Unusual intradural spinal metastasis of a cranial chordoma

Case report

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In this unusual case, multiple intradural spinal cord implants of chordoma were removed 5 years after partial resection of an intracranial chordoma. The "seeding" had occurred in spite of combined surgical and irradiation therapy.

KEY WORDS - spinal cord tumor - chordoma - brain tumor - metastasis

THE chordoma is a tumor originating from the embryonic remains of the notochord. It occurs mostly in the sacrococcygeal region, less frequently in the skull, and rarely as a primary tumor in the vertebral column. It arises as an extradural tumor and compresses the brain, cord, or cauda equina. It may metastasize by a hematogenous route to distant tissues. However, subarachnoid spinal implantation from a cranial chordoma has only been reported once. This unique case is the second such report.

Case Report

This 41-year-old Negro man was admitted to the Hospital of the University of Oklahoma with the complaint of back pain, numbness and weakness of the legs, and impotence for 9 months.

Five years previously in 1964, he had complained of blurred vision, diplopia, and headache, and at another hospital, a neurosurgeon had noted a paralysis of the left fifth and sixth nerves. Skull films disclosed absorption of the left petrous ridge, and a right brachial arteriogram showed separation of the carotid and basilar arteries by a mass. At

Fig. 1. Chordoma of the brain removed in 1964. H & E, ×320.
Spinal metastasis of a cranial chordoma

a left temporal craniectomy, a mass that elevated and displaced the semilunar ganglion was biopsied after splitting the fibers of the second division of the fifth nerve. The mass was found histologically to be a chordoma (Fig. 1). A "moderate amount" of the tumor was removed, and the area was irradiated with 5500 R over 46 days. The headache and diplopia persisted, but otherwise the patient improved.

Examination. The patient was thin, ataxic, and the legs were weak and atrophied. The knee and ankle reflexes were reduced. There was no Babinski sign. The left eye showed ptosis, no motion, and a dilated pupil; acuity was reduced although the fundus appeared normal. The right eye was normal. X-ray films of the thoracolumbar spine were normal, while skull films disclosed erosion of the left petrous ridge with calcification in the middle fossa. An echogram was normal. Lumbar myelography was not successful; a cisternal myelogram disclosed a total block at T12-L1 due to a "multilobulated intradural lesion" (Fig. 2). The cerebrospinal fluid was clear and its protein content 101 mg%.

Operation. Laminectomy at T12-L2 disclosed multiple intradural implants over the dorsum of the cord and conus medullaris and throughout the cauda equina. The implants were firm and readily removed by cup forceps. A subtotal removal was carried out, and the dura left open. The specimen proved to be a chordoma comparable to that removed in 1964 (Fig. 3).

Postoperative Course. There were no postoperative complications. Two months after the laminectomy, a mass was found in the nasopharynx which also proved to be a chordoma; it was partially removed. Outpatient radiation therapy has been attempted but has been inadequate because of the patient's failure to appear for therapy. His neurological examination is relatively unchanged 1 year after surgery.

Discussion

Midway through the 19th century, pathologists8,11,17 first recognized that certain excrescences from the sphenoid bone resembled remnants of the notochord. It was not until 189415 that they were termed "chordomas." It is interesting to note that the first authentic case of a spinal chordoma was recorded by Raul and Diss14 in 1924. Pototschsnig13 in 1919 first recorded the metastatic potential of this tumor.

All authors reporting large series of chordomas recognized that they are least frequent in the spine, particularly in the thoracic area.1,5,7,9,10 Higinbotham, et al.,5 believed the metastatic potential of this lesion is greatly underestimated, for 43% of their series showed evidence of metastases, local or distant. Fox, et al.,4 indicated that these tumors of sacrococcygeal or cranial origin may metastasize to the regional lymph nodes, liver, lungs, heart, bone, adrenals, thyroid, pancreas, kidneys, urinary bladder, peritoneum, pleura, and skin. They noted that only 10 cases in the literature showed evidence of widespread metastasis.

The cases of vertebral origin have arisen in the extradural space and have destroyed or extensively involved the vertebral bony structure. Poppen and King12 noted that the vertebral lesions may be very difficult to differentiate from a metastatic carcinoma. They
pointed out that “penetration of the dura may lead to the correct diagnosis” since dural penetration is rare with any other lesion.

Several authors\textsuperscript{16} have noted eventual spinal dural erosion after a long period of extradural compression. However, there is only one report\textsuperscript{16} of primary intracranial tumor giving rise to subarachnoid spinal implantation. This 59-year-old man had a primary tumor of the clivus with intradural spinal subarachnoid implantation; he died of pyelonephritis and uremia, and the lesions were shown to be chordomas at both sites.

It is worth noting that an acceptable dosage of radiation had been administered to our patient but that this did not affect the ability of the lesion to metastasize.

References

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