



RESEARCH ARTICLE

CASE REPORT OF PRIMARY EWING'S SARCOMA AT THE CERVICAL SPINE IN 19-YEAR-OLD MALE

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Manuscript Info

Manuscript History

Received: 22 March 2020

Final Accepted: 25 April 2020

Published: May 2020

Abstract

Ewing's sarcoma is rare to be in the spine. The age of presentation ranges from 12 to 24 years. It should remain a viable differential diagnosis of an ill-defined spinal mass noted on imaging. The combination of chemotherapy and surgical treatment offers the patient a good chance of survival with a satisfactory quality of life. . This is the case report with detailed history, examination of 19 years-old male patient with Primary Ewing Sarcoma in the Cervical Spine. The present report aimed to highlight on occasional involvement of the spine as a primary site of Ewing sarcoma with a greater challenge in surgical treatment and thus should be considered in the differential diagnosis of spinal tumors and related clinical features.

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Introduction:-

Ewing sarcoma is a cancerous (malignant) peripheral primitive neuroectodermal tumor that usually develops among 85% of cases in bone; it usually affects the long bones, such as femur, tibia, humerus. The bones of the pelvis are also often affected. Occasionally, this tumor may arise in the muscles, soft tissues and spinal cord (this is called extra skeletal Ewing's sarcoma).⁽¹⁾

Comparing with other type of cancers, although the Ewing's sarcoma are rare with an annual incidence of 1–3 per million^(2,3) but it is the second most common bone malignant in children and young adults.⁽⁴⁾ The median age of patients with Ewing sarcoma is 15 years, and majority of cases are adolescents.⁽⁵⁾ Researchers had reported some of Ewing sarcoma in neonates and infants.^(6,7)

However, there are Dramatic improvements in cancer prognosis for children and adolescents.⁽⁸⁾ For Ewing sarcoma, Treatment approaches for Ewing sarcoma consist of chemotherapeutic with the goal of maximizing local control, which includes surgery or radiation.^(8,9) The 5-year survival rate has increased from 59% to 78% for children younger than 15 years and from 20% to 60% for adolescents aged 15 to 19 years.^(9,10)

In Saudi Arabia, one research carried out in western region among the sixty-nine cases of Ewing's sarcoma and showed that the mean age was 22 years with almost 1:1 male to female ratio. Approximately 28.9% of cases presented within the skeleton, and 71.1% cases were presented as a soft tissue disease. Soft tissue affection showed a higher incidence in the head and neck region followed by spinal cord with 8.2 of all extra skeletal Ewing's sarcoma (EES).⁽¹¹⁾ While the spinal canal remains a rare site for Ewing's sarcoma to arise and this is upon performing literature review on different articles. In Saudi Arabia, there are low number of Ewing's sarcoma cases in spinal canal.

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Report of the case:

A 19 years old Saudi male presented to different physicians with an 8-month history of progressive complaints that started as a stiff neck and pain with right upper limb neurological symptoms, pain associate with numbness sensation in the right hands and constant lower back pain that was aggravated by movement. There is no history of trauma or injury and during this time, he did not get any improvements with conservative treatment including medication and physiotherapy.

However, he presented to the orthopedic clinic and the physical examination showed that he appeared alert, oriented person, slim. Generally vitally stable not in sever pain. There is neck limitation of movement (patient avoiding movement).there is tenderness in palpation mainly in the posterior cervical spine. There is pain with range of movement. Neurologically, cervical nerves examination was intact and muscle strength testing revealed 5/5 from C5 to T1. he has normal gait with diffuse tenderness over the upper dorsal region. In addition to that the reflexes and coordination were normal and without any sensory defects.. There was negative upper motor neuron lesion signs.

X-rays of the whole spine, chest, and abdomen were normal. Enhanced cervical magnetic resonance imaging (MRI) (**Fig.1**) of the cervical spine showed mild central wedging of C7 vertebra demonstrating bone marrow edema pattern with enhancement after contrast and no associated pre-or paraspinal or intraspinal soft tissue components. Moreover, T5-6 posterior disc protrusion effacing the anterior subarachnoid space and touching the ventral aspect of the cord. Furthermore, the patient was without evidence of metastatic disease at presentation as determined by CT scan.

CT guided biopsy showed small round cells with round to oval nuclei and Ewing sarcoma was confirmed. The immunohistochemically evaluation revealed diffuse cytoplasmic positivity for CD99 and S100; whereas no expression was seen with CD45, CD20 and CD3 The case was discussed under the hospital tumor board they advised to administer chemotherapy and additional radiotherapy as neoadjuvant therapy.

Subsequently, the patient underwent surgery, the C7 cervical subtotal corpectomy with left anterior cervical approach. Anterior reconstruction was carried out using a titanium cage filled with bone allograft and plate fixation. The tumor was sent to pathology to confirm the necrosis percentage and it was 90 % which indicate tumor had great response to the neoadjuvant therapy.

Following the surgery, the patient had a good initial improvement without post-operative complication. . Afterthought, the patient got follow up with the both Oncology and orthopedic department for surveillance purposes.



Figure 1:- Cervical MRI showing mild central wedging of C7 vertebral body.

Discussion:-

Extra skeletal Ewing's sarcoma of the spinal canal has been a subject of interest because of uncommon location and vague presentation, it remains a rare diagnosis. EES affects males as often as females during the second or third decade of life whereas the primary skeletal Ewing's sarcoma occurs slightly early.^(11, 12) The present case is 19 years old Saudi male with Extra skeletal Ewing's sarcoma of the cervical spine.

Ewing's sarcoma usually presents with local swelling and pain from the rapidly enlarging mass.⁽¹³⁾ When this occurs in the spinal canal as in the present patient, commonly present with progression pain that associated with other neurological symptoms such as weakness, numbness, tingling, or gait disturbances.^(14,15) Even when initial X-ray studies reveal no abnormality, the physicians should be alert to the possibility of spinal malignancy, especially when the patient is in the first two decades of life and symptoms including Local back pain that is not relieved by medication and physiotherapy.

In terms of imaging, MRI is very sensitive in the early detection of ES in the spine, ruling out the possibility of tumor spread to the bone marrow or extension into adjacent soft tissues or spinal cord compression.⁽¹⁶⁾ MRI's findings may determine the treatment plans. ES should be included in the differential diagnosis of nonspecific spinal canal lesions that include neuroblastoma, Ewing's sarcoma, malignant lymphoma, and rhabdomyosarcoma.^(13,17) Our lesion was hypodense on enhanced at C7 vertebrae with contrast administration.

Findings include uniform small round blue cells are the definitive diagnosis that made by histopathological studies.^(1,2) Typically, in order to confirm a diagnosis of Ewing sarcoma, the pathologist must rule out multiple other differentials through the process of multiple immunohistochemically staining. However, there are two cellular markers frequently evaluated by pathologists that are CD-99, which is expressed in both Ewing's sarcoma and lymphoma, and CD-45, which is only expressed in lymphoblastic lymphoma.⁽¹⁾

In the context of treatment, there is strong evidence that early and aggressive combined treatment leads to the most favorable outcomes.⁽¹³⁾ In other cases, there was strong evidence of a rapidly enlarging mass with rapidly progressive neurologic deficit. This warranted the first goal of operation to be decompression of the spinal cord with the proposed laminectomy.⁽¹⁸⁾ The present case, the mass was not producing compression on spinal cord and based on the recommendations of the hospital tumor board that advised to proceed first with possible preoperative chemotherapy and radiation therapy, shrinking the tumor and promoting an increased chance of successful gross total resection. However, the researchers indicated that an aggressive multimodal treatment approach involving surgery, chemotherapy, and local radiation therapy increases the chance of a successful outcome.^(9,10)

Ethical consideration:

The patient and his parents were informed that data from the case would be submitted for publication, and they gave their consent.

Conflicts of interest:

None

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