5 × 1-cm, soft, tan-colored material. Microscopic examination revealed a hypocellular, poorly vascularized tumor. The tumor was composed of stellate and spindle-shaped cells with indistinct cytoplasmic boundaries. The cells were separated by an abundant myxoid stroma that stained strongly with Alcian blue but not with mucicarmine. Reticulin staining revealed
Intramedullary myxoma in a child

Fig. 1. Omnipaque myelogram showing fusiform expansion of the spinal cord shadow in frontal projection. Note the spina bifida and the widening of the interpedicular distances throughout the entire cervical canal.

Numerous fibrils coursing in various directions in some areas. Necrotic foci and mitoses were absent (Fig. 3).

Postoperative Course. The postoperative course was uneventful, and the preoperative symptoms resolved steadily. There was only minimal weakness of the flexor muscles of the fourth and fifth fingers by the 10th postoperative month, and at follow-up examination 2 years after operation he was asymptomatic.

Discussion

Myxomas are rare neoplasms, and even more uncommon in children. Their usual location is in the left atrium of the heart, but they have also been reported in the extremities, soft tissues, gastrointestinal tract, aorta, skin, jaw, nasopharynx, parotid gland, retroperitoneal tissues, spinal facet joint, and skeletal muscle.1-9

The rare occurrence of myxomas in children under 16 years of age has been emphasized by Dutz and Stout,3 who found only 12 cases published in the literature previous to their series of 15 patients. Recently, Chaves, et al.,2 reported two additional cases of childhood myxoma described as congenital, localized in the parotid region and the retroperitoneum. The associated finding of spina bifida and the clinical course of a myxoma which is a benign, slowly growing tumor might suggest a prenatal origin in the present case.

Tahmouresie, et al., 9 reported a case with thoraco-lumbar paraspinal muscle myxoma eroding the spine and causing spinal cord compression. Bell, et al.,1 recently described an epidural myxoma arising from the vicinity of the L3-4 facet joint. The uniqueness of our case is that the tumor originated from within the spinal cord and, to our knowledge, no such case has been reported previously.

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References


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