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Cervical chordoma with retropharyngeal extension presenting with impaired voice

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ABSTRACT

Aims and background. We report an extremely rare case of cervical chordoma presenting with impaired voice.

Method. Case report and a review of the literature concerning the presentation, diagnosis, and treatment of a cervical chordoma.

Results. A singing teacher complaining of dysphonia was examined and surgically treated for a retropharyngeal extension of a cervical chordoma. A local recurrence was treated with proton beam therapy. Among primary malignant tumors of bone, chordomas account for 3-4% of all cases. Chordoma is typically a locally aggressive tumor with a high propensity for local recurrence. Its management involves surgery, radiotherapy, or both.

Conclusion. To our knowledge this is the first report in the world literature of a retropharyngeal extension of a cervical chordoma presenting with impaired voice. This case indicates that bony tumors of the spine may present first to voice-disorder clinicians. Increased awareness of this neoplasm may lead to earlier diagnosis and better treatment.

Introduction

We present a case of cervical chordoma presenting with dysphonia as the predominant symptom. The management and prognosis of these tumors is still poor but increased awareness may lead to earlier diagnosis with smaller lesions that can be better treated.

Case report

A 56-year-old singing teacher presented to us with a 6-month history of progressive dysphonia and intermittent dysphagia. Her past medical history was noncontributory with an asymptomatic mitral valve prolapse, tricuspid insufficiency, and a cesarean section. She denied cigarette and alcohol consumption. She consulted us because of a muffled voice that lacked projection and had modified resonance. Intermittent episodes of coughing during meals had occurred. Physical examination revealed a large, firm, immobile mass involving the right side of the retropharyngeal wall. The mass was at the level of the epiglottis and was covered with normal mucosa. The remaining head and neck exam was unremarkable, including vocal cord mobility and vibration. Neurological examination was also unremarkable.

Computed tomography revealed a well-demarcated soft tissue lesion located in the right prevertebral space and with adjacent osteolysis of C3. Axial imaging with a soft tissue window showed a prevertebral mass with a mass effect on the right posterior pharyngeal wall (Figure 1). The bone window setting showed areas of irregular sclerosis within the vertebral body of C3 and osteolysis involving the anterior and right

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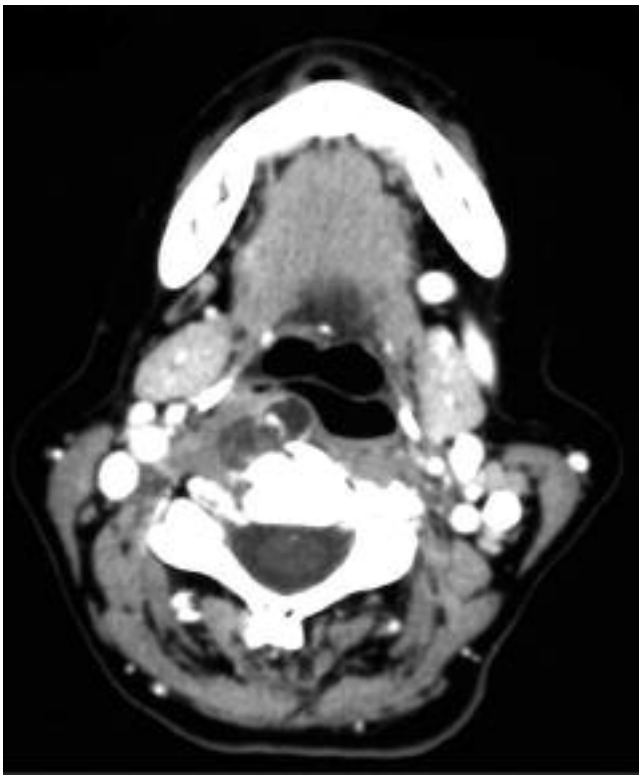

Figure 1 - Axial computed tomography scans of the neck. The soft tissue window shows a prevertebral mass (black arrow) with a mass effect on the right posterior pharyngeal wall.

lateral portion of C3. The mass deformed the posterior wall of the hypopharynx without displacing the larynx. Magnetic resonance imaging of the neck showed on the T2-weighted images a polypoid lesion with extremely high signal intensity including most of the vertebral body of C3 and with extension into the right prevertebral and retropharyngeal space (Figure 2A). After injection of gadolinium, inhomogeneous, bubble-like enhancement of the lesion was seen on the T1-weighted image (Figure 2B). The posterior extension was at the level of the right vertebral artery.

The patient underwent a biopsy procedure through a direct pharyngoscopy approach while the final surgical removal required a cervicotomy and the resection of the vertebral body of C3, followed by a reconstruction with an iliac crest allograft. Macroscopically, the specimen was composed of grey, elastic, myxoid tissue with patchy necrotic and hemorrhagic portions. Microscopic examination revealed tumor cells arranged in cords or nests with a vacuolated cytoplasm (physaliphorous cells) embedded in a myxoid intercellular matrix. The nuclei were round or oval, with a prominent nucleolus (Figure 3A, 3B). Immunohistochemically, the tumor cells expressed keratin, epithelial membrane antigen, S-100 protein but not CD-117 or actin. The MIB-1 labeling

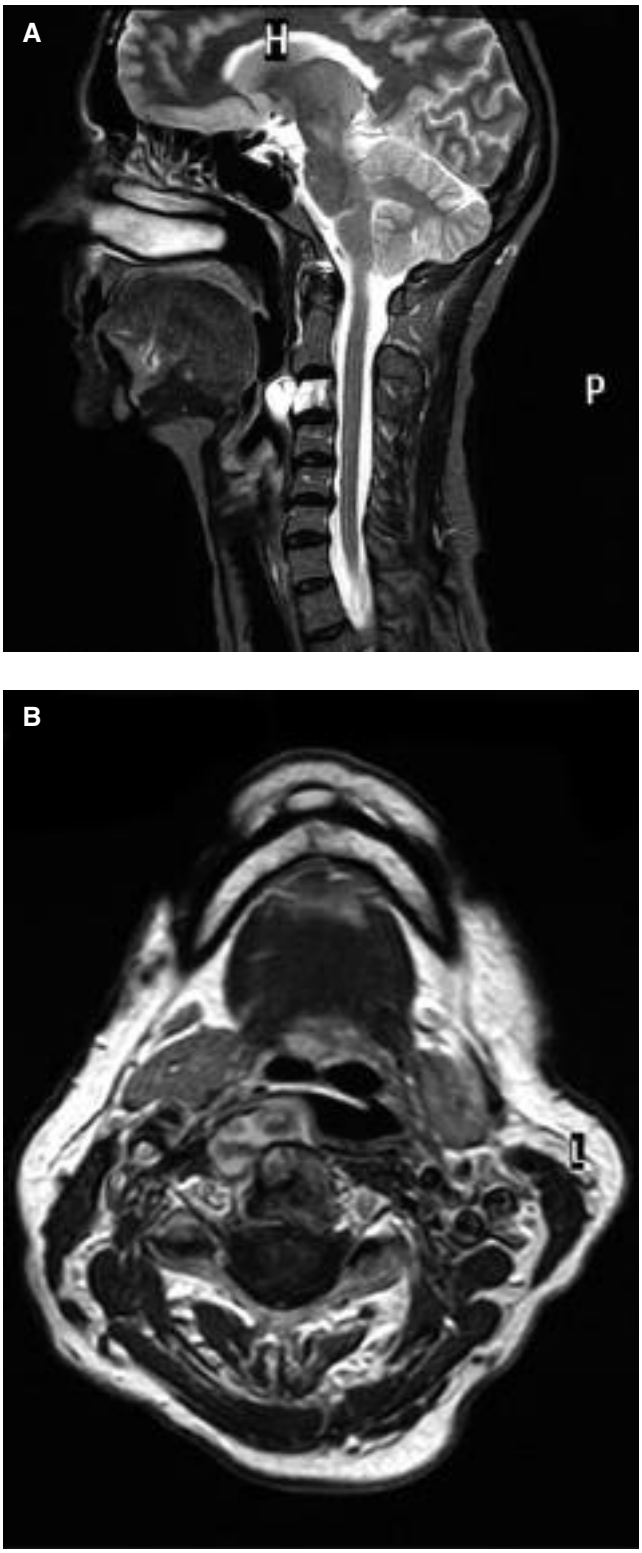

Figure 2 - Magnetic resonance imaging of the neck. A) T2-weighted images show a polypoid lesion with extremely high signal intensity (white arrow) including most of the vertebral body of C3 and with extension into the right prevertebral and retropharyngeal space. B) T1-weighted images with injection of gadolinium show inhomogeneous, bubble-like enhancement of the lesion (white arrow).

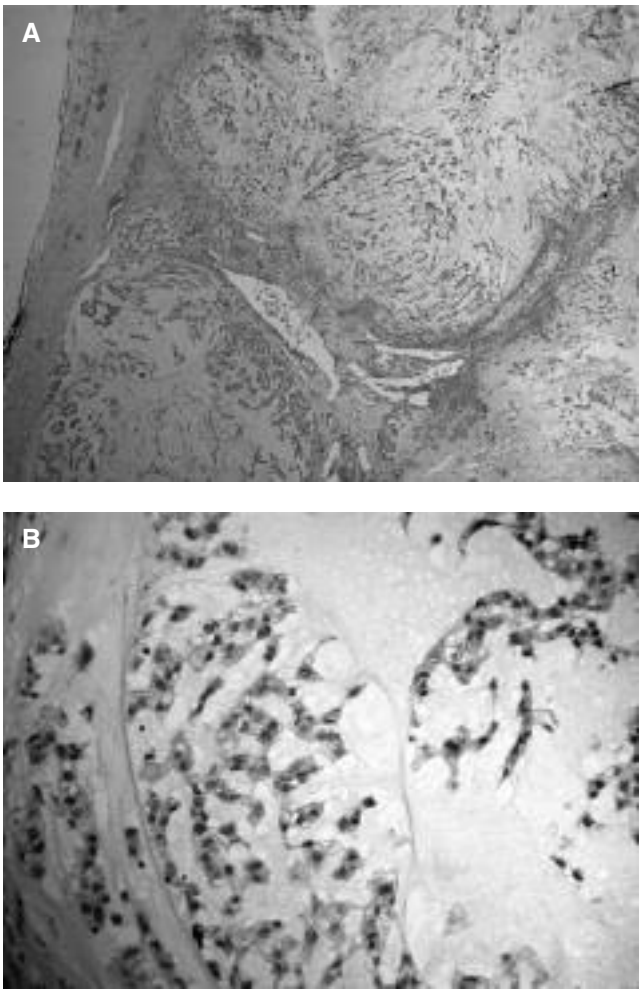

Figure 3 - A) Light microscopy of the tumor (H & E). B) Physaliphorous cells in chordoma (H & E).

index in the cellular area of the tumor was 10%. The final diagnosis was chordoma.

Apart from mild regressive dysphagia, the postoperative period was uneventful. One year after the procedure, the tumor recurred as a small nodule anterior to the axis body, which was treated with proton beam therapy.

Discussion

Among primary malignant tumors of bone, chordoma appears to be the fourth most frequent histological type (after osteosarcoma, chondrosarcoma and Ewing's sarcoma), accounting for 3% to 4% of all cases[1]. In the United States the overall age-adjusted incidence is 8 per 10 million, with a median age at diagnosis of 58.5 years (range, 3-95 years) and a generally progressive increase in incidence with age. The most common sites for the development of chordoma are the skull (32%), spine (33%) and sacrum (29%)[2]. Symptoms are related to tumor localization. Pain is a common symptom in chordoma and can precede the diagnosis by a few months to several years. Spinal chordomas frequently produce compression of the spinal cord and nerves. Although chordomas of the mobile spine have a poor prognosis and traditionally have been difficult to manage, earlier diagnosis of smaller lesions may have a significant impact on their management and prognosis.

Cervical chordomas tend to present as a pharyngeal or lateral neck mass. As the tumor expands, alteration in swallowing due to pharyngeal impingement and various degrees of paralysis of the extremities due to spinal cord and nerve compression appear. Atypical presentations such as dysphonia, as in our case, may be referred to a variety of specialists. It therefore seems important that clinicians be aware of this neoplasm in order to allow early diagnosis and effective treatment.

Chordomas were first reported by Luschka in 1856, prior to their histological description by Virchow[3], who introduced the term "ecchordosis physaliphora" to designate cells in which the central nucleus is surrounded by a large vacuolated cytoplasm. The presence of physaliphorous cells is a hallmark of chordoma.

More than one third of chordomas occur at the base of the skull and especially the clivus. The typical extension of these neoplasms is ventral, towards the nasopharynx and occasionally into the nasal cavity or maxillary antrum. Cervical spine chordomas are very rare and only few cases have been reported[4,5]. Symptoms vary with the site of the lesion and the degree of local extension. Upper cervical and cranial base lesions may cause visual disturbances (loss of acuity, diplopia, and limitation of visual field), headaches, pain, endocrinopathies, and a variety of nasal symptoms. Lower cervical lesions may be associated with dysphagia or dyspnea[4]. Spinal lesions may result in radicular pain and sensory motor problems[5]. Dysphonia related to a retropharyngeal mass as the first symptoms of chordoma has not been yet reported. This case illustrates that dysphonia may be a predominant symptom in the initial presentation of malignant tumors of the cervical spine.

CT images give information on bone invasion and destruction. MRI can detect epidural spread and can be used to evaluate encasement of the vertebral arteries before surgery[6]. The possibility of a chordoma should be considered when a destructive lesion of the cervical vertebral body is associated with a large well-defined soft-tissue mass that is extremely hyperintense on T2-weighted MR images and presents inhomogeneous enhancement after contrast material injection[6].

The presence of physaliphorous cells and the trabecular or cord-like arrangements with a myxoid matrix are typical for chordoma. In addition, immunohistochemical examination may be more reliable by showing positive staining for keratin, epithelial membrane antigen and S-100 protein, thereby excluding other tumors such

as chondrosarcoma, pleomorphic adenoma, or mucin-producing adenocarcinoma.

The current management of cervical spine chordoma involves surgery, radiotherapy, or both. Chemotherapy has been of little benefit in this tumor type. Surgery with complete tumor resection is considered the treatment of choice. However, resection of vertebral lesions may be less efficient than for sacral lesions as vertebral tumors are often more developed at the time of diagnosis. As the incidence of local recurrence and metastasis is correlated with the degree of tumor resection, extensive procedures often have to be undertaken.

Some authors reported significant improvement in recurrence-free survival with maximal surgical excision followed by proton beam radiation therapy or stereotactic radiosurgery[7-9]. Chordomas are considered to be radioresistant tumors and local control can only be achieved with high radiation doses, above 7000 cGy. To achieve this goal, proton beam radiation therapy offers the advantage of increasing the dose delivered to the tumor while minimizing radiation to surrounding normal tissues[8]. Stereotactic radiosurgery with the use of a linear accelerator or gamma knife is limited to cranial chordomas because the irregular and infiltrative nature of this tumor makes it difficult to target[9]. Fractionated stereotactic radiotherapy using intensity-modulated radiation therapy and micro-multileaf collimators may help optimize delivery of radiotherapy to tumors in the future. The small numbers of patients reported and the short follow-up make optimal treatment difficult to define. It seems reasonable to reserve external radiation for recurrences and inoperable tumors.

Chordoma is typically a locally aggressive tumor with a high propensity for local recurrence. The reported 5-year overall survival rates are 60% to 70%, but the 10-year survival drops to 35-40%[2]. Tumor recurrences occur typically in the form of multiple bone and soft tissue masses or subcutaneous nodules. Chordomas metastasize most often to lymph nodes, lung, bone, and liver. The rate of metastasis varies widely among series, with a higher incidence among chordomas of the mobile spine than sacral lesions. The presence of microscopic tumor necrosis and an MIB-1 labeling index of more than 5% are considered as poor prognostic factors[10].

References

1. Dorfman H: Chordomas and related lesions. In: Bone tumors, Dorfman H, Czerniak B (Eds), pp 974-1008, Mosby, St Louis, 1998.
2. McMaster ML, Goldstein AM, Bromley CM, Ishibe N, Parry DM: Chordoma: incidence and survival patterns in the United States, 1973-1995. Cancer Causes Control , 12: 1-11, 2001.
3. Virchow R: Untersuchungen über die Entwicklung des Schädelgrundes im gesunden und krankhaften Zustande und über den Einfluss derselben auf Schädelform, Gesichtsbildung und Gehirnbau, p 128, G. Reimer, Berlin, 1857.
4. Hester TO, Valentino J, Strottmann JM, Blades DA, Robinson MC: Cervicothoracic chordoma presenting as progressive dyspnea and dysphagia. Otolaryngol Head Neck Surg, 120: 97-100, 1999.
5. Zacay G, Eyal A, Shacked I, Hadani M, Faibel M, Kronenberg J, Talmi YP: Chordoma of the cervical spine. Ann Otol Rhinol Laryngol, 109: 438-440, 2000.
6. Wippold II FJ, Koeller KK, Smirniotopoulos JG: Clinical and imaging features of cervical chordoma. AJR Am J Roentgenol, 172: 1423-1426, 1999.
7. Colli B, Al-Mefty O: Chordomas of the craniocervical junction: follow-up review and prognostic factors. J Neurosurg, 95: 933-943, 2001.
8. Hug EB, Loredo LN, Slater JD, DeVries A, Grove RI, Schaefer RA, Rosenberg AE, Slater JM: Proton radiation therapy for chordomas and chondrosarcomas of the skull base. J Neurosurg, 91: 432-439, 1999.
9. Debus J, Schulz-Ertner D, Schad L, Essig M, Rhein B, Thillmann CO, Wannenmacher M: Stereotactic fractionated radiotherapy for chordomas and chondrosarcomas of the skull base. Int J Radiat Oncol Biol Phys, 47: 591-596, 2000.
10. Bergh P, Kindblom LG, Gunterberg B, Remotti F, Ryd W, Meis-Kindblom JM: Prognostic factors in chordoma of the sacrum and mobile spine. A study of 39 patients. Cancer, 88: 2122-2134, 2000.