

Chordoma of the Cervical Spine in a Competition Athlete: Case Report and Long-term Follow Up

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Abstract

Introduction: Chordoma is a rare type of low-grade malignant neoplasm that arises from the remnants of the embryonic notochord. Observed mainly in the clivus and sacrum but can occur anywhere along the spine. Several treatment approaches are described. Treatment outcomes are significantly influenced by the size and location of the tumor.

Clinical presentation and follow up: We report a 19-year-old female professional athlete with a cervical chordoma, involving C2, C3 and C4 vertebrae with spinal cord compression. Diagnosis was established by open biopsy. The patient was surgically treated in three steps: one anterior resection of the lesion was carried out, followed by a posterior resection and finally an arthrodesis and anterior fixation. The patient was referred to rehabilitation and one year after the first surgery she resumed competitive sport activities. No recurrences were observed within fifteen years of follow-up.

Keywords: Chordoma; Cervical spine; Tumor

Abbreviations: CT: Computerized Tomography; MRI: Magnetic Resonance Imaging; US: Ultrasonography

Introduction

Chordoma is a malignant tumor that usually involves the axial skeleton, and its origin is related to notochord remnants. It is a slow-growing, locally invasive and aggressive tumor. It is observed mainly in the sacrum and in the clivus but also may appear along any region of the spine [1]. The diagnosis of this tumor is usually late, due to the slow growth and its location [2]. Various treatment approaches have been attempted, including radical excision, radiotherapy and chemotherapy. Treatment outcome is significantly influenced by the size and location of the chordoma [3]. Complete tumor resection is considered very difficult because its proximity to the spinal cord and a high rate of local recurrence is usually observed [2,4].

In this study it is reported a case of a female professional athlete with cervical chordoma involving C2, C3 and C4 vertebrae that have been undergoing surgical resection.

Case Report

In 1999, a 19-year-old, female, professional athlete underwent to a surgical procedure to remove her tonsils. During the procedure, an increased volume in the lower right side of the oral cavity was observed. A cervical spine CT scan was performed prior spine surgeon evaluation. The physical exam showed swelling in the right cervical region, distal to the lower mandible angle and anterior to the sternocleidomastoid muscle. No pain on palpation and a mass adherence to deep layers was noted. The neurological exam was normal. Radiographs of the cervical spine showed no bone changes. Both CT as MRI demonstrated a rounded extra-osseous lesion penetrating the spinal canal by the foramen of C2-C3 and C3-C4 causing cord compression. It was also found that the tumor encapsulated the right vertebral artery (Figure 1). Bone scintigraphy revealed absence of local changes or signs of metastasis. The patient was instructed stopping sport activities immediately. A biopsy guided by ultrasound followed by confirmation by a transoral open biopsy allowed the diagnosis of chordoma. No sign of metastasis was identified in the staging of the chest and abdomen. Arteriography has demonstrated that the tumor was poorly vascularized

and the contralateral vertebral artery (left side) was the prevailing. Embolization of the right vertebral artery was carried out early in two weeks prior to tumor resection. Firstly the tumor was approached anterolaterally, resecting the extra-osseous lesion, and removing tumoral fragments from within the vertebral canal and opening C2-C3 e C3-C4 foramina. The extra-osseous lesion had a good cleavage plan, which was not observed in the bone structure (Figure 2). Fixation was not required in this procedure since there was no instability. After that a CT scan was performed for postoperative control purpose and tumor was still observed in the right posterolateral region. Thus, a posterior approach was carried out three months after the first procedure and C3 lamina and C2-C3 and C3-C4 facet joints were resected only on the right side, preserving ligaments, muscular, articular and bone structures on the left side. The athlete had only a decrease in tactile sensitivity in the C3 root area on the right side. The patient was referred to rehabilitation to recover physical conditioning and therefore return to her sports activities. Dynamic radiographs were taken during rehabilitation and identified a listhesis of C2. There were no signs of tumor relapse on CT and MRI. Due to the instability and in order to allow the return to sports practice other procedure was performed for fixing vertebrae C2, C3 and C4, in December 2000 (Figure 3). The patient was referred to physical therapy and recommended to continue using cervical orthosis for 3 months. In March 2001 the patient was allowed to gradually resume physical activities. In May 2001, one year after the first procedure, MRI showed no signs of local tumor relapse. The athlete returned unrestricted to competitive sports practice one year after the first procedure and 15 years of follow-up showed no recurrence and a mild degeneration of adjacent levels (Figure 4).

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Figure 1: A) Radiography of the cervical spine shows no bone changes. B,C) CT and MRI showing a roundish extra-osseous lesion penetrating the spinal canal through the C2-C3 and C3-C4 foramen causing cord compression. Tumor was encapsulating the right vertebral artery.

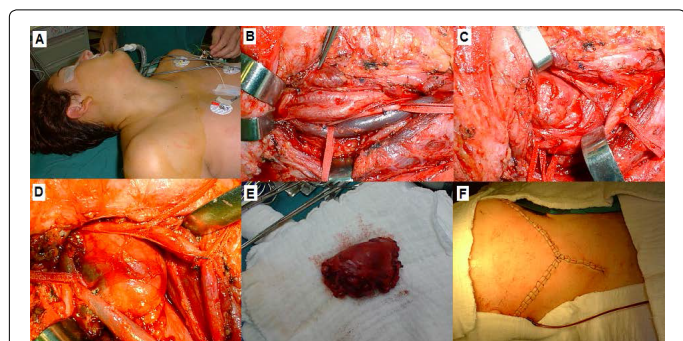


Figure 2: Anterior approach for chordoma resection. (A) Patient positioning. (B) Isolating neurovascular structures (Carotid Bulb, Jugular Vein, Vagus and Hypoglossal nerves) (C,D) Tumor exposure (E) Resected mass (F) Final suture.

Discussion

Chordoma represents 1-4% of primary bone tumors. Patient age at detection is between 30 and 70 years; they are found more often in men than women (ratio 1,6:1). Most Chordomas affect sacrococcygeal region (~50%) and the base of the skull (~35%), being uncommon in the mobile segments of the spine (~15%) [1,3,5-7]. Cervical spine chordomas represent approximately 5% of all chordomas, they are usually large and cause the destruction of multiple vertebrae invading soft perivertebral tissues and breaking into the epidural space causing pain and weakness in the neck and shoulders [8,9]. In the case reported above, the aspect of the tumor was different. A cystic and well-defined tumor was observed with minor bone involvement causing no pain. It is similar to the description of the variant chondroid chordoma, which was originally reported by Heffelfinger et al. in 1973, having a better prognosis than conventional chordomas [7,10]. This injury combines aspects of a chordoma in association with the cartilaginous tissue producing a hyaline matrix [10,11].

The reported metastatic rate of chordomas ranges from 10% to 48%. Survival rates at 5 and 10-years are approximately 50 and 35%, respectively [9,12]. Several factors can influence the prognosis and recurrence rate; the chondroid type is more often observed in patients

younger than 40 years, which appears to be a significant factor of better prognosis. The size and location of the tumor at first presentation is also important in prognosis, since radical resection at this time, is either associated with lower relapse rates, and tumors involving neurovascular structures may preclude margin-free en bloc resection [12].

The diagnosis was initially suggested due to the increased swelling of the neck. Radiography did not contribute for the diagnosis, however, CT and MRI allowed the identification of tumor extension, its lobulation and presence in the vertebral canal causing cord compression. This picture has been reported in literature [3,8,10,13,14], but there are no pathognomonic images for this kind of injury. Arteriography allowed the assessment of tumor vascularization assisting with surgical planning, as shown in literature [13-15], since the tumor encapsulated the vertebral artery on the right side, which was embolized before tackling the tumor [15]. The diagnosis and therapy used depended on the evaluation of the fragments removed during biopsy.

The surgical technique was chosen according to the localization of the tumor [16,17] and the need of stabilization due to the instability caused by bone resection. It was not possible to carry out en bloc resection with a safety margin according to the technique described by Fujita et al. [18], since the lesion was extra-compartmental, involving neurovascular structures [2]. After tumor resection, stabilization and arthrodesis of the approached segments are important procedures [5,19,20]. Titanium plate and screws were used allowing the performance of magnetic resonance to control relapses in the postoperative period. A synthetic device of bone matrix was used in the disc spaces. Radiotherapy was not used in the postoperative period. Some authors have reported beneficial effects of using this treatment [4,16,20], however Fagundes et al. [21] concluded that local control of the tumor alone prevents recurrences.

In our case an aggressive anterior approach was performed for tumor resection in first surgery, followed by a posterior approach after three months to remove the remaining tumor. This treatment added to embolization of right vertebral artery (that might be giving some nutrition to the tumor), was enough to eliminate the chordoma without radiotherapy, and no relapses were observed over fifteen years of follow up.

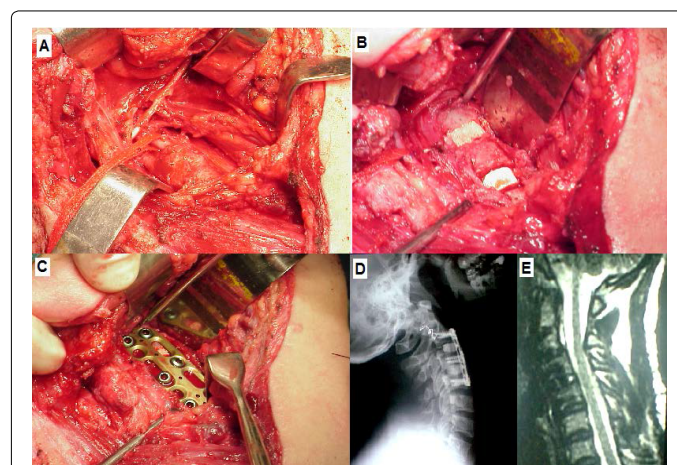
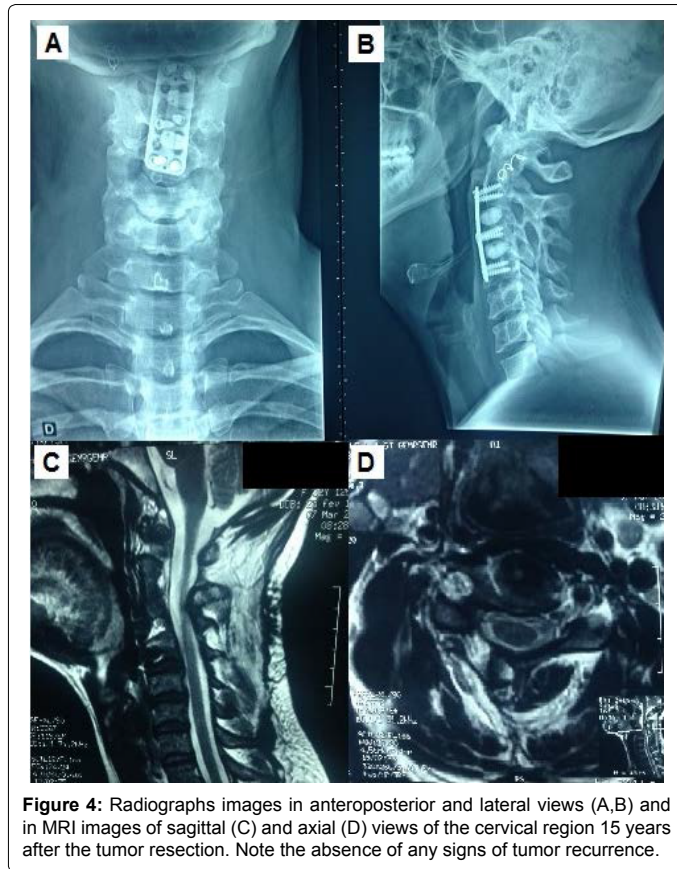


Figure 3: Third procedure demonstrating the anterior aspect of the neck in the second approach (A) and the arthrodesis with cages (B) and after the plate (C). Note no signs of recurrence (A-C), Sagittal Radiography (D) and MRI (E) after C2-C4 fixation.



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