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Case Report Open Access

Primary Spinal Epidural Rhabdomyosarcoma of the Upper Thoracic Spine

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Abstract

Introduction: Rhabdomyosarcoma is a highly aggressive and rapidly growing sarcoma with skeletal origin that occasionally appears in the spinal epidural space.

Method: We report a 20-year-old girl who presented with back pain, progressive paraparesis, and urinary retention.

She had muscular weakness in her lower extremities and absent deep tendon reflex.

An epidural dumbbell-shape mass at T3-T4 level was observed on MRI.

The patient underwent T3-T5 hemilaminectomy.

Result: Histopathological examination Immunohistochemical staining confirmed the diagnosis of alveolar Rhabdomyosarcoma. She received radiotherapy and chemotherapy. The patient died 3 months after.

Conclusion: Primary spinal epidural RMS is an extremely rare and very aggressive tumor. The treatment should focus on extensive resection with intensive combination of radiotherapy and chemotherapy.

Keywords: Rhabdomyosarcoma; Thoracic spine; Epidural tumor

Introduction

Rhabdomyosarcoma (RMS) is a highly aggressive and rapidly growing neoplasm of skeletal muscle origin that occasionally appears in the vertebral column and spinal epidural space [1]. This report deals with a case of rhabdomyosarcoma in the upper thoracic spine with a particular interest, not only for the rarity of type but also the location of this tumour.

Case Report

We present a rare case of Rhabdomyosarcoma in a 20 years young girl, who presented with back pain at the level of T4, included an acute demonstration of complete paraplegia. Power of the lower limbs was 1/5 both distally and proximally, with absent deep tendon reflexes and severe asymmetrical sensory loss below the T4 dermatome bilaterally. Spine MRI, before and after gadolinium (Gd) administration, revealed an epidural dumbbell-shape mass at T3–T4 area compressing the cord laterally and extending through the T3–T4 neural foramina on the right side. Leptomeningeal metastasis was found (Figure 1). A large, firm, dark-red vascular epidural tumor was found compressing the cord; T3-T5 laminectomy with large surgical biopsy, and foraminal parts of the tumor was removed. The history followed by partial recovery gradual relapsing of neurological deficit, one month later. Histopathological examination revealed undifferentiated small round and oval tumoral cells (Figure 2). Immunohistochemical staining was positive for vimentin, desmin, and myogenin. The final diagnosis was alveolar Rhabdomyosarcoma. The patient was treated with chemotherapy followed by radiotherapy. But a major progression of the disease leads to the death 3 months after diagnosis.

Discussion

Primary spinal epidural RMS is an extremely rare tumor and only few cases have been reported in the literature [1-3]. RMS occurs sporadically and no predisposing or risk factors have been recognized in the majority of cases [4], it is thought that RMS arises as a consequence of regulatory disruption of skeletal muscle progenitor cell growth and differentiation [5]. The disease has been associated with familial syndromes, including Li-Fraumeni syndrome, neurofibromatosis type 1, and hypomelanosis of Ito [6]. RMS has also been reported to occur as a congenital tumor. Congenital dysrhaphic malformation of the spine may predispose children to intra-lesional development of RMS [7].

The median age of presentation is 6 years; however, this disease follows a bimodal distribution with peak incidences between 2 and 6 years and again between 10 and 18 years of age [8]. There is a slight male to female predominance (5:3) and no known predilection for race [9]. RMSs are classified into four types: embryonal, botryoid, alveolar, and pleomorphic. Alveolar RMS account for 10-20% of all RMSs and affect chiefly children and young adults between 10 and 25 years of age [10]. Alveolar RMS is associated with a significantly higher risk of relapse and a much higher risk of metastasis [10]. Clinically, spinal RMS can present with pain, limb weakness, and bladder or bowel dysfunction. The pain may be localized or dermatomal. The limb weakness is usually spastic, but may be flaccid if the conus medullaris or the cauda equine is involved as in our patient. Other symptoms and signs include localized swelling, scoliosis, torticollis and sensory disturbance [2,3,6,11]. Radiologically, primary spinal RMS is usually hypointense on T1Wand hyperintense on T2W images and shows homogeneous or inhomogeneous enhancement after Gadolinium injection [2,3]. There are a number of differential diagnoses for this tumor in the spinal canal including hemangioma/vascular malformation, peripheral neuroectodermal tumors, Ewing's sarcoma, lymphoma, neuroblastoma, and Meningiomaand immunohistochemical study is mandatory for a definitive diagnosis [2,12].

This malignant tumor invades local structures and metastasizes to remote sites by lymphatic and hematogenous spread. Children with metastatic RMS have poor survival rates [13]. The most common

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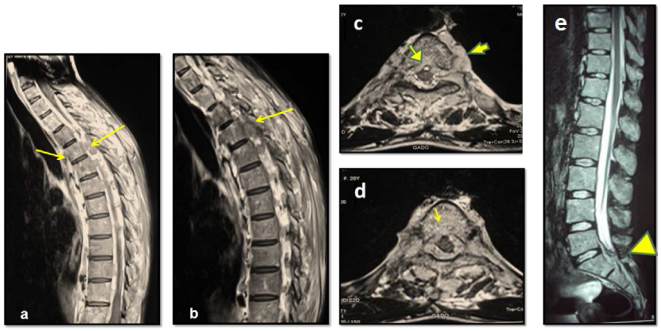

Figure 1: (a) Sagittal T2-weighted MRI showing an isosignal epidural mass at T3–T4 level. (b) Sagittal and axial (c and d) contrast enhanced T1-weighted MRI showed an epidural dumbbell-shape mass at T3- T4 level extending through the right T3-T4 foramen with marked enhancement and involvement of T4 vertebra. (e) Sagittal T2-weighted MRI showing lumbar epidural metastasis.

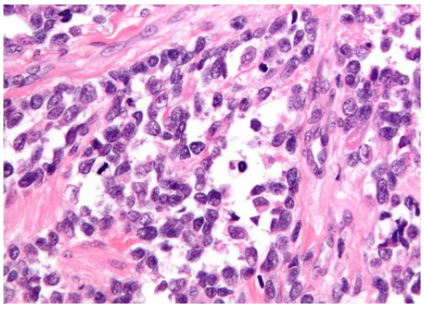

Figure 2: Histopathology examination of the tumor. Fibrous septa with loose clusters of rounded cells in center (hematoxylin-eosin, 100X).

metastatic sites from primary rhabdomyosarcoma are lung, bone, bone marrow and liver. Our patient had l*epto-meningeal* dissemination [13].

Treatment of spinal RMS requires a multidisciplinary approach and includes a combination of surgery, chemotherapy, and radiation based on the Intergroup Rhabdomyosarcoma Study groups which divide patients into low-risk, intermediate-risk, and high-risk groups [14]. Using this multimodality approach, the cure rates for RMS have steadily increased from only 25% in the 1970s to 70% in the 1990s [15].

Conclusion

Primary spinal epidural RMS is an extremely rare tumor that should be included in the differential diagnosis of spinal epidural tumors. It is a very aggressive tumor. The prognosis for a patient with RMS is related to patient age, site of origin, extent of tumor at the time of diagnosis, tumor histology, and presence or absence of metastases. Improved outcomes may be achieved by advances in multidisciplinary (pediatric oncology, pathology, radiotherapy, and surgery) management and supportive care.

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