Case Report

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A case report of Sustained triumph: 15-year recurrence-free survival following surgical resection of a cervical chordoma in a 15-year-old girl

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Introduction and importance: Chordoma of the cervical spine is a rare condition associated with poor long-term outcomes. This is mainly attributed to its pathological involvement of vital structures such as the cervical roots and vertebral artery (VA). Although the most appropriate management in these cases is total en bloc excision, attaining complete resection is relatively challenging due to the vicinity of the critical anatomical structures mentioned above.

Case description: A 15-year-old female with middle cervical spine chordoma was treated by a multidisciplinary team involving neurosurgery and head and neck surgeons utilizing anterior and posterior approaches followed by high-beam X-ray radiotherapy. Histopathological examination matched the description of a chordoma. Fifteen years after the initial excision, the patient maintained her normal neurological function without local recurrence or metastasis.

Clinical discussion: The patient underwent surgery in two stages. A posterior approach for C3, C4, and C5 laminectomies was performed in the first stage, with the second stage involving head and neck surgery for complete resection of the tumor. The patient also underwent radiotherapy 3 months after surgery for a total duration of 1 month. The patient is currently 30 years old with no evidence of chordoma recurrence.

Conclusion: Patients afflicted with cervical chordomas often find themselves undergoing multiple operations due to high recurrence rates. Fortunately, the utilization of en bloc resection coupled with adjuvant radiotherapy presents a hopeful treatment modality that can serve to substantially reduce recurrence rates, increase survival rates, and ultimately enhance the quality of life.

Keywords: case report, chordoma, recurrence free

Introduction

Our case presentation demonstrates a unique presentation involving the mobile spine in a pediatric patient, which constitutes a minority of typical chordoma presentations[1–3]. In addition, the case demonstrated a lack of recurrence for a substantial period of time despite the hurdles faced with executing the optimal mode of treatment by en block excision of the tumor that is followed by radiotherapy[4]. Such cases are scarcely reported in the literature.

Here, we report the case of a 15-year-old female patient with C3, C4, and C5 vertebral chordomas with tumor intrusion into the neural canal and foramina transversarium, compressing the spinal cord and engulfing the VA. The patient's tumor was excised at Jordan University Hospital, a tertiary university hospital and further treatment was done using radiotherapy. She had correction by fusion of a swan neck deformity 2 years after surgery. Fifteen years after surgery, the patient is neurologically intact with no evidence of tumor recurrence. This case report has been written to be in line with the SCARE criteria[5].

HIGHLIGHTS

- A unique case of 15-year recurrence free of cervical chordoma post-en bloc resection radiotherapy.
- Cervical chordomas are rare neoplasms with poor survival rates following treatment.
- The most generally accepted treatment of chordoma is total en bloc spondylectomy with a wide margin with postoperative radiotherapy.
- Total en bloc spondylectomy of chordoma has the potential to significantly improve survival rates when it is possible to perform.

Case presentation

Patient information

A 15-year-old Jordanian high-school female patient with a free past medical and family history of neurological diseases presented

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to the outpatient clinic complaining of neck pain radiating to the right shoulder of 1-month duration with no relief by non-steroidal anti-inflammatory inhibitors. Otherwise, the patient's drug history is clear. Two weeks prior to her visit, she had right-sided weakness and electrical sensation on neck flexion and extension. The patient has full insurance covering all diagnostic and therapeutic procedures provided by the neurosurgery department at Jordan University Hospital.

Clinical findings

On physical examination, there was right shoulder abduction and elbow flexion weakness graded as 1-3/5 and right lower limb weakness of 4/5 on the Medical Research Council (MRC) scale of muscle power accompanied with a high steppage gait. Her reflexes were exaggerated on the right side of her body, and a right Babinski sign was noted.

Computed tomography (CT) of the cervical spine revealed a widening of the right foramen transversarium of C5 (Fig. 1).

Cervical spine magnetic resonance imaging (MRI) demonstrated an extradural extraspinal tumor, most likely a chordoma, with an intraspinal extradural component in addition to an extraspinal component in a dumbbell fashion (Figs 2,3).

Magnetic resonance angiography (MRA) of the neck vessels revealed compression of the right vertebral artery (VA) and increased vascularity in C3/C4 vertebrae from both VAs. Conventional angiography also demonstrated right VA concentric narrowing at the C3/C4 level corresponding to the circumferential encasement of the right VA, with a slight displacement of the right VA anteriorly and laterally (Fig. 4). A balloon occlusion test was performed, but no deficits were detected.

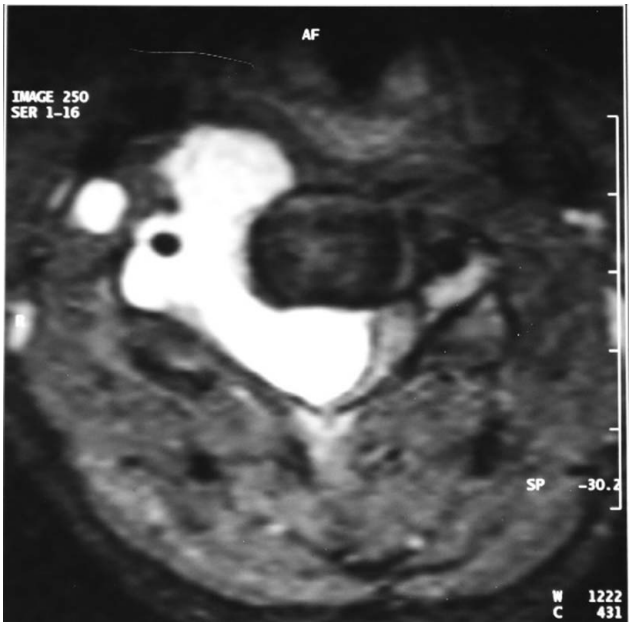

Figure 1. Computed tomography image of an axial view of the C5 vertebra showing the widening of the foramen transversarium.

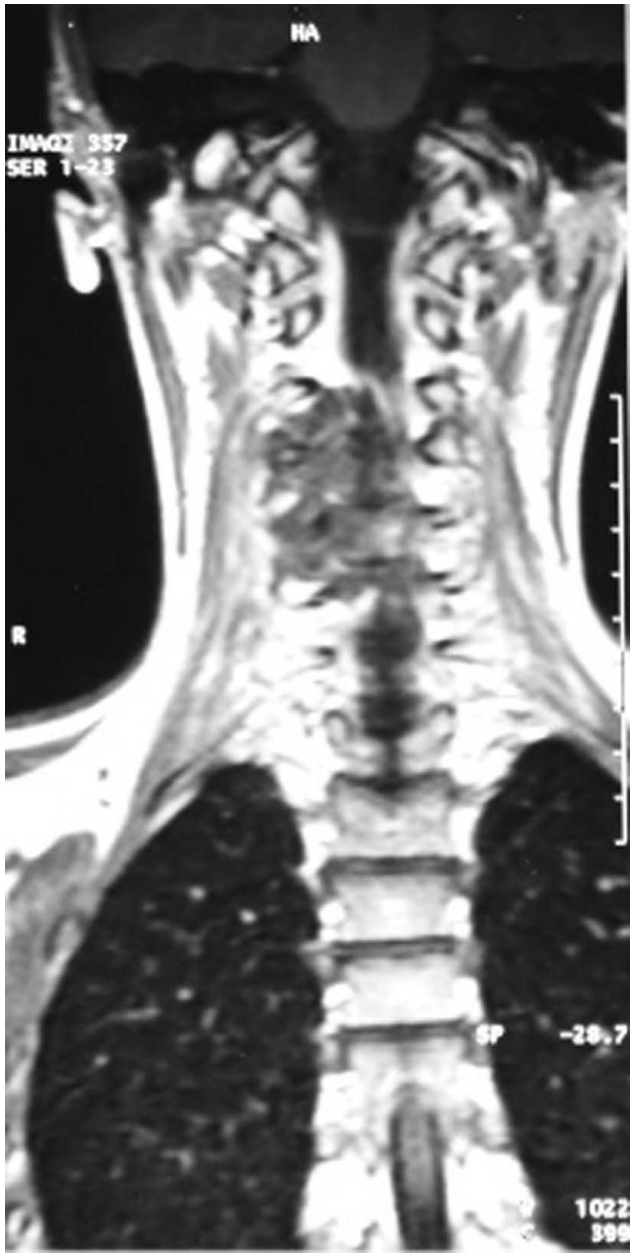

Figure 2. Coronal view of T1 magnetic resonance imaging of the cervical vertebrae showing a homogenous mass protruding from C3/C4/C5 vertebrae.

Follow-up and outcome

The patient was treated with surgical resection in two stages; the first was conducted in a sitting position. A posterior approach for C3, C4, and C5 laminectomies was performed for the excision of the extradural part, removing the intraspinal portion as far as could be seen in the intervertebral foramen. The tumor was soft and amenable to suction.

The second stage, performed 2 weeks later, was carried out by a head and neck surgical team using a right anterior neck incision in the supine position. The thyrocervical trunk was identified and ligated, and dissection of the tumor from the VA was achieved with complete removal of the extraspinal part in one block.

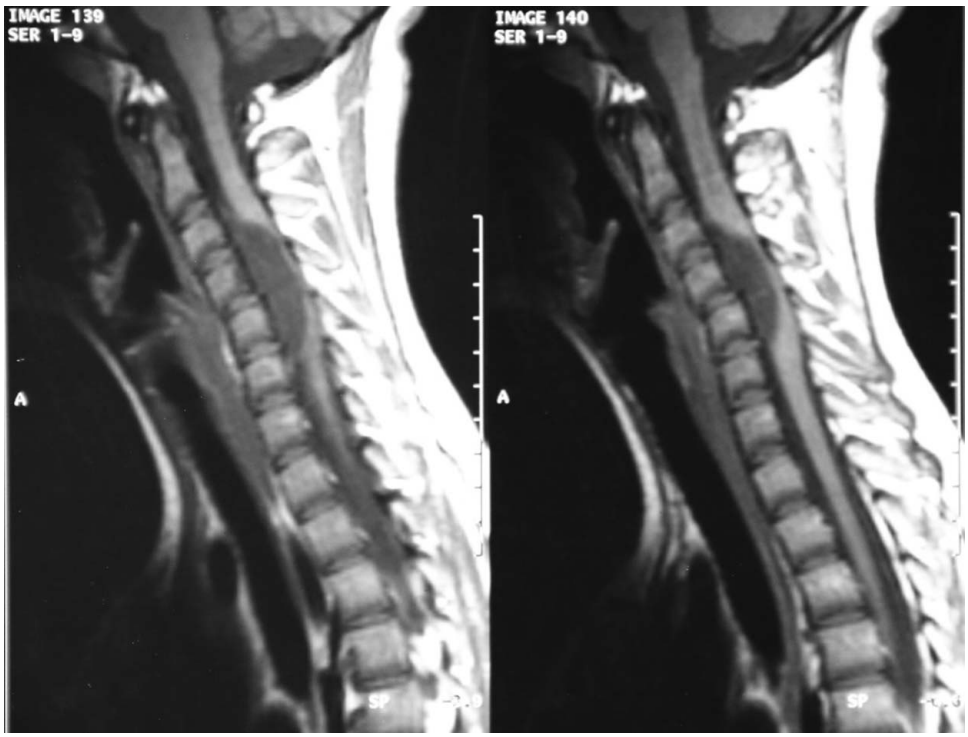

Figure 3. Sagittal view of T1 magnetic resonance imaging of the cervical vertebrae showing the cervical chordoma involving C3/C4/C5 vertebrae.

Immediately during the postoperative period, the patient developed Horner's syndrome. The power became 4/5 in the previously affected limbs. The pathological examination confirmed the diagnosis of chordoma, showing physaliphorous cells in myxoid background, with vacuolated bubbly cytoplasm and mild nuclear atypia, all of which are indicative of a conventional chordoma. Immunohistochemical analysis revealed that the tumor cells were strongly positive for cytokeratin, positive for S100 protein, and epithelial membrane antigen with the nuclear protein Ki 67 <2% (Fig. 5).

Three months after surgery the patient underwent radiotherapy. A total dose of 50.4 Gray (Gy) in 28 fractions over

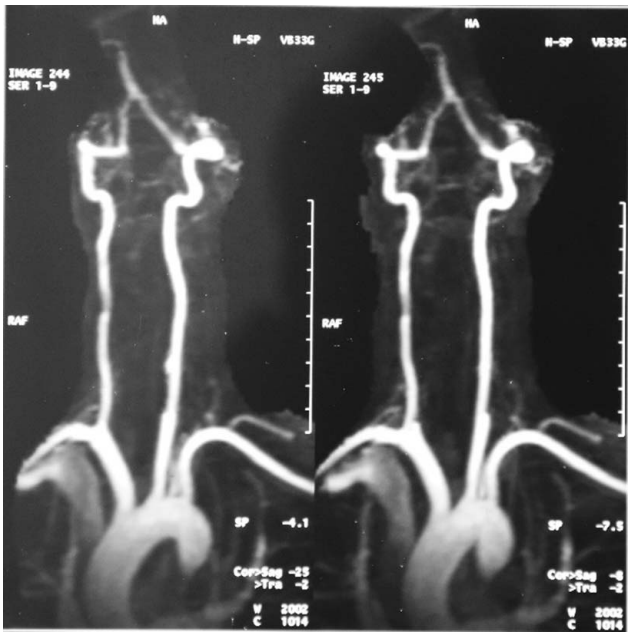

Figure 4. Magnetic resonance angiography image with a pointed arrow at the site of defective circulation within the vertebral artery at the level of C3/C4 cervical spine.

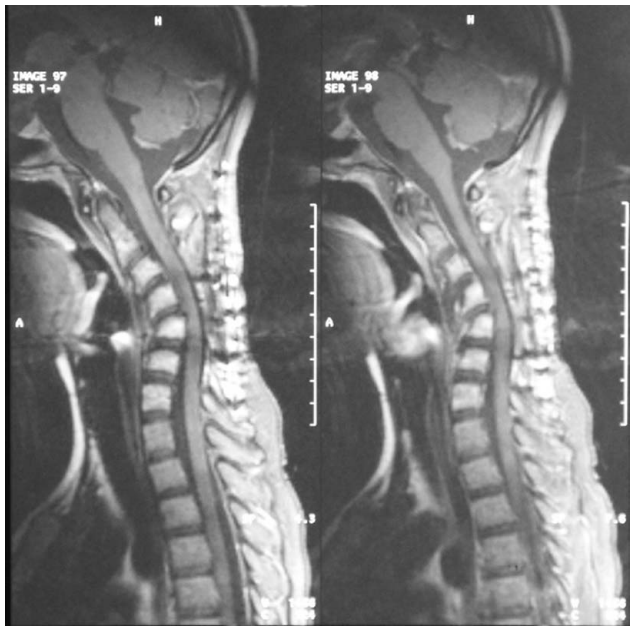

Figure 5. Postoperative sagittal view of magnetic resonance imaging of the cervical vertebrae following en bloc resection of the cervical chordoma.

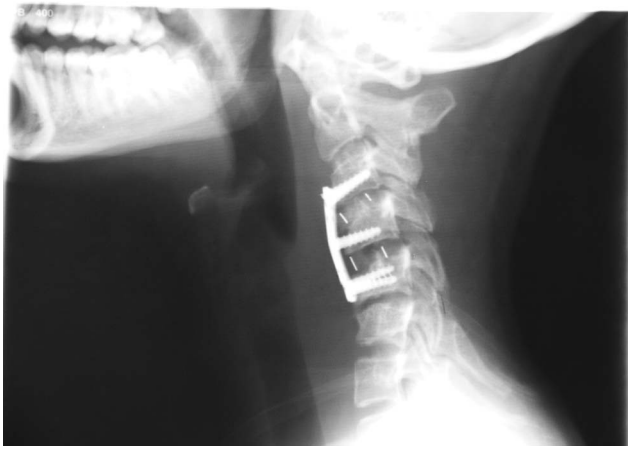

Figure 6. X-ray sagittal view of the cervical vertebrae following anterior cervical discectomy and fusion at the level of C3–C4/C4–C5 and C5–C6.

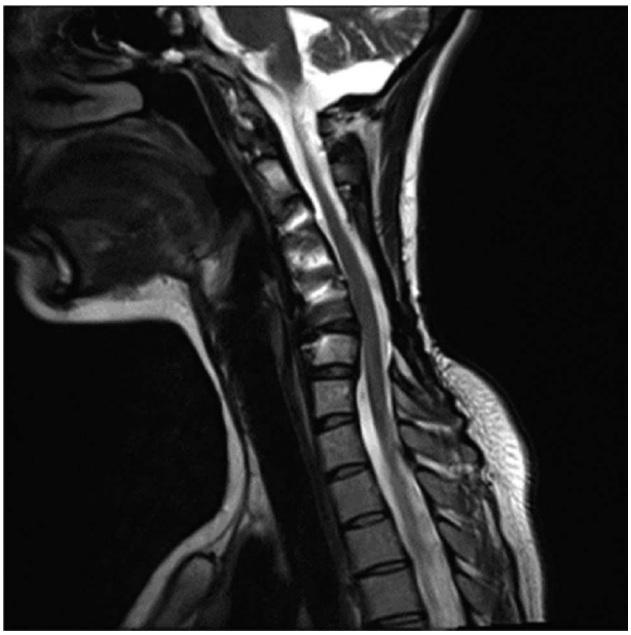

Figure 7. Sagittal view of cervical spine magnetic resonance imaging with contrast 20 years following en bloc resection of cervical chordoma.

1 month was delivered to a field between C2 and C7 via two opposing directions, right and left lateral. The patient tolerated the radiotherapy course well without any complaints or complications.

The patient was followed up closely and showed complete recovery of neurological deficits 6 months following surgery. However, 24 months following surgery, she complained of neck pain, and examination revealed loss of the cervical lordosis. Plain cervical X-rays and spinal CT showed a swan neck deformity, which was treated with anterior cervical discectomy and fusion of the level C3–C4/C4–C5 and C5–C6 (Fig. 6).

Repeated MRI examinations were performed every year and then every 2 years without any evidence of recurrence. When seen last month, 15 years after her first presentation, she was well

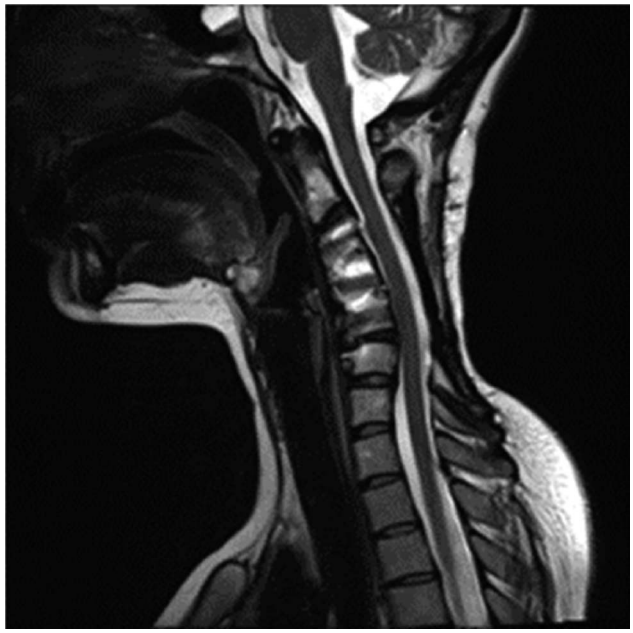

Figure 8. Sagittal view of cervical spine magnetic resonance imaging with contrast in 2023.

without any deficits, had married, and was inquiring about the practicality of raising a family (Figs 7,8).

Discussion

Chordomas are tumors of notochordal origin with genes involved in the expression of the transcription factor Brachyury[6,7]. The most common sites of tumor development include the sacrococcygeal region (50–60%), sphenoid-occipital region (25–35%), and mobile spine (15%)[1,2]. Lesions of the cervical spine are rare, comprising between 3 and 7% of all chordomas and between 20 and 50% of spinal chordomas[4,8].

In an analysis of pediatric and adult chordoma patients, the age-adjusted incidence rate was 0.08 per 100 000 population with male preponderance (male:female ratio of 1.7:1)[3]. The median age of diagnosis was 58.5 years, and less than 5% of chordomas are diagnosed in the pediatric population[3,9].

Due to their anatomic and pathologic diversity, management varies on an individual basis and is influenced by the treating center's multidisciplinary expertise, involving the capability of performing surgery by specialized surgeons, the availability of radiotherapy, and molecular therapy following molecular sequencing[7,10].

The mainstay of treatment involves maximal safe surgical resection, followed by radiation therapy, preferably with proton beams[3]. Conventional chemotherapy is of limited value, and intensive research now focuses on molecularly targeted therapies against specific genes, proteins, and signaling pathways[6].

Historically, complete surgical resection was and still is the most common option for the treatment of chordomas, mainly due to the poor response achieved by either chemotherapy or radiotherapy when used solely to prevent tumor recurrence[11]. In addition, there are no randomized controlled trials that concluded with finding the optimal form of treatment for chordomas[12,13].

Surgical intervention stands as an effective and reliable treatment modality for patients afflicted with spinal chordoma[9,14]. Tomita *et al*. described an innovative surgical technique termed total en bloc spondylectomy for malignant vertebral tumors in the thoracolumbar spine[15–17].

Total en bloc spondylectomy with a wide margin is generally agreed upon to be the most appropriate modality of treatment with clean margins and postoperative radiation therapy, providing the best long-term survival[18]. However, due to the vital structures involved in cervical chordomas, the technique may still prove to be challenging[18]. Refined high-dose photon radiotherapy following tumor resection is effective in the long-term control of chordoma and low-grade chondrosarcoma[18].

Although the aforementioned technique is highly efficacious, it remains a difficult operation with inherent challenges. Chordomas of the cervical spine usually involve vital structures such as the vertebral arteries. Many authors argue that the aggressive surgical approach is limited by the risk of vascular and neurological compromise of the neurovascular structures[10,19–22]. VA sacrifice may predispose to strokes, particularly if the tumor involves a dominant VA[19] and spinal cord ischemia due to variability of the radiculomedullary branches[23].

So, several authors have recommended cerebral angiograms and performing temporary balloon occlusion tests as part of the preoperative workup in patients with cervical chordoma to determine the feasibility of vessel sacrifice to achieve en bloc resection[10].

Regarding post-resection instability, a study conducted in Seoul, South Korea investigated the optimal timing of fusion surgery of clival chordoma with occipital–cervical joint (OCJ) instability. The study ultimately advocated occipital–cervical fusion before resection of clival chordoma with OCJ involvement[24].

Prognostic factors in patients with cervical chordoma were reviewed in multiple studies in the literature. Zhou *et al*.[25], a study of 52 cases from a single institution with a mean follow-up of 50 months (median 31 months) and range of 1–228 months, found that contiguous vertebral segment involvement, intralesional surgical margin, lack of treatment with adjuvant radiotherapy, and incisional biopsy significantly increase local recurrence. While tumor location in the upper cervical spine significantly increases tumor-related mortality[25], the CT-guided fine needle aspiration biopsy and total spondylectomy with marginal excision may improve the survival of patients with cervical chordoma[25].

Despite the modest survival rates described in the literature, our case had a substantial period of no recurrence, which highlights the importance of performing en bloc resection of cervical chordomas in addition to the need for further studies looking into circumstances surrounding such patients with high survival rates.

Conclusion

In conclusion, patients afflicted with cervical chordomas often find themselves undergoing multiple operations due to the high recurrence rates. Fortunately, the utilization of en bloc resection coupled with adjuvant radiotherapy presents as a hopeful treatment modality that can serve to substantially reduce recurrence rates, increase survival rates, and ultimately enhance the quality of life[26].

Ethical approval

Ethical approval was obtained from the Institutional Review Board of Jordan University Hospital under the reference number 10/2023/25/21. Departmental approval was obtained under the reference number 4762/2023/67.

Consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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This study did not receive any funding support.

Author contribution

M.H.: investigation and writing – original draft; L.R.: writing – review and editing, validation, and visualization; M.H.: writing – original draft and resources; M.A.-M.: writing – review and editing, and validation; W.M.: conceptualization, writing – original draft, visualization, and validation.

Conflicts of interest disclosure

There are no conflicts of interest to declare.

Research registration unique identifying number (UIN)

Not relevant.

Guarantor

Walid Maani.

Data availability statement

Not relevant.

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