

MR Imaging of extraosseous ewings sarcoma with Histo-pathological correlation- A rare Case Report

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Abstract :

We report a case of Extraskeletal Ewing sarcoma (ES) in a 24-year-old male patient who presented with a painful rapidly enlarging mass in the left thoracolumbar paraspinal area. MRI showed a heterogeneous soft tissue lesion involving the left dorsal paravertebral muscles with extension into the ipsilateral psoas muscle extended over multiple vertebral segments in a cranio-caudal direction (T11-L3). MRI with contrast enhancement showed a solid tumor surrounding areas of necrosis/cystic degeneration. There was no intraspinal extension or neural foraminal invasion. Pathologically, the lesion was proved to be a Extraskeletal Ewing's sarcoma. Herein, we discuss the clinical, radiological, and pathological features of this disease entity.

Keywords : Extraskeletal Ewing sarcoma (ES), MRI, Paraspinal musculature.

Introduction :

The extraskeletal variant of Ewing sarcoma (ES) is a rare lesion with scant reports in the radiology literature, especially on MR studies. Skeletal ES more frequently occurs in the long bones of the lower extremities in young patients (mean age: 10 years). The soft tissue variant of this malignancy affects an older age group (mean age: 20 years) and has a wider distribution of localization. It can occur in the head/neck, trunk, diaphragm, retroperitoneum, pelvis and extremities. Most reports emphasize the paraspinal area as the most frequent site of origin. We present the imaging features of a rare case of extraosseous paravertebral ES.

Case History :

A 24-year-old male presented with a 2 month history of a painful rapidly enlarging mass in the left paraspinal thoracolumbar area. Physical examination revealed a firm, non mobile subcutaneous mass.

MRI and Histopathological Findings

Preoperative imaging included MR (Fig. 1-3). An inhomogeneous soft tissue lesion of the left dorsal paravertebral muscles with extension into the ipsilateral psoas muscle extended over multiple vertebral segments in a cranio-caudal direction(T11-L3). The margins are

smooth and become evident in the T2w images and post contrast. The mass is mostly T1 isointense and moderately T2 hyperintense relative to muscle, indicative of high cellularity. Thin hypointense septae separate locules of solid tumor from degenerated/necrotic areas. There was no extension into neural foramina or in the spinal canal.

An open biopsy was performed. Macroscopically, the tumor was firm, whitish-gray in color and vascular. Microscopic examination revealed small, uniform cells arranged in solid nests separated by fibrous strands. A thin layer of cytoplasm enveloped round nuclei containing small nucleoli. A high mitotic activity was seen. Periodic acid-Schiff (PAS) and PAS-diacetate stains were positive for cytoplasmic glycogen.

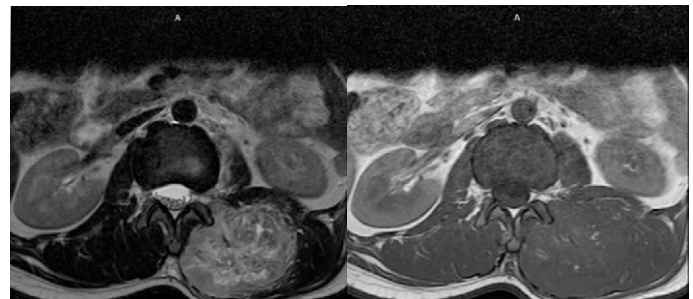


Fig 1: T1W and T2 W axial images show an inhomogeneous soft tissue lesion of the left dorsal paravertebral muscles with extension into the ipsilateral psoas muscle. The margins are smooth and become evident in the T2w images. The mass is mostly T1 isointense and moderately T2 hyperintense relative to muscle, indicative of high cellularity.

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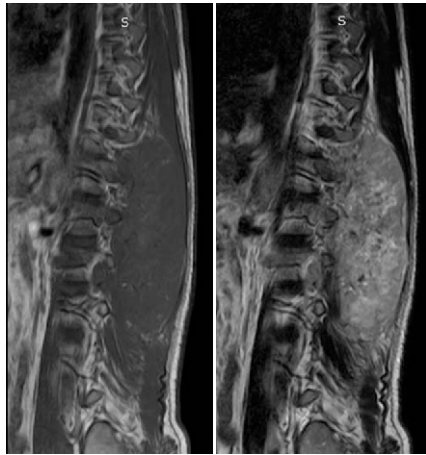


Fig 2 : T1W and T2W sagittal images show an inhomogeneous soft tissue lesion of the left dorsal paravertebral muscles extending over multiple vertebral segments in a cranio-caudal direction(T11-L3). The margins are smooth and become evident in the T2w images. The mass is mostly T1 isointense and moderately T2 hyperintense relative to muscle, indicative of high cellularity.



Fig 3 : Post contrast T1 axial and sagittal images show significant enhancement of the mass with non-enhancing central necrotic area.

Discussion : ES falls in the category of small round (blue) cell tumors of children and adolescents. It differs in presentation from skeletal ES in several respects. The average age of occurrence of EES is 20 years, in contrast to 10 years for skeletal ES. EES occurs equally in both sexes, whereas skeletal ES has a 2:1 male predilection^{1,2}. EES commonly affects the soft tissues of the trunk, such as paravertebral and intercostal regions, head and neck, pelvis, and peritoneum. In contrast, skeletal ES has a predilection for the long bones of the lower extremities. Other rare reported locations of EES include the mediastinum, heart, external genitalia, and broad ligament^{2,3}. Macroscopically the tumor is lobulated with a rich vascular supply. Areas of hemorrhage, necrosis and cystic degeneration are common findings. Some

tumors appear well contained by a fibrous pseudocapsule. Microscopically compact clusters of uniform, round or oval cells supported by trabeculae of fibrous tissue are seen. The cells are small with high nucleocytoplasmic ratio, finely divided chromatin and minute nucleoli. Despite a scant amount of cytoplasm it is easy to demonstrate the intracellular glycogen stores. The radiologic findings reflect the histopathologic composition of the tumor. On MR images, the EES is generally of low to isointense signal compared with muscle on T1 weighted images, of high signal intensity on T2-weighted images, and exhibits heterogeneous enhancement⁴⁻⁶. It is known that both local recurrences and distant metastases are common in EES⁷. It is stated that the 5-year survival rate of EES is 61%, whereas that of skeletal ES is approximately 10%⁸. In conclusion, the diagnosis of ES should be considered for aggressive paravertebral tumors in the appropriate age group (teenagers to young adults). The age and paravertebral location may be the clues to diagnosis.

Conclusion :

EES is a rare disorder and carries a poor prognosis. Radiology plays an important role in its management in terms of diagnosis and monitoring treatment. Identifying EES involving the paravertebral muscles is important and is usually clinched by a combination of clinical, radiological, and histological features.

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