Metastatic chordoma to the mandible


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The chordoma is a slowly growing spinal cord tumor that rarely metastasizes. This is the first report in the literature of a chordoma with metastasis to the mandible. The mandibular involvement occurred 4 years after the primary sacral chordoma was controlled by excision and radiation. The jaw lesion did not respond to radiation but has been controlled by resection.

The chordoma is a rare spinal cord tumor that originates from remnants of the primitive fetal notochord. It grows slowly but eventually is fatal. Chordomas usually arise in the line of the craniovertebral, vertebral, and sacrococcygeal groups. Chordomas are considered locally aggressive tumors that rarely metastasize. Chambers and Schwinn and Markwalder and associates reviewed the literature from the years 1907 to 1978 for metastatic chordomas and were not able to cite one case metastatic to the jaw.

The purpose of this report is to review the pathophysiology of the malignant chordoma and present the first case in the literature with metastasis to the mandible.

CASE REPORT

In 1976 a primary sacral chordoma was diagnosed in a 64-year-old Filipino woman. The patient underwent resection of the tumor, followed by postoperative irradiation to the area. She remained clinically free of disease for 4 years. In September, 1979, the patient was presented to the head and neck tumor conference at the University of California San Francisco Medical Center with a growth in the region of the right mandible. A 5.0 by 6.0 cm. firm, nontender, fixed mass was felt overlying the mandible deep to the right parotid gland. No lymph nodes were palpated, and there was no evidence of any other tumor elsewhere. The patient felt well. Radiographic survey, including a CT scan, showed a 3.5 by 2.0 cm. bone-destructive lesion of the right mandibular angle with poorly defined margins (Fig. 1). A biopsy of the tumor was consistent with a metastasis of the sacral chordoma, revealing similar histologic features: a lobular arrangement of tumor cells in a vacuolated mucinous matrix (Fig. 2, A), sheets and syncytia of polygonal tumor cells as well as larger cells with vacuolated cytoplasm and scattered typical physaliferous cells,
and occasional mitotic figures and cells with irregular, hyperchromatic nuclei (Fig. 2, B). The patient underwent tumor debulking and superficial parotidectomy, followed by postoperative irradiation to the region of the right posterior mandible.

In January, 1981, the patient had some renewed pain in the right mandible, which was effectively relieved with Tylenol and codeine. On examination in February, 1981, there was some expansion of the ascending right ramus of the mandible. The Panorex film taken at that time showed a well-circumscribed 2 by 2 cm. radiolucency in the right angle of the mandible. It was recommended that the patient be followed closely, since curative procedures were not deemed practical.
In July, 1981, a CT scan of the head and neck and sacral areas confirmed a right mandibular lesion; the sacral region showed postsurgical and postradiation changes without evidence of tumor. It was recommended again that the patient be given palliative treatment only.

In November, 1981, the patient returned to the Oral Medicine Clinic because of increased intraoral pain and numbness of the right lower lip as well as definite enlargement of the right mandibular mass. On intraoral examination, there was a 2 by 2.5 cm. rubbery, firm mass fixed to the ascending ramus of the mandible (Fig. 3). The mass was tender and interfered with mandibular occlusion. The pain was moderate. A CT scan revealed progressive disease in the right mandible. The sacral area remained clear.

In December, 1981, the patient underwent a right hemimandibulectomy because of severe pain and dysfunction. The histopathologic examination of the tissues removed proved the mass to be a persistent chordoma. Since the resection the patient has been asymptomatic.

DISCUSSION

The notochord is a column of cells of mesodermal origin formed along the central dorsal axis, situated in front of the neural tube. This embryonic cord extends from the buccopharyngeal membrane to the coccyx and provides a support to the developing body of the embryo. In vertebrates, the notochord undergoes regression by the sixth week of fetal life and its function is assumed by the vertebrae and intervertebral discs.

The vertebræ and discs are formed in mesoderm which develops around the notochord. Although the notochord disappears, remnants remain between the vertebral bodies, forming the nucleus pulposus of the intervertebral discs. These remnants, which may persist anywhere along the axial skeleton, especially in the areas of the head and the tail folds where there is much developmental activity, give rise to the chordoma.

The typical chordoma is a lobulated, partially translucent mucoid tumor. The microscopic picture shows a lobular arrangement of polygonal tumor cells in a mucinous matrix. Tumor cells may contain numerous mucus-filled cytoplasmic and nuclear vacuoles that displace the nucleus, producing the typical "physaliferous" cells. Sometimes the cytoplasm forms a syncytium in which the nuclei are scattered. Extracellular mucus is nearly always present. Binkhorst and colleagues state that the histologic signs of malignant chordoma are definitive aplasia, mitotic figures, and dark-staining, irregular nuclei. The chordoma in our patient was characterized histologically by irregular hyperchromatic nuclei and occasional mitotic figures.

Regarding origin of chordoma in the cranio cervical region, Binkhorst and co-authors described one group as an ectopic localization in the mandible and frontal sinus. However, their conclusion about this origin was based on a report made in 1948 by Adams, whose conclusions about localization of chordoma in the mandible were made from a literature search in which he found one case in the lower jaw. In his report, however, Adams did not give any reference to support the existence of that one case of jawbone chordoma.

Small reported a case of a soft mass located in the tail of the parotid gland. This mass was diagnosed as a malignant chordoma. The diagnosis was made from an aspiration biopsy. No information was given regarding bone involvement.

Regardless of location, the classic radiologic finding of chordoma is that of an osteolytic lesion with a soft-tissue mass accompanying the bony lesion. In the sacrococcygeal zone the tumor grows slowly, with mild symptoms that are not realized for years before the final diagnosis is made. The tumor destroys the sacrum by an osteolytic process, and when the mass reaches the spine, pressure gives rise to pain and paresthesia. Primary chordomas of the nasopharyngeal area have been reported.

The question arises whether our case is that of a metastasis of chordoma from the sacrococcygeal zone to the mandible or a case of a second primary chordoma arising in the cranial end of the notochord. It must be recalled that the chordoma is a rare, slowly growing neoplasm that is locally invasive and rarely metastasizes. Only about 10 percent of all chordomas metastasize, with the sacrococcygeal lesions metastasizing more frequently than the craniocervical and vertebral areas. It has been reported that radiation treatment raises the risk of metastasis. It seems likely that our case repre-
sents a metastasis rather than a second primary neoplasm arising in the same patient.

REFERENCES