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Case Report and Review of the Literature

A Primary Dumbbell-Shaped Cervical Extraskelatal Ewing's Sarcoma in an Adult: A Case Report and Literature Review

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ABSTRACT

Dumbbell spinal tumors get their hourglass or dumbbell shape from being constricted in the middle, with one part inside the spinal canal and the other extending outside of it.

This is the case of a 37-year-old female who presented to the clinic with a left cervicobrachialgia and acroparesthesia. While radiographic imaging initially reported a well-defined dumbbell-shaped C7-T1 foraminal tumor consistent with a neurinoma, the pathology report obtained after surgical intervention came back positive for Ewing's sarcoma, with no other locations of the tumor. The diagnosis was unexpected given the shape of the tissue and the rarity of the extraskelatal form of Ewing's sarcoma (EES). Only 4 previous cases to date have identified a primary cervical dumbbell-shaped extraskelatal Ewing's sarcoma in adults.

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Introduction

Ewing's sarcoma is nowadays a well-known malignant entity, classified as a primitive neuroectodermal tumor (PNET) [1]. While the most common form, the osseous Ewing's sarcoma (OES), arises in 85% from the diaphysis of long bones of the lower extremities primarily in children and adolescents in the first two decades of life, the quite rare extraskelatal or extraosseous Ewing's sarcoma (EES) can develop in around 15% of cases in soft tissues outside the bones, with cases reported in the esophagus, the larynx, the chest wall, the kidney, the head and neck region, the pelvis, the peritoneum and the paravertebral space [2-4]. It has also been described in the spinal epidural space, with a slight predilection for the thoracic spine [5, 6]. Fewer than 30 cases have been mentioned in the literature in the cervical area of adults, with only 4 presenting as a dumbbell-shaped mass in the foraminal region [1, 2].

Case Report

This is the case of a 37-year-old female who presented as an outside patient with a left cervicobrachialgia and acroparesthesia. An

electromyography (EMG) done revealed no particular radiculopathy or carpal tunnel syndrome. A cervical MRI (Figure 1) ordered showed a well-defined mildly enhancing soft tissue lesion obliterating the left foramen at C7-T1 in a dumbbell shape, with minimal medial extension toward the right anterior aspect of the cervical canal. The signal characteristic and its morphology oriented towards a nerve sheath tumor and were mostly consistent with a neurinoma. A posterior C5-T1 cervicothoracic laminectomy and arthrodesis associated with radical tumor resection was performed with decompression and preservation of the left C8 nerve root. Following the surgery, the patient completely recovered from the neurological symptoms, and showed no motor nor sensory deficit in her left arm. The pathology report later described a small round blue cell neoplasm, with positive stains for CD99 and NKX 2.2, highly suggestive of Ewing's sarcoma. A total body scan followed and revealed no other extension of the tumor. She was started on a course of chemotherapy, with a whole-body PET-scan performed a month later showing slight hypermetabolism in her left knee, confirmed to be non-suspicious on MRI. A cervical MRI done 5 months after the surgery noted the absence of residual tumor in the left C7-T1 vertebral foramina.

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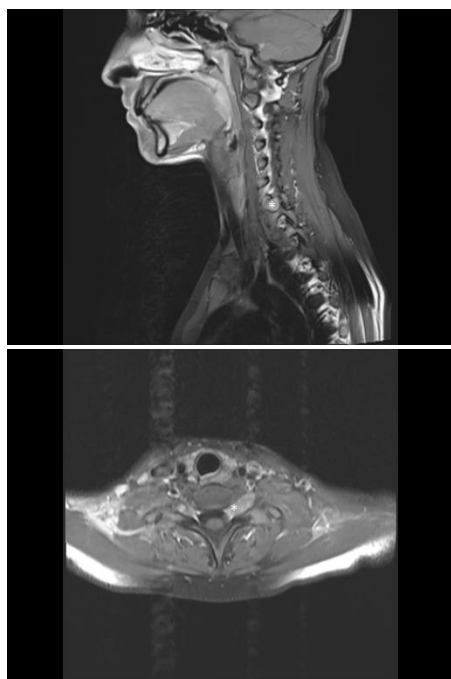


Figure 1: Sagittal and axial cervical MRI sequences with injection of gadolinium. The extraspinal dumbbell-shaped foraminal C7-T1 tumor is highlighted with an (*).

Discussion

With an annual incidence of 1-3 per million, Ewing's sarcoma is a rare diagnosis [7]. It's primary extraskelatal or extraosseous form (EES) is found in only 15% of the cases. It affects males and females equally, mostly in their second or third decade of life, while the classic skeletal

or osseous form (OES) manifests earlier on in life [2, 3, 8, 9]. Radiologically, the EES mass presents no specific findings according to the cases reviewed in the literature, and tends to enhance heterogeneously [8, 10]. Our lesion was described for instance as hypointense on T2 with intralesional cystic changes, hyperintense on T1-weighted images, and demonstrated mild enhancement following gadolinium administration on the cervical MRI performed. The definite diagnosis can only be made through histopathological analysis [11]. Traditionally, light microscopy with the aid of immunohistochemical stains was suitable for diagnosis of Ewing's sarcoma in both its forms. The most commonly used cellular marker is the CD99 (a 32-kDa cell surface glycoprotein encoded by the MIC2 gene), whose expression can be found in both Ewing's sarcoma and lymphoma [1, 11]. But now translocation analyses, notably the t(11;22) (q24;q12) which is positive in 88-95 % of ES/PNET cases, are being used not only for the diagnosis and classification of small round cell tumors, but to ascertain their prognostic significance, detect micrometastasis, and monitor minimal residual disease, with potential for targeted therapy [12]. Standard treatment often involves a multidisciplinary approach of surgery, chemotherapy, and radiation therapy with the particular preferential order determined on a case-by-case basis [1]. Survival rates for the EES vary across the literature papers, coming just around the 50% mark for most of them at 5 years of diagnosis [13].

Our case is special given the fact that the tumor both on the pre-operative MRI and during the surgery was confined to the vertebral foramina in an hourglass shape, without abutting into the spinal canal, typically mimicking a schwannoma. The diagnosis hence came as a surprise, especially in the absence of other known locations of the tumor. Only 4 previous cases of a cervical spinal epidural extraskelatal Ewing's sarcoma (EES) presenting as a dumbbell-shaped mass in the foraminal region have been described in the literature before (Table 1) [14].

Table 1: A review of the literature of the primary cervical dumbbell-shaped extraskelatal Ewing's sarcoma.

	Case 1	Case 2	Case 3	Case 4	Current case
Gender (M or F)	M	M	M	M	F
Age (years)	46	27	27	24	37
Symptoms	Pain Radiculopathy	Pain Radiculopathy	Pain Radiculopathy	Pain Radiculopathy SC compression	Pain Radiculopathy
Cervical level	C3-C6	C1-C4	C7	C5	C7-T1
Tumor location	Extra and intraspinal	Extra and intraspinal	Extra and intraspinal	Extraspinal	Extraspinal
Surgery	Anterior Posterior	Anterior Posterior	Anterior Posterior	Posterolateral	Posterior
Resection	Piecemeal	Piecemeal	Piecemeal	Piecemeal	Piecemeal
Metastasis	Lung	None	None	Brain	None
Year	2008	2010	2009	2011	2018
Author	Zhu <i>et al.</i> [14]	Zhu <i>et al.</i> [14]	Zhu <i>et al.</i> [14]	Zhu <i>et al.</i> [14]	Bteich <i>et al.</i>

Conclusion

Extraosseous Ewing's sarcoma (EES) is a rare form of Ewing's sarcoma which can manifest in the spinal canal or region as a rapidly-growing mass that has no specific radiographic nor histopathologic features, but which should be kept in the differential diagnosis of cervical tumors, even when affecting the vertebral foramina in a dumbbell shape. Surgery

should be done quickly to contain the spread of the malignancy, and multi-adjuvant therapy initiated after a full body check-up, which can be adjusted depending on the response to treatment. Survival is fair with less than 50% alive at 5 years from diagnosis.

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