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PAGES: 192-197

ORIGINAL PDF URL: <https://dergipark.org.tr/tr/download/article-file/2156415>

## Neck mass as a consequence of surgical implantation of cervical chordoma: A case report

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### ABSTRACT

Chordoma is a rare primary malignant tumor of notochord origin along the craniospinal axis. Extranotochordal chordoma is extremely rare. These slow-growing but highly destructive tumors have worrisome local recurrence rates following successful primary treatment. Symptoms and findings are due to localization of the lesion. The primary treatment option is surgery because of weak chemotherapy and radiotherapy response. The size of surgical resection in these tumors has a major effect in ensuring disease-free surveillance. Recurrence throughout the surgical site is a rare form of treatment failure. A case of cervical chordoma diagnosis and chordoma recurrence on follow-up is presented in this study because of its rare clinical course.

**Keywords:** Chordoma; implantation; neck; surgery.

Chordomas are rare, slow-growing, locally invasive tumors originating from ectopic notochordal residues along the axial skeleton.<sup>[1]</sup> They originate from sacral (29.2%), cranial (32%) and spinal (32.8%) regions.<sup>[2]</sup> Even if they are histologically benign, chordoids are highly invasive and locally destructive. Chordomas rarely occur in soft tissue and are extremely rare in the field of head and neck surgery.<sup>[3]</sup> Only a few cases of soft tissue chordoma have been reported in the head and neck zone, which includes the lateral nasal wall, paranasal sinuses, oropharynx, parapharyngeal space and soft tissues.<sup>[4,5]</sup> Symptoms and signs are due to effects of the lesion on surrounding tissues.<sup>[6,7]</sup> Complete tumor removal on initial surgery is important to ensure good prognosis, since it makes the

probability of recurrence almost impossible.<sup>[7]</sup> However, optimal surgical access and complete resection is often restricted by proximity of the tumor to vital organs.<sup>[8,9]</sup> Even with adjuvant radiotherapy (RT), the local recurrence rate is approximately 29% of the basilar cord and occurs in 95% of all treatment failures.<sup>[10]</sup> Recurrence at the surgical entry site is a rare form of treatment failure. We present a rare case of cervical chordoma.

### CASE REPORTS

A 59-year-old male patient presented with asymptomatic right neck mass. The patient underwent transcervical C5 corpectomy + C5-7 stabilization with adjuvant RT for a cervical chordoma at a different medical center four

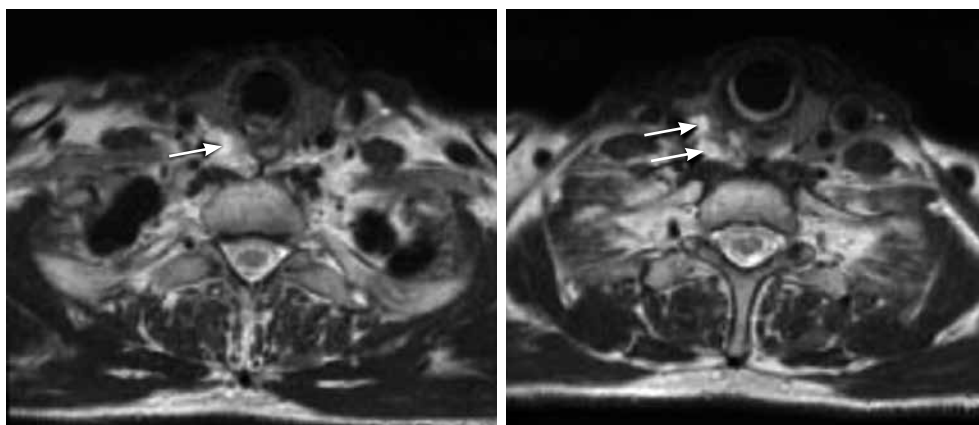
Received: November 04, 2018 Accepted: January 05, 2019 Published online: February 12, 2019

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**Doi:** <http://dx.doi.org/10.5606/Tr-ENT.2018.40085>

### Citation:

Kabaş Uysal Z, Tatar B, Etit D, Kumbul YÇ. Neck mass as a consequence of surgical implantation of cervical chordoma: A case report. Tr-ENT 2018;28(4):192-197.

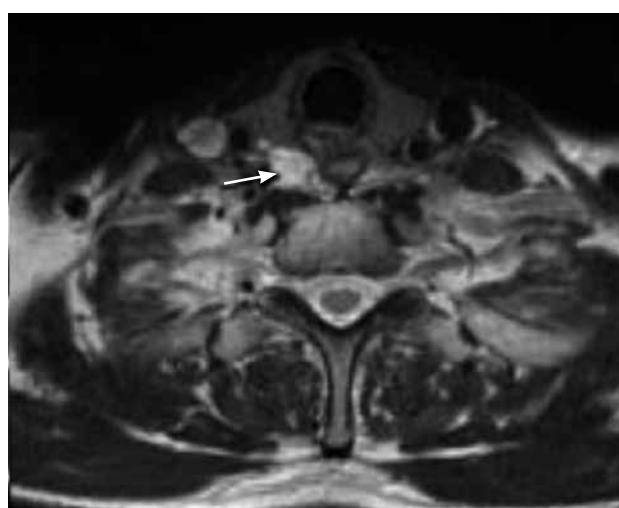


**Figure 1.** T<sub>2</sub> weighted axial magnetic resonance imaging images at the level of C5-C6 showing hyperintense lesion (arrows), four months before our surgery.

years ago. On examination of patient who was referred to us because of the mass detected on follow-up, there was a mobile mass of approximately 2.5×1.5 cm in the posterolateral side of the right thyroid lobe and its vicinity. A fine needle aspiration biopsy (FNAB) report was compatible with chordoma. Metachromatic myxoid fibrillary material showed multinucleated, microvacuolized tumor cells that stained positive with cytokeratin and EMA and negative with S-100 and GFAP. Fine needle aspiration biopsy examination performed under the right lobe of the thyroid was reported as mass with cystic content. The patient's neck magnetic resonance imaging (MRI) revealed a 2.5×1.5 cm lesion with a lobulated contour on the lateral aspect of the right thyroid lobe and an 8 mm sized lesion on the lateral side as a satellite (Figure 1). The tumor, which grew conspicuously in comparison to the MRI eight months earlier, had spread up to the prevertebral area (Figure 2). The patient was evaluated in a multidisciplinary tumor panel. As a result of the mass biopsy, the impression was chordoma recurrence after surgical site implantation.

A right thyroid lobectomy incision was performed. In order to preserve the right recurrent laryngeal nerve, it was decided to reach the mass from the right side of the thyroid lobe and from the medial to the carotid sheath, and right thyroid lobectomy was performed. When the trachea and esophagus were flipped to the left, a cystic mass of approximately

3 cm was reached in the anterior of the cervical vertebra and in the medial of the carotid sheath. The mass did not cause any destruction of surrounding tissues, and it was removed along with its capsule (Figure 3a-e). Histopathology confirmed the presence of chordoma that stained positive with S100, keratin AE1/AE, keratin 8/18, keratin 7 (Figure 4a-c). Histopathological examination of the thyroid lobe was benign. The surgical margins were negative. The patient had no complaints after the operation and was discharged and referred for RT. He was healthy



**Figure 2.** T<sub>2</sub> weighted axial magnetic resonance imaging images at the level between vertebrae C5-6 showing that the tumor (arrow) and its borderline with the prevertebral area, one year before surgery.



**Figure 3.** Intraoperative photographs showing: (a) localization of tumor between the carotid sheath and the right lobe of thyroid, (b) view of tumor after lobectomy, (c) view of part of tumor adjacent to the carotid sheath. Notice the localization of tumor; (d) removal of the tumor, and (e) view of surgical site after tumor removal.

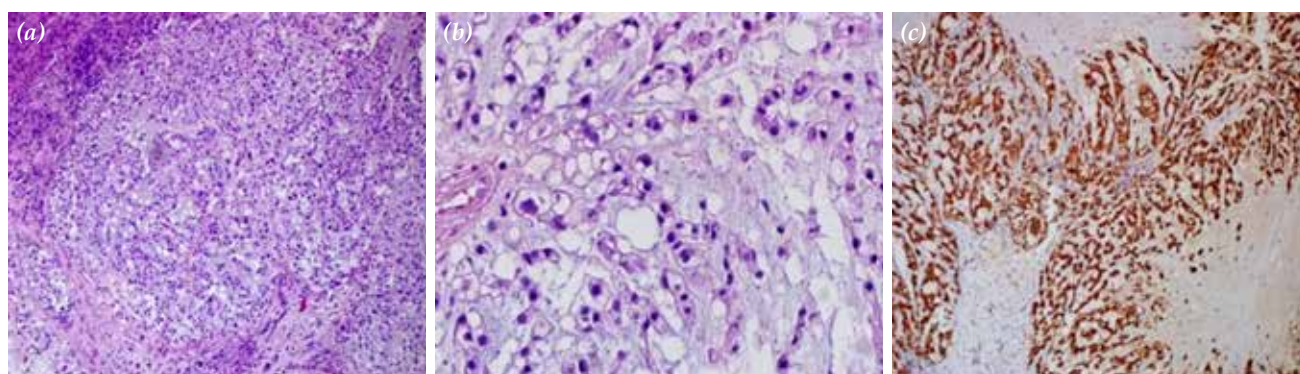
on follow-up and showed no signs of recurrence or residual on six-month postoperative MRI images (Figure 5).

### DISCUSSION

The major differential diagnosis of our case included distant metastasis, regional lymph

node metastasis, and previous surgical implant recurrence.

According to the 2017 ESMO Chordoma Global Consensus Group, local recurrence is defined as the spread of tumor to the adjacent areas of primary site and/or tumor recurrence or progression of primary tumor at the same



**Figure 4.** (a) Histopathological images of physaliferous cells with the large vacuolated cytoplasm and mucinous stroma (H-E×100), (b) pan keratin positive, (c) S-100 positive (H-E×200).





**Figure 5.** T<sub>2</sub> weighted axial magnetic resonance imaging images at the level of C5-6 showing that no residue tumor at the sixth month after operation.

site after RT.<sup>[11]</sup> This includes the progression or recurrence of treated lesions; lesions that develop iatrogenic insemination along the biopsy or surgical path, near the germ limits; and metastases in the near vicinity.<sup>[11]</sup> In most cases, tumor spread is through direct physical contact rather than through lymphatic, hematogenous or subarachnoid routes.

Long-term recurrence-free survival (RFS) of mobile spinal chordomas 5 and 10 years after optimal local treatment has been reported at 58% and 32%, respectively.<sup>[12]</sup> Primary determinants of local control of primary chordomas in all zones include tumor size, resection width, quality of surgery, and quality of RT and patient age.<sup>[13]</sup> Treatment center experience may also play a role in the likelihood of recurrence.

Fischbein et al.<sup>[9]</sup> suggested that four criteria must be met in order to apply the term surgical site recurrence: (i) disease along the surgical access pathway for original tumor resection; (ii) presence of a normal intervening tissue of at least 2.5 cm between disease and location of original tumor; (iii) recurrence at least within 24 months after surgery; (iv) no evidence of perineural, lymphatic or distant metastases at the time of diagnosis.

All these conditions have been met in our patient. A rare number of chordoma cases

with longitudinal surgical implantation using the above criteria have been reported for surgical pathway recurrence.<sup>[10]</sup> Boyette et al.<sup>[10]</sup> reported two local recurrences and recurrent clival chordomas that occurred as a neck mass with surgical implantation after performing more than one resection. Measures have been proposed to prevent surgical implantation, including replacing instruments, gloves, and contaminated towels and drapes before closing, as well as filling with surgically tunneled fibrin glue.<sup>[14]</sup> However, further research is needed to determine the effectiveness of filling the surgical tunnel as a preventive tool. The radiation zone should be considered to include the surgical path.<sup>[14]</sup> Radiologic follow-up evaluation should be recommended to facilitate early detection of surgical path recurrence. Chordomas are isointense or hypointense to brain on T<sub>1</sub>-weighted MRI.<sup>[15]</sup> Final diagnosis requires tissue biopsy but remarkable T<sub>2</sub> hyperintensity on MRI would be reduced recurrence. If a soft tissue lesion is detected along the surgical pathway, the differential diagnosis should consider body reaction to operative materials, postoperative granulation tissue formation and surgical path recurrence. Surgical pathway recurrence is a rare condition requiring a high index of suspicion, and control is best performed by surgical intervention and postoperative radiation. Although the surgical pathway as in this case can be controlled locally, there is often a poor prognosis, mainly because of primary or distant failure.<sup>[15]</sup> The current treatment approach is wide local excision of the tumor followed by high dose RT. The main goal of this treatment modality is curative palliation due to high recurrence rates and the survival rate is 5-10 years.<sup>[16]</sup>

Chordoma Consensus Group recommended treatment plan for local recurrence chordoma in 2017 ESMO: total resection with negative surgical border should be performed.<sup>[11]</sup> Previously digested with resection or other high dose RT (mobile backbone) are expressed in significant exclusion criteria for curative treatment for re-resection of the tumor rupture. Primary treatment with high-dose preoperative RT, may also be suitable for patients were not taken before and/or post-surgical treatment.<sup>[17]</sup>

This plan is standard therapy plan for some referral centers in prime disease<sup>[18]</sup> and after failed prior surgical procedures in the event of an actual R<sup>0</sup> resection of local recurrence treated as a chance to do is possible particularly suitable.

The complex anatomy of the head and neck region complicates the extensive excision of malignancies. We recommend a clinical and radiological examination of lymph nodes in all soft tissue tracts to remove metastasis that may potentially affect initial tissue planes. Moreover, due to the rare occurrence, complex diagnosis and follow-up of these lesions, it is recommended that such cases be routinely monitored at a specialized oncology center. This case suggests that despite the rarity of surgical site recurrence, a high index of suspicion is required and should be considered in the differential diagnosis of a patient with a history of a chordoma resection that has been referred more than two years after initial treatment.

In conclusion, surgery is still the basis of treatment for these tumors. Published case series indicate that local recurrence and reporting surgical outcomes for chordoma affects 50% of patients who undergo macroscopic complete resection with or without RT.<sup>[11]</sup> Although various surgical techniques are available depending on the location of the lesion and the surgical resection site is very important for ensuring disease-free period, it is appropriate to discuss the risks (especially local recurrence) that may develop during the postoperative period and plan a surgical approach accordingly. Therefore, defining best evidence-based practices for optimal management of the patient during initial treatment is crucial for improving patient outcomes.

#### Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

#### Funding

The authors received no financial support for the research and/or authorship of this article.

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