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Intraspinal Chordomas

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Summary

Intraspinal chordomas are rare tumors, comprising about 1% of central nervous system tumors. They are derived from notochordal elements which have failed to degenerate and are mainly found in the sphenooccipital and sacrococcygeal regions. These tumors are very slowly growing, are virtually impossible to excise completely and only metastasise in about 10% of cases. This paper reports on 3 cases of chordomas at 3 different levels and thus causing 3 different clinical pictures. The therapeutic possibilities are outlined.
Key words: *Intraspinal chordomas; Paraplegia; Tetraplegia; Hemiplegia; Metastasis.*

Introduction

Spinal cord tumors are uncommon. Different reports put the average annual incidence at between 0·9 and 2·5 per 100 000 population (Kurland, 1958; Leibowitz, 1971). Intraspinal chordomas represent approximately 2% of all spinal cord tumors (Rand, 1963, Leibowitz, 1971) which means that the annual incidence in Norway (population 4 million) should be between 0·7 and 2 and the prevalence about 8–11.

Chordomas are derived from remnants of the foetal notochord which in the foetus extends from the buccopharyngeal membrane to the last coccygeal segment. The notochord becomes incorporated along most of its length into the vertebral column and ultimately contributes to the formation of the nucleus pulposus of the intervertebral disc. At the ends of the skeleton the fate of the notochordal tissue is more variable. In rare instances there may remain a nest of notochordal cells. The location of aberrant notochordal tissue at these two sites, the base of the skull and the sacrococcygeal region correlates well with the location of chordomas in later life. 35–40% of all chordomas arise at the base of the skull, about an equal percentage appear in the sacrococcygeal region and only about 20% elsewhere in the spine.

Chordomas usually appear in middle life. Local pain without any neurological signs is a common presenting complaint and the diagnosis is often not made until the patient develops signs of nerve root or spinal cord compression. There are no typical radiological findings (Hudson, 1983). As with many other neoplasms one may find foci of vesicular osteolysis accompanied by reactive zones of sclerosis. Soft tissue extension is difficult to detect. At operation the

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macroscopic appearance is of a tumor with a nodular but smooth surface of greyish-white colour. The cut surface is translucent and mucinous in consistency. The microscopic appearance is one of cords and masses of large epithelial-like cells that are sharply delineated and have a dark, eccentrically placed nucleus. The tumors grow slowly, survival of up to 23 years having been observed. Chordomas do not usually metastasise, according Maccarty (1961) this occurs in less than 10% of cases.

Case reports

Case 1. A female, born 1953, developed her first symptoms of lower back pain in February 1978. Lumbar myelography showed an intraspinal tumor at the level of L5 vertebra. In August 1978, an operation was performed, a laminectomy of S1–S4 and removal of tumor was carried out. Postoperatively she was free of pain and had no neurological sequelae. She became pregnant and was delivered by caesarian section in July 1981. Towards the end of her pregnancy she developed urinary retention, sacral paraesthesia and increasing muscular weakness in her lower limbs. There were no radiological signs of recurrence of the tumor at this stage. She was able to walk without support until late in August 1983 when she was readmitted because of increasing weakness in her lower limbs. A laminectomy of L5 was done and approximately 3 kgm of tumor was removed. The tumor was situated intraspinally in the sacral region and retroperitonelly in the pelvis. Postoperatively she had an incomplete paraplegia below L5 with sacral analgesia, absent bulbocavernous reflex and hyperactive bladder. Six months later she was again able to walk but had decreased power distal to her knees and in the gluteal regions. Her next admission to our hospital was in January 1985 and she has remained an inpatient since then. There has been a progressive worsening in her neurological and general condition and serial CT-scans has shown the tumor to be increasing in size. After her second operation she had refused x-ray therapy because she wanted more children. Since we did not have any specific therapy to offer her, she was allowed to use injections of Iscador (Mistletoe) for some months without any effect on her condition. She was reoperated on in September 1985 when 5 kgm of tumor was removed from her pelvis. Her condition is now as previously described with an incomplete paraplegia below L4 motor and sensory, urinary incontinence and obstipation. She has severe back pain and requires narcotic analgesics. On her last admission a swelling was noticed below her right knee. X-Ray studies showed a probable metastases and she has since developed multiple skeletal metastases which have been confirmed by isotope scan and biopsy.

Case 2. In 1979, a male, born 1925, developed pain in his left upper limb and shoulder increasing in intensity and distribution until it involved the left side of his thorax in 1983. He was not properly investigated until he showed signs of an upper motor lesion in his lower limbs in May 1983. CT-scan showed a posteriorly placed mediastinal tumor with bony destruction and involvement of intraspinal structures. At operation in May 1983 a laminectomy from Th1–Th4 was carried out and an intraspinal chordoma was partially removed. During the postoperative months he experienced increasing thoracic pain and weakness of his lower limbs, and he was reoperated on in March 1984. He received high voltage irradiation postoperatively. His condition improved somewhat but after

a few months he once again experienced increasing pain, difficulties in walking and ataxia in his lower limbs. His condition gradually deteriorated and when last examined in December 1985 he had an incomplete spastic tetraplegia below C7. The sensory level was at C7 on the left side and at Th5 on the right. He was transferred to a nursing home and has since died. Just like the other two patients, his pain was intense and he required large doses of narcotic analgesics.

Case 3. A male patient, born 1953, had his first symptoms when he was 10 years old in 1964. These were; neck pain, decreasing power in upper and lower limbs, ataxia and dysarthria. He received high voltage irradiation towards the base of his skull and all symptoms and signs disappeared for about 3 years. In 1967 a biopsy from a recurring epipharyngeal tumor showed that it was a chordoma and x-rays showed an occipital tumor stretching as far down as C3. He was once again irradiated and received a total dose of 7700 RAD in 1964 and 1967. After this he was free of symptoms for about 13 years, his only complaint being occasional neck pain. He received an education and was working full time until 1980. Since then he experienced radicular symptoms in his upper limbs and weakness of all four limbs. Part of the tumor was removed in 1981 and he was left with a left sided spastic hemiplegia. He has slowly deteriorated and was reoperated on 3 times during 1985. Once to remove part of the tumor, and twice to insert and check a ventriculoperitoneal shunt that was found necessary when he developed hydrocephalus. He is now hemiplegic, has left sided tongue atrophy and atrophy of his right trapezius muscle. There is no sensory loss and he has normal bladder and bowel control. He experiences much pain and uses narcotic analgesics.

Conclusion

All three patients had their chordomas for several years (range 6–23) and each had several operations to alleviate symptoms. This is typical for chordomas since they appear to be impossible to remove completely because of their inaccessibility and involvement of nerves and blood vessels. Multiple operations are performed to relieve symptoms and these often result in prolonged periods of improvement. High voltage irradiation is also recommended, preferably postoperatively (O'Neill, 1985). Our last two patients received radiation therapy and this certainly seemed to help for a while. However, the long term prognosis still remains poor particularly since chemotherapy does not appear to affect these tumors.

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