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Giant Rhabdomyosarcoma in A Child: A Rare Entity

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Rhabdomyosarcoma (RMS) is a highly aggressive and rapidly progressive sarcoma. RMS can occur in any part of the human body, however, paraspinal or paravertebral or epidural RMS is very rare in children. We present a case report of 3 year old male child with right paraspinalRMS with intracanalicular extension at L3– S1 level, presented with painful swelling at lower back region. We will review the literature and discuss on epidemiology, diagnosis, management strategy and prognosis of RMS.

Key words: epidural, embryonal, outcome, rhabdomyosarcoma, paraspinal, paravertebral, Rhabdomyosarcoma, Sarcoma, treatment

Rhabdomyosarcoma (RMS) is a fast growing, malignant tumor of mesenchymal cell origin. RMS accounts for slightly less than half of the soft tissue sarcomas occurring in children and it is the third most common extracranial solid tumor in children after neuroblastoma and Wilm's tumor, but primary paraspinal or paravertebral location of RMS is rare in children[1,2,3]. The most common presentation of RMS is a soft tissue mass with or without accompanying signs and symtoms of organ dysfunction depending on the site of tumor origin. Paraspinal RMS may present with local swelling at paraspinal area, rarely with features of spinal root or cord compression symptoms[4]. Primary goal of treatment should be total excision of mass followed by chemo and radiotherapy. About 70% of RMS is chemo and radiosensitive and 5 year survival after multimodality treatment is about 60-70%[5].

Case report:

This 3 year old male child presented with lump in the back for last two months. According to his parents, they noticed lump in the lower back since two months which was gradually increasing in size which was painful on palpation. There was no significant past medical and family history. There was no history of weakness of lower limbs and bowel and bladder dysfunctions. On local examination, there was a mass at left paraspinal area at the level of L3 – S1 level which was tender, non pulsatile, non collapsable, non mobile, hard in consistency and overlying skin was normal.The size of the mass was 5.2 x 6.3 x 5 cm. The transillumination test was negative and no bruit on auscultation was heard.There was no motor and sensory deficit with normal reflexia. He had normal bowl and bladder functions.

MRI of spines showed moderately enhancing bilobed left paraspinal mass of about 6x5x5.2x5x4x6 in size with intracannalicularextention at L4-L5 level. Findings of MRI was suggestive of either paraspinalneurofibroma or neurogenic tumor or soft tissue sarcomas **(Fig.1a,b&c)**.

Plain CT scan of spines showed no involvement of adjacent spines **(Fig.2a & b)**

Hematology, biochemistry and serology were within normal limits.

The child underwent gross total excision of mass under general anesthesia. Tumor was solid, moderately vascular, bilobed and located at left paraspinal area. There was no muscle, bone and neural tissues involvement. Postoperative recovery was uneventful and the child was discharged on 7th post op day after removing stitches.

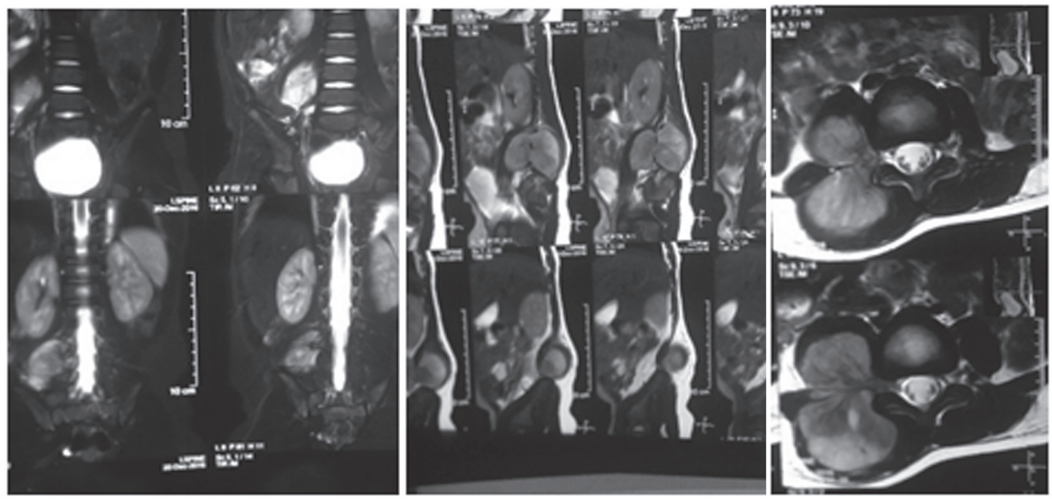

Figure 1: 1a. MRI of spine T2W2 coronal image showing isointense mass in the right lumbar region.1b. MRI of spine T2W2 sagittal images showing para vertebral mass over the lumbar sacral region.1c. MRI of Spine T2W2 axial image showing isointense para vertebral mass with intracanalicular extension.

Histopathological picture was suggestive of high grade sarcoma(Fig.3a,b&c) and pathologist recommended for imumunohistochemistry (IHC) for confirmation and typing. IHC showed Desmin, Myogenin and S-100 markers positive in neoplastic cells which are consistent with embryonalRhabdomyosarcoma.

The child was sent to pediatric oncologist for a course of chemo and radiotherapy. Imaging was repeated after a course of chemotherapy which revealed residual mass in paravertebral area at L3 – S1 level on left side. Then further treatment strategy was discussed among treating neurosurgeons and oncologist. Oncologist strongly favored for second surgery to have effective response of second course of chemo and radiotherapy. Retroperitoneal total excision of residual mass was performed with the help of pediatric surgeon. Then again he was sent back to oncologist for remaining course of chemo and local radiation.

Discussion:

Rhabdomyosarcoma (RMS) is a highly aggressive malignant tumor that develops from striated muscle (skeletal) cells that have failed to fully differentiate. It is mainly a disease of children accounting for 5-8% of childhood tumors and occurs below the age of 18[1,2,3]. RMS is the most common type of soft tissue sarcoma in children and it can begin in anywhere in the body. It commonly occurs in the head, neck, genital or urinary organs. It can also occurs in anus, legs, chest, abdomen, however, paraspinal or paravertebral RMS is very rare and only few cases have been reported in literature[1,2,3,4].

There are three main types of RMS and they are embryonal, alveolar and anaplastic. Embryonal sarcoma is the most common type of RMS and most often found in head and neck or in genital or urinary organs or epidural or paraspinal. Embryonal RMS usually affects children in their first 5 years of life accounting about 60-70% of childhood RMS[5]. Embryonal RMS is characterized spindle shaped cells with a stromal rich appearance and morphology is similar to the developing muscle cells of a 6-8 week old embryo. People who have inherited diseases like Li-Fraumeni Syndrome, pleuropulmonaryblastoma, neurofibromatosis type I, Costello syndrome, Beckwith – wiedemann syndrome and Noonan Syndrome are at risk of having embryonal RMS. Alveolar RMS is the second most common type and comprises 20-25% of RMS related tumors[6,7,8]. It is equally distributed among all age groups. Alveolar RMS is characterized by densely packed, round cells that arrange around spaces similar in shape to pulmonary alveoli. Alveolar RMS tends to form more often in extremities, trunk and peritoneum. It is more aggressive than Embryonal RMS. Anaplastic (undifferenciated) RMS, also known as pleomorphic RMS is the final variant of RMS. It occurs most often in adults, rarely in children. Alveolar RMS is defined by presence of anaplastic cells with large, lobulatehyperchromatic nuclei and multipolar mitotic figures. It is the most aggressive type of RMS and will often require intensive treatment. There is also an extremely rare subtypes of RMS that has been described as sclerosing RMS characterized by hyaline sclerosis and pseudovascular development. Its origins are unclear, but some studies have pointed to an association with embryonal RMS[7,8,9].

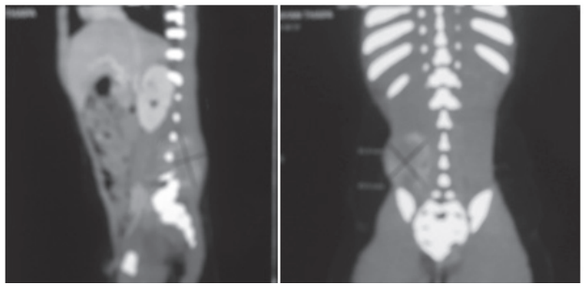

Figure 2: 2a & b. CT Scan of whole spine showing isodense paravertebral mass on right lumbosacral region without involvement of spines.

Prognosis of paraspinal RMS depends on age, tumor site, extent of resection, tumor size, regional lymph nodes involvement, presence of metastasis, site and extent of metastasis and histological characteristics of tumor cells[6,8].

Primary spinal epidural embryonal RMS is an extremely rare tumor and only few cases have been reported in literature[1,2,3,4,9]. Clinically RMS can present with local swelling and pain in the paraspinal area, later on can develop weakness of the limbs and bladder and bowel dysfunction. On MRI of spines Embryonal RMS is usually hypointense on T1W1 and hyperintense on T2W2 images and shows homogenous or inhomogenous enhancement after Gadolinium injection[7]. Often, CT Scan of spine is necessary to appreciate bone involvement. There are number of differential diagnosis for paraspinal or epidural masses to be considered like Peripheral neuroectodermal tumors, Ewing's sarcoma, lymphoma, neuroblastoma, meningioma and nerve sheath tumors. Embryonal RMS invades local structures and metastases to remote sites like lung, bone, liver etc. Treatment of spinal Embryonal RMS is gross total resection followed by chemo and radiotherapy. Fortunately, RMS is highly chemosensitive with approximately 80% of cases responding to chemotherapy. Modern survival rates with adjuvant therapy are 5 year in 60-70%. Approximately 30% of children with RMS will experience tumor recurrence and most of these patients will eventually die of progressive disease[1,2,5,6,8]. Recurrence usually occurs within three years of diagnosis[6,8].

Conclusion:

Primary paraspinalEmbryonal RMS is extremely rare tumor. It is very aggressive lesion, but 5 year survival is 60-70% if multimodality treatment like surgical resection, chemotherapy and radiotherapy are used in time.

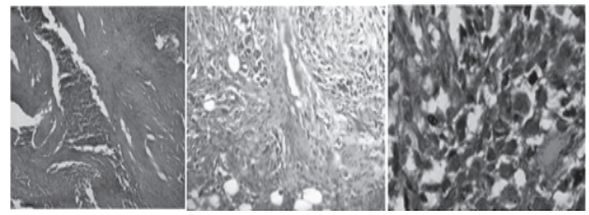

Figure 3:Histological appearances. Fig 3a,b&c : These tumor cells exhibit moderate to marked pleomorphism. The cells are arranged in diffuse sheets, aggregates and singly scattered. Bizzare appearing multinucleated, floret cells are seen. Some of the cells show abundant vacuolated cytoplasm and centrally located nuclei. Focally lipoblasts like cells are seen. Mitotic figures are frequent (1-3/HPF). Foci of necrosis and hemorrhages are also present (H&E).

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