Chondrosarcoma of the cervical spine

Case report

RUSSELL L. BLAYLOCK, M.D., AND LUDWIG G. KEMPE, M.D.

Division of Neurological Surgery, Medical University of South Carolina, Charleston, South Carolina

The authors present a case of chondrosarcoma involving the lateral mass of the C-2 vertebra, treated by an apparent total removal of the tumor. A review of the literature revealed that these patients can be cured in a significant number of cases if a radical removal is accomplished.

KEY WORDS • **chondrosarcoma** • **cervical spine** • **spinal tumor**

PRIMARY malignant tumors of the bone involving the vertebral axis are extremely rare. Of this group of osseous tumors, the chondrosarcoma has been reported to involve the vertebral axis in less than a dozen cases. We are presenting a case of chondrosarcoma primarily involving the lateral mass of the C-2 vertebra.

Case Report

This 43-year-old man had been in good health until early in 1971, when he began to have recurring left suboccipital headaches; over a period of 1 month these became persistent. He was seen by a neurosurgeon at that time and was diagnosed as having occipital neuralgia. He subsequently underwent an occipital neurectomy that left him essentially free of pain and otherwise asymptomatic until March, 1973, when the suboccipital headache and cervical pain returned. He was admitted to another hospital and during his work up was found to have a mass involving the C-2 vertebra.

An attempted removal was done by way of a posterior cervical laminectomy. Only partial removal was accomplished and the final pathological report was chondrosarcoma. He received no postoperative radiation therapy. He was told that he had a malignancy of the cervical spine and that his condition was terminal. After that time he experienced progressive dysphagia and was admitted to our hospital on October 23, 1974, because of a mass appearing at the angle of the jaw.

Examination. On admission to our hospital, the patient was found to have no physical abnormalities other than a firm, immovable, nontender mass in the region just posterior to the ramus of the mandible and anterior to the sternocleidomastoid muscle. The tumor mass could also be felt in the region of the left tonsillar fossa. Examination of the cranial nerves revealed no abnormalities and the rest of the neurological examination was entirely within normal limits.

Radiographic examination of the cervical spine showed a stippled calcification, outlined in Fig. 1. The majority of the tumor appeared

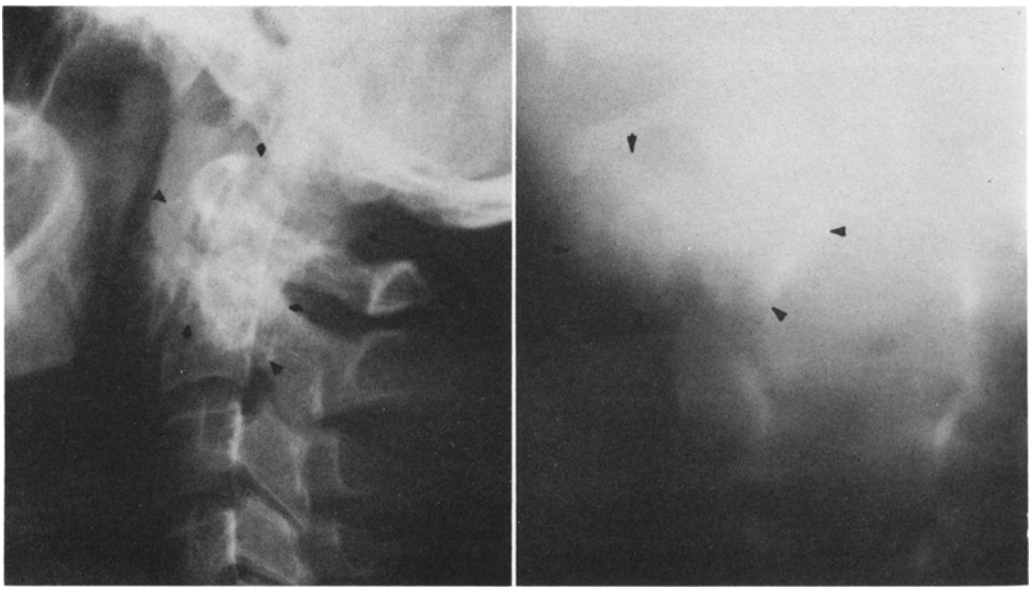

FIG. 1. *Left:* Plain film of the cervical spine, lateral view, showing stippled calcification of the tumor mass *(arrows). Right:* Anteroposterior tomogram demonstrates the extent of the tumor that appears to arise from the lateral mass of the axis.

to involve the lateral mass of C-2 and was osteoblastic in appearance. Vertebral arteriograms were done by means of femoral catheterization. These showed a very small rudimentary left and a normal right vertebral artery. The left carotid was seen to be displaced anteriorly by the tumor. There was no tumor blush seen on these arteriograms. A previous bone scan was reported to have shown increased osteoblastic activity in the upper cervical region.

Operation. At surgery the patient was placed in the right lateral decubitus position with the left mastoid process uppermost (Fig. 2). The skin incision is made approximately 3 cm above the mastoid tip and 1 cm behind the ear. The incision was then carried inferiorly over and just anterior to the sternocleidomastoid muscle. The periosteum was then removed from the mastoid process and the tip of the mastoid process was rongeured away. The sternocleidomastoid muscle was then divided approximately 1 cm from its insertion, and reflected inferiorly, whereupon the tumor mass came into view. The carotid sheath and the 9th, 10th, and 11th nerves were displaced anteriorly. The tumor capsule was smooth and firmly attached to the lateral mass of C-2. There was no evidence of inva-

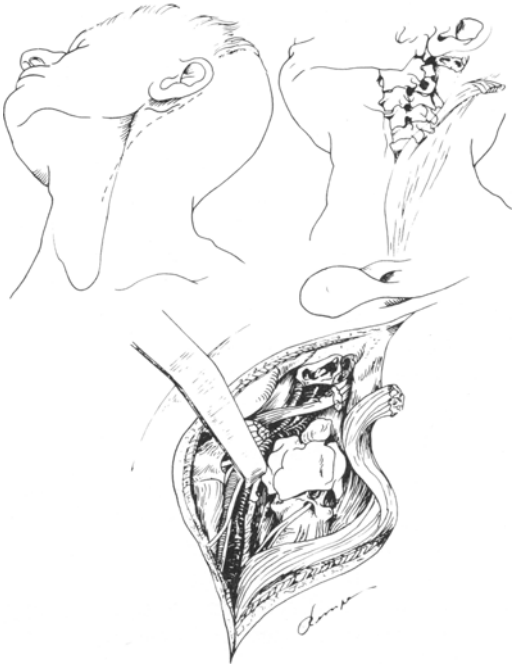

FIG. 2. Diagrams showing operative exposure of the tumor. *Upper Left:* Placement of skin incision. *Upper Right:* The sternocleidomastoid muscle has been divided and reflected inferiorly, exposing the lateral aspect of the upper cervical spine. *Lower:* The carotid sheath and lower cranial nerves have been retracted anteriorly, exposing the tumor mass.

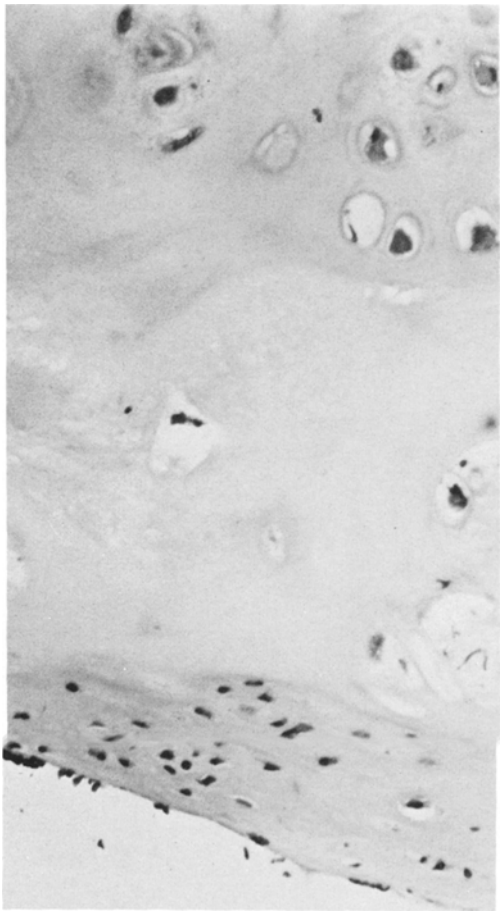

FIG. 3. Photomicrograph of tumor section, showing numerous cartilagenous cells with a moderate amount of matrix. H & E, × 300.

sion of surrounding structures. The capsule was then incised and by subcapsular piecemeal dissection the bulk of the tumor mass was removed. The vertebral artery was never seen during the dissection. As we neared the facet joint space, the tumor was more firm and this was removed by neuroaerotome drill until what appeared to be normal bone was identified. At this time the entire capsule was apparently removed. Pathological diagnosis from the surgical material was well-differentiated chondrosarcoma (Fig. 3).

Postoperative Course. The patient made an uneventful recovery. Neurological examination revealed no deficits and all cranial nerves were functioning. Postoperative films of the cervical spine demonstrated good stability of the cervical spine and no evidence of residual tumor. He was discharged on the 7th postoperative day with the wound well healed. No postoperative radiation was given. Correspondence with the patient 1 year postoperatively found the patient free of further symptoms and without evidence of recurrence.

Discussion

Chondrosarcoma represents only 7.6% of all primary malignant bone tumors, but half of those reported to be successfully treated.[3] Therefore, total removal of these tumors should always be attempted.

Chondrosarcomas arising from the spinal column are extremely rare. Törmä,[8] in his report based on 250 histologically verified malignant tumors of the spine and spinal extradural space, found chondrosarcoma to have occurred only 11 times. Coley.[2] did not report a single chondrosarcoma in his series of osteogenic tumors of which 283 were osteogenic sarcomas and of these only two were of the spinal column itself.

Chondrosarcomas have been found at every level of the spinal column, most commonly in the thoracic spine.[4,8] This tumor occurs as a primary malignant tumor in the earlier age groups but as a secondary malignant change in such conditions as osteochondroma, Paget's disease, or diaphysial achalasia. The bones most often affected are the pelvis, ribs, sternum, and femurs.[5] Dahlin and Henderson[3] report the closer the tumor is to the axial skeleton and the larger it is, the more likely it is to be malignant. Most feel that these tumors may arise from previously existing benign cartilaginous lesions, such as chondroma, osteochondroma, multiple exostoses, or Ollier's disease. Coley[2] feels that malignant changes may occur over an interval of 18 months to 30 years, while Seiforth[6] reports an interval of 3 to 5 months. In multiple cartilaginous exostoses or chondrodysplasia, the chondrosarcoma arises from the cap of the exostoses. If these cases are followed for a long enough period, a good number will be found to develop chondrosarcomas.

Radiographically, there is often frank destruction of trabecular bone and cortex with an expanding lesion which contains irregular flecking or mottling of the calcified tissue. Histologically it appears as an

atypical, more cellular enchondroma with islands of mature hyaline cartilaginous matrix interspersed with other areas where the cartilage is poorly developed and contains atypical anaplastic cells.

Chondrosarcomas have been classified by Thomson and Turner-Warwick[7] into three types:

Type 1 is a low-grade, well-differentiated tumor containing increased numbers of cartilaginous cells with well-formed matrices. Approximately 75% of patients with this type were alive 10 years after treatment.

Type 2 is an average-grade tumor, with less matrix and increased cellularity. The cells vary in size and shape with nuclear irregularities. Less than 50% of patients with this type were alive at 5 years after treatment, and only one-third were alive at 10 years.

Type 3 is a high-grade tumor, with a poorly differentiated picture. Anaplastic cells are common, mitoses frequent, and only an occasional island of cartilage is seen. Its behavior is similar to that of the osteosarcoma. Of this group, only one of 10 patients survived 3 years.

Metastasis tends to occur late in the disease and takes place by way of large veins.[1] When metastasis does occur it may spread over long distances, especially to the lungs.

References

1. Ackerman LV, del Regato JA: **Cancer. Diagnosis, Prognosis, and Treatment, ed 3.** St. Louis: CV Mosby, 1962, p 1136
2. Coley BL: **Neoplasms of Bone, ed 2.** New York: Paul Hoeber, 1960
3. Dahlin DC, Henderson ED: Chondrosarcoma: a surgical and pathological problem. Review of 212 cases. **J Bone Joint Surg 38A**:1025–1058, 1956
4. Robbins SL: **Pathology, ed 3.** Philadelphia: WB Saunders, 1967, pp 1348–1349
5. Schwartz SI: **Principles of Surgery.** New York: McGraw-Hill, 1969, pp 1649–1650
6. Seiforth B: Primäre Wirbelsäulentumoren Klinische und röntgenologische Differential Diagnose. Inaugural dissertation, Koln, 1964
7. Thomson AD, Turner-Warwick RT: Skeletal sarcomata and giant-cell tumor. **J Bone Joint Surg 37B**:266–303, 1955
8. Törmä T. Malignant tumors of the spine and the spinal extradural space. A study based on 250 histologically verified cases. **Acta Chir Scand (Suppl) 225**:1–176, 1957

Address reprint requests to: Russell L. Blaylock, M.D., Division of Neurological Surgery, Medical University of South Carolina, 80 Barre Street, Charleston, South Carolina 28401.