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Chordoma of the Atlas

A CASE REPORT

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Benign and malignant tumors of the atlas (the first cervical vertebra) are so rare that a computer search of the world medical literature revealed only two reported cases, a giant-cell tumor³ and a myeloma¹.

We have encountered a chordoma of the atlanto-axial region of the cervical spine, as reported herein. Besides the rarity of such a case, the roentgenographic manifestation — subtotal lysis of the anterior elements of the first and second cervical vertebrae — is noteworthy.

Case Report

A sixty-three-year-old white woman had a history of deep pain in the neck, beginning after she had slipped on the ice and landed flat on her

pre-auricular regions, where there was also numbness.

Physical examination two months after injury revealed tenderness in the upper cervical region, some muscle spasm, and some decreased sensation in the first and second cervical dermatomes. Roentgenograms showed diffuse destruction of the lateral masses of the atlas and odontoid process of the axis (Fig. 1-A). The laboratory studies were all within normal limits.

A transoral biopsy of the lesion of the upper cervical spine was done under general anesthesia. The histological diagnosis was chordoma (Fig. 1-B). A week later, the patient had a spine fusion from the occiput to the fourth cervical vertebra under general anesthesia and she tolerated the procedure well. Radiotherapy was advised but was refused by the patient. The postoperative course was uneventful and a Minerva cast was applied prior to discharge.

The patient wore the Minerva cast for approximately six months,



FIG. 1-A

Roentgenogram of the cervical spine, showing destruction of the lateral masses of the atlas and the odontoid process of the axis.

back. She was seen by her family physician who told her that she had a "cracked bone" in the neck and she was treated with a cervical collar. Because of persistence of pain over a two-month period during which she had two periods of hospitalization for traction and physiotherapy, she was referred to us. The pain had spread over the left mandibular and

after which she wore a cervical collar. On follow-up slightly more than five years later, she was quite well and did not have any neural deficit. Follow-up roentgenograms showed almost complete destruction of the anterior arch and lateral masses of the atlas and odontoid process of the axis, but the fusion from the occiput to the fourth cervical vertebra appeared to be solid (Fig. 1-C). The patient died suddenly at home seven months later. The cause of death was unknown. No autopsy was performed.

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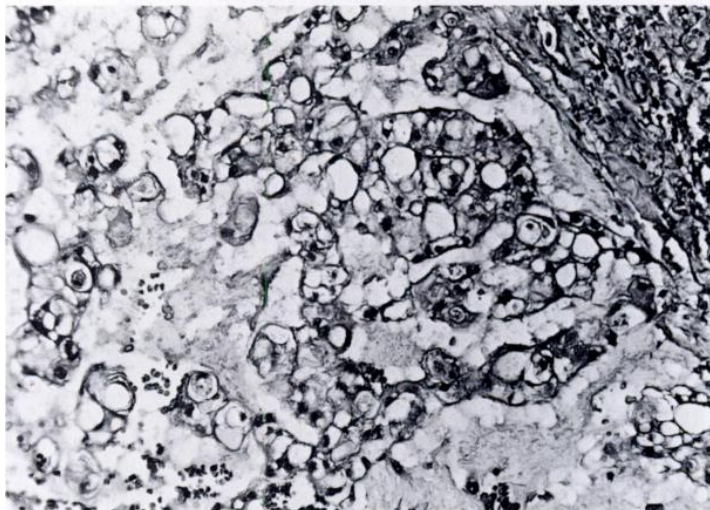


FIG. 1-B

Photomicrograph showing the lobulated pattern, intracellular and extracellular mucinous substance, eccentric and pyknotic nuclei, and bubble-like physaliferous cells of chordoma (hematoxylin and eosin, $\times 280$).



FIG. 1-C

Roentgenogram showing almost complete destruction of the anterior arch and lateral masses of the atlas and the odontoid process of the axis. Note the solid posterior fusion from the occiput to the fourth cervical vertebra.

Discussion

The tumor of the atlas reported by Hastings and associates, a myeloma, caused destruction of the left articular mass of the atlas and the patient became quadriplegic because of a subluxation. In our patient the nearly complete disappearance of the anterior supporting structures of the atlas and odontoid process did not cause a subluxation, but we think that the fusion from the occiput to the fourth cervical vertebra was indispensable for sustaining the patient's life during five years of follow-up.

We are not certain whether the chordoma originated in the atlanto-axial region or in the spheno-occipital area with secondary involvement of the atlas and axis. Nevertheless, it presented a challenging problem in diagnosis (a transoral biopsy) and therapy (occipitocervical fusion). The atlas by the Netherlands Committee on Bone Tumors mentioned a chordoma of the second cervical vertebra which apparently did not involve the atlas. No mention was made of the patient's clinical course.

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