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Adult primary cervical extra-osseous Ewing's sarcoma: A case report and short literature review

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ABSTRACT

INTRODUCTION: Primary spinal epidural extraskeletal Ewing's sarcoma (EES) is extremely rare, with a peak incidence in the second decade of life. EES in old people is challenging to treat due to the lack of specific guidelines. In this paper, I present a unique case of adult primary cervical epidural EES with a 13-month follow-up. A short literature review of the therapeutic approaches and prognosis is also presented.

PRESENTATION OF CASE: I present a case of a 49-year old male patient who presented with right upper limb pain, numbness, hand grip weakness, and hyperreflexia of 3 months duration. Enhanced cervical magnetic resonance imaging showed a homogenously enhancing epidural and paravertebral soft tissue mass extending from the C6 to the T2 that appeared hypointense on T1 and hyperintense on T2. The patient underwent biopsy that confirmed EES via histopathology. Treatment with chemotherapy and radiotherapy resulted in tumor resolution and symptom relief.

DISCUSSION: EES is a type of PNET. Surgical removal is generally the treatment of choice, followed by adjunctive chemotherapy and radiotherapy. In old patients with large tumors, a more conservative approach with biopsy, adjuvant chemotherapy and radiotherapy is recommended.

CONCLUSIONS: Adult primary cervical epidural EES is a neurosurgical challenge due to the extension to the surrounding vital structures making the tumor not amenable for total resection as in the present case. In the absence of specific therapeutic guidelines, our case highlights the need to individualize the treatment modality according to age, tumor extension, and feasibility of total tumor resection.

1. Introduction

Extraskeletal Ewing's sarcoma (EES) is a rare and highly malignant primitive neuroectodermal tumor (PNET) arising from the soft tissue. It has a predilection to the thoracic spine, and primary spinal location is rare [1]. Moreover, primary cervical localization is considered extremely rare, with only 13 cases reported in the medical literature from 1969 to 2015 [2]. Cervical spinal EES in old people is challenging to treat due to the lack of treatment guidelines. In this paper, I present the 47th case of a primary spinal EES in an adult, with the aim of enhancing the establishment of multidisciplinary approach for the best management strategy and possible molecular research that compare ES to EES in relation to treatment response and cell of origin.

This work has been reported in line with the SCARE criteria [3].

2. Presentation of case

2.1. History and physical examination

A 49-year-old male patient presented to the outpatient clinic with neck and right upper extremity pain, and generalized bilateral upper limb numbness of 3 months duration. The patient scored 5/5 on motor power examination, except the right hand grip, which was 4/5; atrophy of the right forearm; hyperreflexia of the right upper limb; and no other signs of myelopathy.

Enhanced cervical magnetic resonance imaging (MRI) (Fig. 1) showed partial collapse of the C7 with a homogenously enhancing epidural and paravertebral soft tissue mass extending from the C6 to the T2 that appeared hypointense on T1 and hyperintense on T2 with extension into the spinal canal through the right and left C6-C7 exit neural foramina causing cord compression with no myelopathy. The intervertebral disc space was not involved. Bone scan and positron emission tomography (PET) scan excluded bony and other soft tissue involvement, including C7 compressed vertebral body. Initial and follow-up bone scans showed no evidence of metastatic or osteoblastic activity and no uptake in C7 verte-

Abbreviations: CT, computed tomography; EES, extraskeletal Ewing's sarcoma; PNET, primitive neuroectodermal tumor; PET, positron emission tomography; MRI, magnetic resonance imaging.

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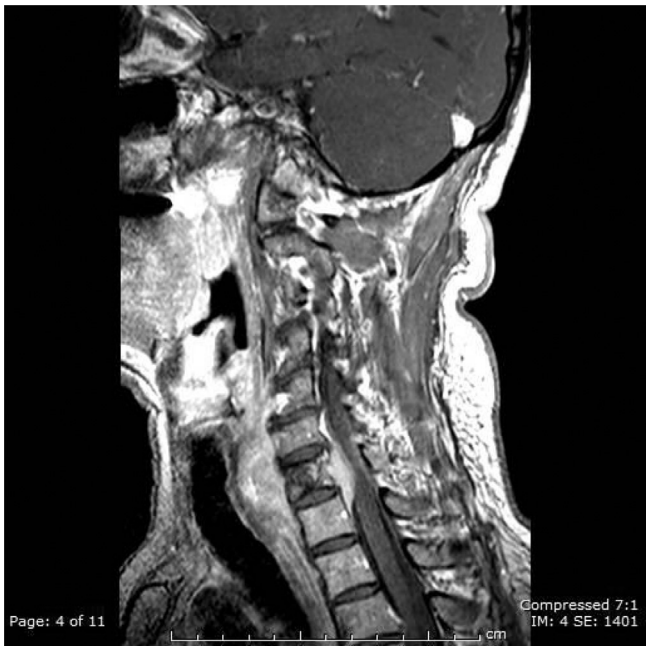

Fig. 1. Sagittal enhanced T1- weighted image of cervical spine show homogeneous extradural mass compressing the spinal cord. C7 vertebrae collapse with preserved intervertebral disks.

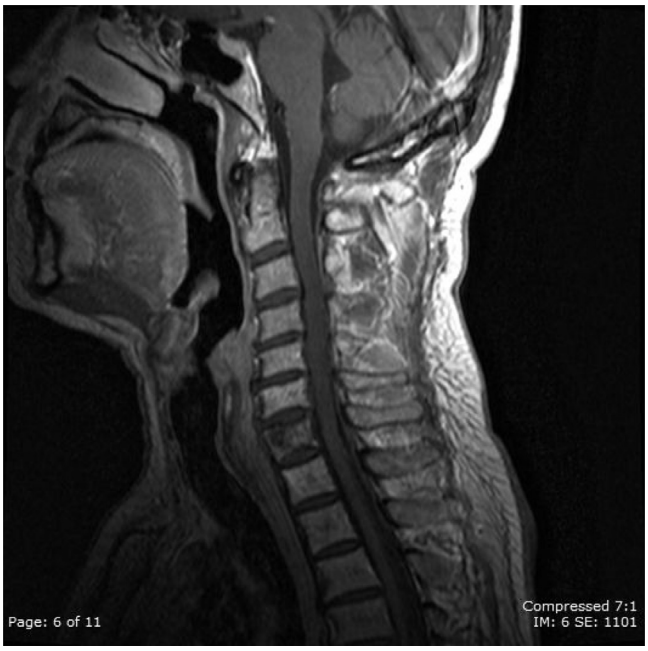

Fig. 2. sagittal enhanced T1–weighted image of cervical spine shows complete resolution of the intra-spinal mass with small pre-vertebral residual. C7 vertebrae partially collapsed.

brae. Computed tomography (CT) scan of the chest/abdomen/pelvis showed no evidence of other diseases.

Differential diagnosis: lymphoma, metastasis, multiple myeloma, TB, nerve sheath tumor, ES.

2.2. Surgical decision and operation

C7 anterior corpectomy with cage and plate followed by posterior laminectomy was considered. However, the tumor was circumferentially involving the C7 vertebrae and vertebral arteries with no uptake in the C7 bone. Consequently, total excision was not possible even with combined anterior and posterior approach.

The patient underwent biopsy via an anterior approach, which confirmed EES. A course of adjuvant chemotherapy was administered with close clinical follow-up. Profound clinical and radiological improvements were noted. The pain disappeared and numbness decreased. Follow-up cervical MRI one year after chemotherapy and radiotherapy (Fig. 2) showed that the intraspinal mass had disappeared completely.

2.3. Histopathology

The histopathology showed small, round, blue tumor cells characterized by high nuclear/cytoplasmic ratio, a thin rim of cytoplasm, and indistinct cell membranes that were diffusely positive for CD99 markers with weak focal positivity for the synaptophysin marker. However, they were negative for PAN-CK, EMA, S100, LCA, TdT, desmin, myogenin, and TTF-1 immunostaining. Ki-67 proliferative index was estimated to be 40%. Such results confirmed the diagnosis of ES.

3. Discussion

Primary spinal EES is an extremely rare malignancy, with a peak incidence in the second decade of life [4]. In this paper, I present a case of an adult with primary cervical EES with a 13-month follow-up. EES is a type of PNET that appears as a nonspecific spinal tumor on CT, MRI, and PET scan [5]. In the present case, the patient was aged 49 years, and the tumor showed homogenous enhancement of the soft tissue mass with no bony involvement under bone and PET scan. Definitive diagnosis was made via histopathological studies, with special consideration to differentiate EES from lymphoma [6].

Surgical removal is generally the treatment of choice for EES, followed by adjunctive chemotherapy and radiotherapy [7]; however, the choice of treatment is individualized because no therapeutic guidelines have been established. The clinical history, tumor extension, feasibility of total tumor resection, and age of the patient should be considered when determining the appropriate treatment modality [8]. Surgical decompression and tumor resection is initially necessary in patients with progressive neurological deficits and when the tumor is amenable for total or near total resection, particularly in young patients. However, in old patients with large tumors, a more conservative approach with biopsy and adjuvant chemotherapy and radiotherapy is recommended. The average 5-year survival rate for primary spinal ES is 45% after treatment [9]. Favorable predictors of survival are localized disease at presentation, time to local therapy <4 months, primary origin in bone, and objective response to chemotherapy [10,11].

4. Conclusion

Primary spinal EES is extremely rare. In this paper, I present a unique case of primary cervical spinal EES in a patient older than 40 years. No specific treatment guidelines have been established for EES. Consequently, treatment is individualized depending on patient age, progression of neurological deficit, tumor extension, and total resectability of the tumor.

Conflict of interest

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The author declares no potential conflict of interests. This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Funding

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Ethical approval

All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Consent

Written informed consent was obtained from the patient for 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.

Author contribution

Eloqayli H is the sole and corresponding author. I am Eloqayli H did study design, data collection, data analysis or interpretation, writing the paper.

Guarantor

I am Eloqayli Haytham accept full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

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