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Article · August 2022

DOI: 10.52910/ajhs.87

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A Case of Dorsal Spine Rhabdomyosarcoma in a 2 years old Child

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ARTICLE INFO

Article History
Submitted: 20 May, 2022
Accepted: 16 June, 2022
Published: 8 August, 2022

Source of support: None
Conflict of Interest: None

ABSTRACT

Rhabdomyosarcoma is highly aggressive malignant form of mesenchymal tumor arising from skeletal muscle cells rhabdomyoblast, that have failed to fully differenciate. It is most commonly seen in the children before 12 year of age. Primary spinal rhabdomyosarcoma is very rare. A 2 year old girl presented with mass over right upper back and inability to move lower limb. Patient underwent dorsal laminectomy and excision of the mass. Histology and molecular pathology helps in definite diagnosis and further need of chemotherapy or radiotherapy.

Keywords: Dorsal spine; Rhabdomyosarcoma.

INTRODUCTION

Rhabdomyosarcoma (RMS) is highly aggressive malignant form of mesenchymal tumor arising from skeletal (striated) muscle cells rhabdomyoblast, that have failed to fully differenciate.[1–3] RMS is the most commonly seen in the children before 12 year of age. RMS primarly found in head, neck, orbit, genitourinary tract, genitals and extremities. They can be primary spinal RMS or metastatic spinal RMS. They have a worst prognosis in adult than in the children with a 5 year survival rate in adult and child is 27% and 64% respectively. In this case report, we describe a patient who underwent surgical resection of a RMS from dorsal spine. We discuss the presentation, diagnosis, and management.

CASE REPORT

A 2-year-old girl presented with mass over right upper back since 3 months and inability to move lower limb since 15 days. She had no history of urinary retention. She had no prior history of radiation exposure. On examination, she had severe pain and tenderness over dorsal region. Motor power in the lower limb was 0/5 both distally and proximally with absent deep tendon reflexes

Magnetic resonace imaging (MRI) of the dorsal spine revealed showed T1 isointense area which appered heterogeneously hyperintense in T2 and T2 STIR sequences, located along the dorsal spine.The intradural component is located at right lateral thecal sac extending from D3-D5 vertebral levels with large extraforaminal component in the posterior mediastinum and in right paraspinal intrinsic muscles of the back with associated widened neural foramina at D4 level.The intradural component has compressed and displaced the adjacent cord towards left side,at the adjacent levels.Post contrast study shows heterogenous enhancement of mass lesion. Patient underwent dorsal laminectomy. Intradural component of the tumor excised, followed by excision of intrapleural component. Pleura was repaired with fascia graft. Tumor was grayish white mild- moderately vascular soft to hard tumor extending to pleural cavity.

Histopathological examination revealed embryonal Rhabdomyosarcoma; predominantly round cells arranged in diffuse pattern.The cells were intermediate in sizes.

The nuclei were round to oval and pleomorphic. Some foci showed spindle nuclei also. In some areas some of the cells are large with deeply eosinophilic cytoplasm with eccentrically placed nuclei(Rhabdomyoblast like cells). It also showed frequent mitotic figures and necrosis. Focal area shows tumor cells infiltration in the surrounding adipose tissue.Necrosis and bony trabecula were also observed. Her immunohistochemical report showed positive for desmin, myogenin, CD99 and FLI-1 and negative for synaptophysin,CD45, CK. The features are consistent with embryonal RMS. The metastatic survey on chest, abdomen, and pelvis revealed negative findings. Postoperative course was uneventful and her motor power of lower limbs has gradually improved. Postoperatively, the patient was scheduled to refer to the pediatric oncologist and received radiotherapy (54Gy in 30 fractions) and chemotherapy by cyclophosphamide (12.5 mg) and vinorelbin (14 mg) for 12 cycles. MRI of whole spine was done after 1 month of operation showed no evidence of tumor regrowth.

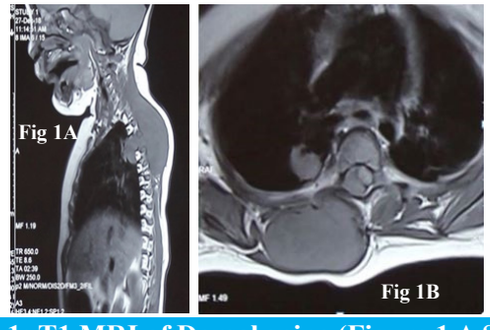

Figure 1: T1 MRI of Dorsal spine (Figure 1 A Sagittal view, Figure 1 B axial view)which shows isointense area at D2-D6 vertebral levels.

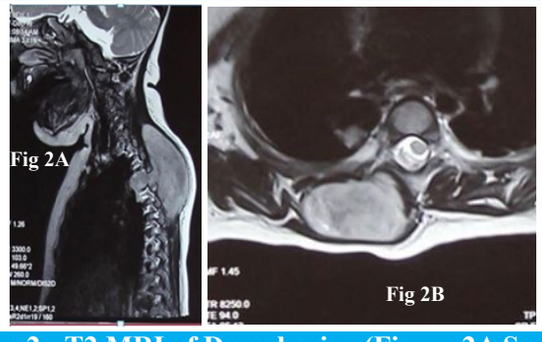

Figure 2: T2 MRI of Dorsal spine (Figure 2A Sagital view, Figure 2B axial view) which shows heteroenous intensity.

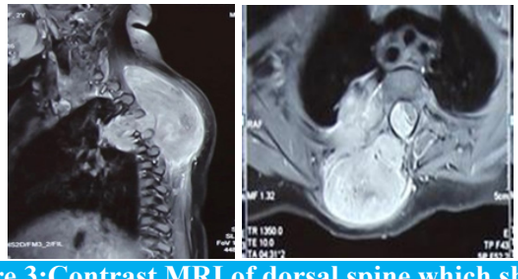

Figure 3:Contrast MRI of dorsal spine which shows heterogenous enhancement.

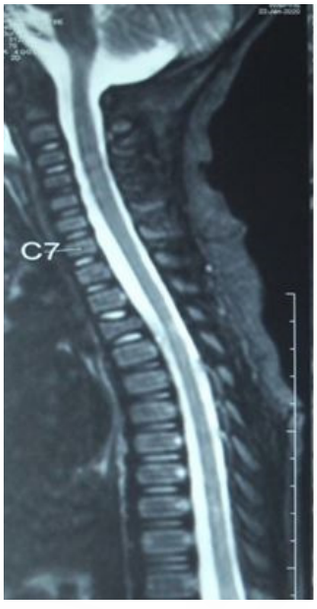

Figure 4: Post operative MRI of whole spine which shows gross total excision with no evidence of regrowth.

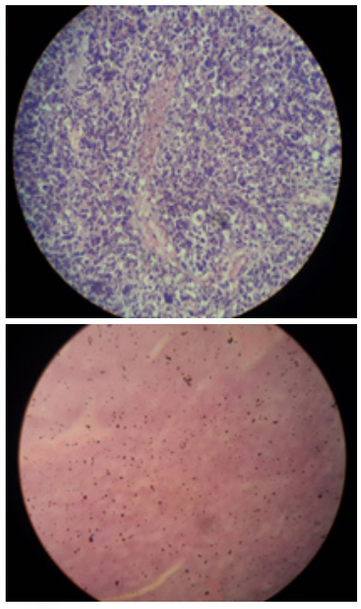

Figure 5: Histopathological examination showing predominantly round cells arranged in diffuse pattern.

Figure 5A & 5B shows the histopathological examination where he cells were intermediate in sizes.The nuclei were round to oval and pleomorphic. Some foci showed spindle nuclei also. In some areas some of the cells are large with deeply eosinophilic cytoplasm with eccentrically placed nuclei(Rhabdomyoblast like cells). It also showed frequent mitotic figures and necrosis.

DISCUSSION

RMS is a fast-growing, primitive, high-grade, malignant mesenchymal tumor. Rhabdomyosarcoma is a fast-growing, primitive, high-grade, malignant mesenchymal tumor. Primary spinal RMS is an extremely rare and only few cases have been reported. RMS is highly aggressive malignant form of mesenchymal tumor arising from skeletal (striated) muscle cells rhabdomyoblast, that have failed to fully differenciate.[1–3] RMS primarly found in head,neck,orbit,genitourinary tract,genitals and extremities. No risk factors,but associated with some congenital anomalies like NF1,Beckwith-wiedemann syndrome, Li-Frakmeni syndrome,DICER syndrome, LOS TELLU Syndrome.[5,6] 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. There is a slight male to female predominance (5:3) and no known predilection for race. Four types are distinguished: 1) Embryonal most common(>70%) with three subtypes a)Spindle cell (50-60%), b)Botryoid (5-(10%) with good prognosis) and c) Anaplastic. 2)Alvelor 20%, 3)Pleomorphic (5%) and 4)

Mixed type. RMS can be categorized as embryonal, alveolar, spindle cell/sclerosing, and pleomorphic according to the current WHO classification.[7] The Embryonal type is most common with characteristic round cells looking like lymphocytes and spindled cells with a elongated nuclei and eosinophilic cytoplasm. The Alveolar type has areas of spaces lined by non cohesive round or oval cells.[8] Pleomorphic RMS is common subtype in adults,that shows large plemorphic rhabdomyoblasts with eosinophilic cytoplasm. [9–11] The mixed type involves more than one histologic subtype.

Clinically, tumors present with wide range of symptoms ,depending upon the location. Limb and trunk tumors may present with painless swelling. spinal RMS may present with localized or dermatomal pain,torticollis,scolios,sen sory disturbance and spastic limbs weakness or flaccid limbs weakness if conus medullaris or cauda equine is involved along with bladder or bowel incontinence.[12–15]

In our case, the patient presented with swelling in the right upper back since 3 months which was soft on palpation and was non tender along with bilateral lower limbs weakness (spastic)since 15 days with motor power of 0/5 in bilateral lower limbs. Radiologically in MRI primary RMS in T1 is isointense to slight hyperintense, T2 shows hyperintensity, and contrast MRI shows marked contrast enhancement with intralesional hemorrhage or necrosis. Embryonal RMS shows enhancement and Alveolar and pleomorphic RMS shows areas of necrosis.[5,16] Though MRI helps to describe localization and extent of such lesions however it doesnot provide definite diagnosis. Differencial diagnosis in the spinal canal include Hemangioma, peripheral neuroectodermal tumors,Ewing's-sarcoma,Lymphoma,neuroblastoma and meningioma.[12,17,18] Diagnosis of spinal RMS can be done on the basis of Histopathological appreance and immunohistochemical(IHC) markers .[19]Immunohistochemical staining with antibodies against myogenin and myo D1 is considered to be an important diagnostic criteria for RMS and for differerential diagnosis with other tumors.[20] Treatment for RMS needs multidiscipilinary approach, where surgery , radiotherapy and chemotherapy each has its own specific role.The goal of surgery for spinal RMS includes complete excision of tumor and preserve neurological function and regard for stability in the growing spine in case of pediatric spinal RMS. For the patients with microscopic or gross residual disease following initial surgery, radiotherapy helps in local control. Follow up imaging is important to monitor tumor regression during or after completion of chemotherapy and radiotherapy and to detect tumor recurrence or metastases.[8]

CONCLUSION

Primary spinal Rabdomyosarcoma is rare.Histology and molecular pathology helps in definite diagnosis and further need of chemotherapy or radiotherapy.

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NPRS= Numeric pain rating scale, GROC = Global rate of change, QOLS: Quality of life Scale