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Research Article

A RARE CASE OF RHABDOMYOSARCOMA PRESENTING AS AN INTRADURAL SPINAL TUMOR

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Abstract

Spinal cord compression is a very serious complication of malignancy in adults. A Sarcoma presenting with an epiduralmass, and spinal cord compression is very rare. We report a case of a 29 year old female patient who presented to us with para-spinal and epidural rhabdomyosarcoma (RMS) causingspinal cord compression and a complete brachial plexopathy. She came with a 15 month history of right upper limb pain and weakness, neck mass associated with neck pain and a 2 week history of bilateral lower limb weakness associated with urine and stool incontinence. Imaging studies showed a large lobulated intra-dural and extra-medullary enhancing dumbbell-shaped tumor at C6/C7 levels compressing the spinal cord at that level .There was a right supraclavicular mass with central necrosis and involving the exiting regions of the spine. There was a syrinx in the spinal cord above the level of the lession extending up-to C2. The patient was taken for laminectomy and durotomy and excision of the intra-dural portion of the tumor. Histology confirmed the diagnosis of rhabdomyosarcoma. After surgery there was improvement in signs and symptoms. Unfortunately she died while still waiting for the second stage of the operation to excise the extra-spinal tumor component.

Keywords: Rhabdomyosarcoma, Extradural Spinal Tumor, Dumbbell, Spinal Cord Compression

INTRODUCTION

Rhabdomyosarcoma is a highly aggressive and rapidly growing sarcoma with skeletal muscle origin that occasionally appears in the spinal epidural space (Kim *et al.,* 2000). 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 and complications of this tumor. Spinal tumors comprise 15% of all CNS tumors. Their annual incidence is 2-10 per 100000. Ninety percent of these patients are older than 20 years. Most common spinal tumor location is extradural (55-60%), where cancer metastasis to spine leads the way. Primary vertebral bone tumors are less frequent extradural spinal tumors. Extramedullary, intra-dural spinal tumors (EISTs) are rare. They comprise about 40-45% of all spinal tumors. The fourth to sixth decades of life represent the peak incidence of occurrence. Men and women are equally affected. Spinal cord compression can occur in rhabdomyosarcoma. It can be due to direct spread of the tumor, spread to the epidural space by infiltration of the vertebral foramina or metastatic vertebral bone disease with cord compression. Most intra-dural extramedullary tumors are benign, and they exhibit no specific symptoms. Radicular pain and worsening sensory and motor loss are common manifestations. Patients may have bladder or bowel complaints. Therefore, most of the patients are wrongly diagnosed with cervical spondylopathy or intervertebral disk herniation. MRI is very crucial to confirm the diagnosis of intra-dural extramedullary tumors. Once the diagnosis is confirmed, the best treatment for nonmalignant intra-dural extramedullary tumor is surgery. The goal of surgery is complete surgical resection while preserving spinal stability, without worsening the preoperative neurological status.

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CASE REPORT

We report a case of a 29 year old female patient who presented to us with a 15 month history of right upper limb pain and weakness, neck mass associated with neck pain and a 2 week history of bilateral lower limb weakness associated with urine and stool incontinence. She was previously well until about a year and three months prior when she started experiencing right sided neck pain mainly at night .The pain would radiate to her right upper limb particularly her right thumb and middle finger .This was then accompanied by progressive numbness and weakness of the same limb until about 2 months after onset of symptoms that she became completely paralysed in that limb. This was followed by gradual onset painless weakness and paraesthesia of the left upper limb.2 weeks prior to presentation she developed lower limb paraesthesia and weakness associated with bowel and bladder dysfunction (incontinence). She developed dyspnea as well around the same time. There was no history of trauma or any known malignancies. She did not have any history of TB contact and was HIV negative. On examination patient was ill looking, with a moderate respiratory distress (respiratory rate of 25 breaths per minute, intercostal and supraclavicular recessions), paradoxical breathing. She was pale and wasted. There was a right sided mildly tender supraclavicular firm mass which was 8 x 6 cm in its widest diameters, immobile and attached to underlying structures. Her LOC was 15/15,all cranial nerves were intact and her eye examination was unremarkable.

Upper limb examination

Right upper limb muscles were wasted and had visible unprovoked fasciculations. Sensory level was C5 on the right and C6 on the left. There was arreflexia in the right upper limb and hyperreflexia in the left upper limb. Proprioception was lost in the right upper limb.

	Right	Left
C5	0/5	5/5
C6	0/5	5/5
C7	0/5	2/5
C8	0/5	1/5
TI	0/5	1/5
Tone	Hypotonia	Hyperetonia

Lower limb examination. Power was 0/5 across all myotomes. There was bilateral hypertonia and hyperreflexia. Perianal sensation was absent. Chest examination showed reduced air entry on the right hemi-thorax and reduced chest excursions on the same side. Examination of the back revealed a large grade 4 necrotic sacral pressure ulcer.

Full blood count showed a WCC-13,0..HB-9,2.MCV-98,2.PLT-311.

Urea and electrolytes which were normal NA-141..K-4,4..U-5,0..CR-46,7

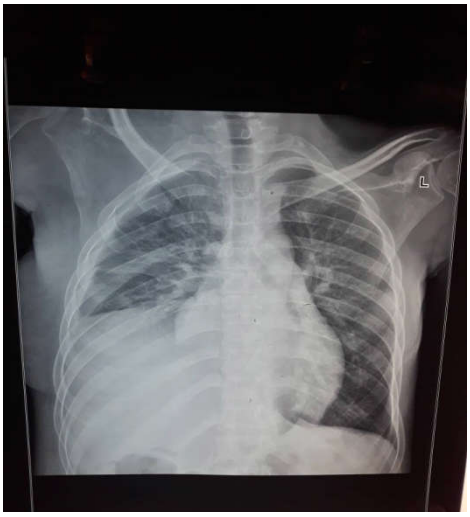

Fig. 1.Chest x-ray showing elevation of the right hemi-diaphragm indicating right phrenic nerve palsy

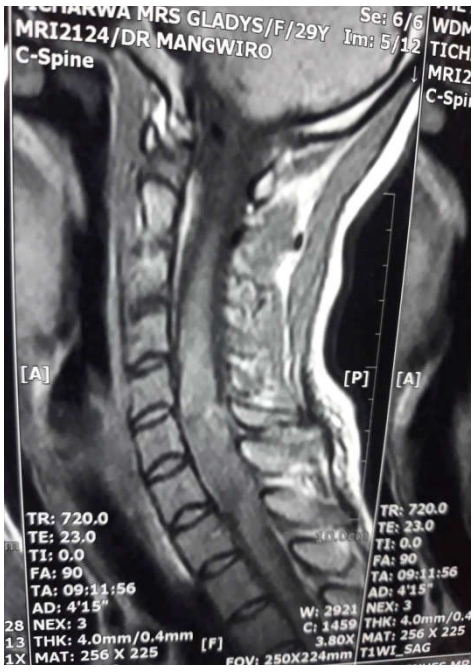

Fig 2. T1-W MRI scan showing an enlarged spinal cord at c5-c7 with signal change within the cord

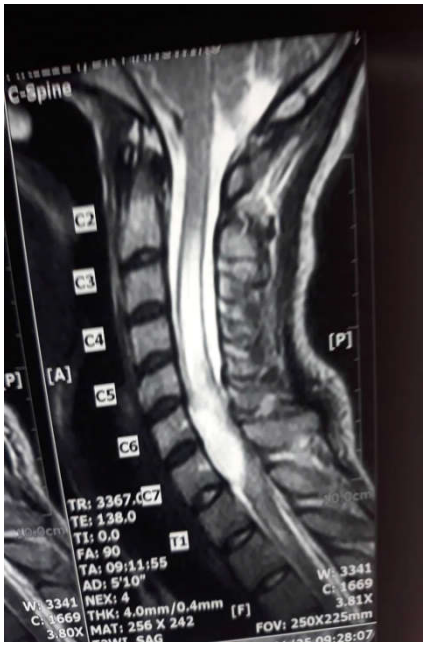

Fig 3. T2W MRI showing the Hyperintense tumor at C6/C7 compressing the spinal cord at that level with extra-spinal extension and Syringomyelia up to the level of C2

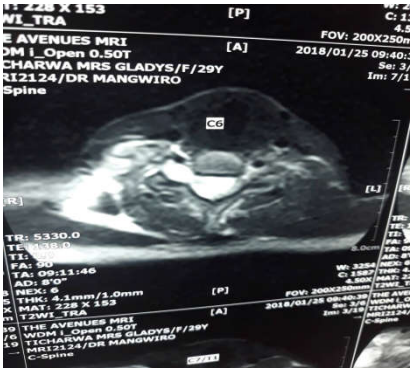

Fig 4. AXIAL T2 MRI showing a dumbbell tumor, compressing and displacing the spinal cord to the left. Also evident is the extra-spinal soft tissue component of the tumor

MRI showed a large lobulated intra and extramedullary enhancing lession, dumbbell-shaped at C6/C7 levels compressing the spinal cord at that level with a syrinx in the spinal cord above the level of the lession extending to C2.There is a right supraclavicular mass with central necrosis and involving the exiting regions of the spine. Our working differential included a schwannomaand a neurofibroma. Patient was put on intravenous antibiotics, steroids, analgesia and commenced on venous thromboembolism prophylaxis. Laminectomy and excision of the intra-spinal tumor was done to decompress the spinal cord. Second stage in conjunction with the head and neck surgeons was going to be scheduled to do neck dissection and excise the extra-spinal component of the tumor. Intra-operatively, patient was put in a prone positionand a midline skin incision done over the spinous processes of C4-T1.Sub-periosteal dissection was done to expose the laminae. Laminectomy was done at C5-C7. Midline vertical durotomy was done and a tan greyish gelatinous tumor was identified on the right side of the spinal cord displacing the cord to the left side, postero-laterally. Tumor-spinal cord interface identified and intra-dural tumor completely excised until exiting nerve roots were freed and visible. Cord visibly started to pulsate soon after decompression.

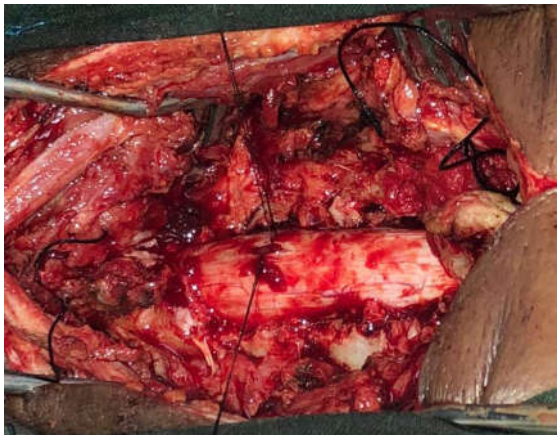

Fig. 5.Laminectomy done,spinal cord visible and durotomy being done

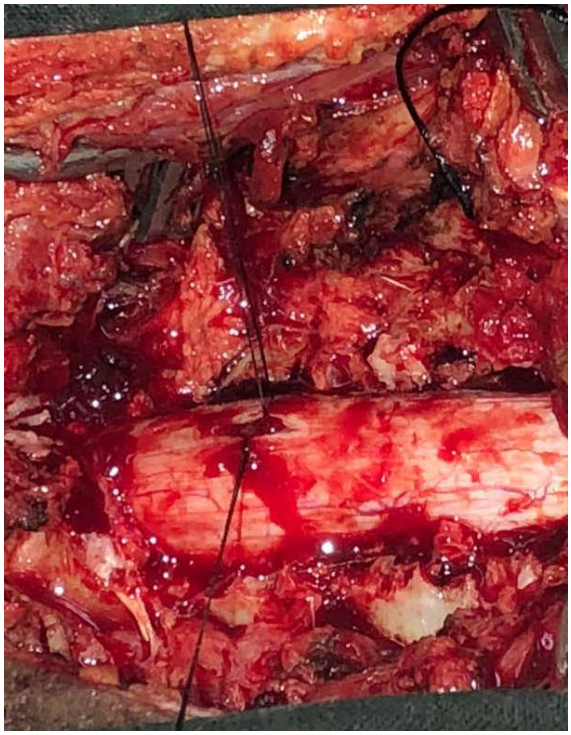

Fig.6.Durotomy

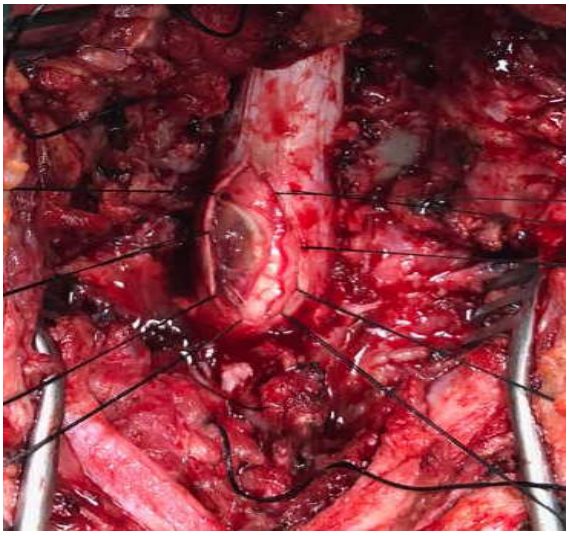

Fig.7.Durotomy done, dark greyish intra-dural extramedullary tumor visible, compressing the spinal cord which is seen as a squashed pinkish tissue

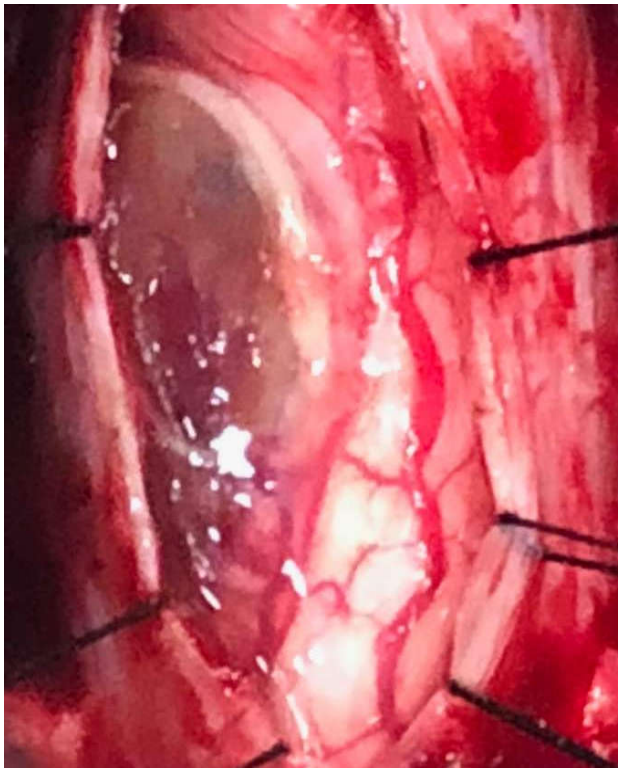

Fig.8.Tumor – spinal cord interface clearly visible

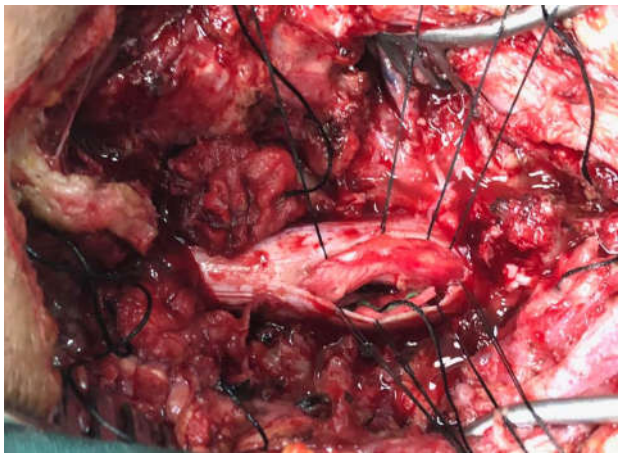

Fig. 9. Tumor excision completed

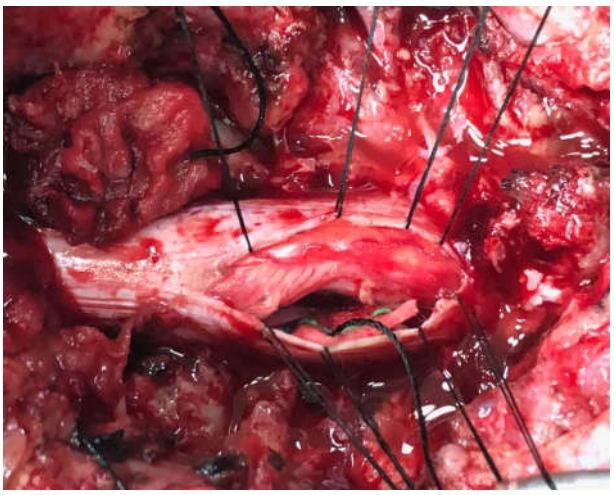

Fig. 10. Normal spinal cord showing areas where tumor was indenting and compressing it

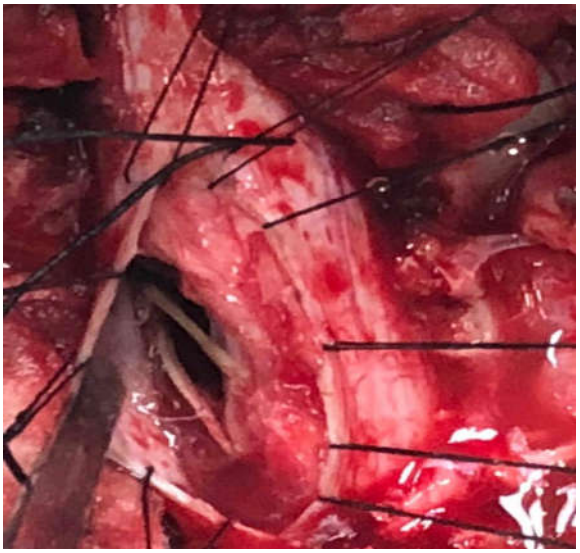

Fig 11.Exiting nerve roots completely freed from the tumor

Postoperatively

Patient stayed in HDU for a few days as she was having episodic respiratory distress. She was continued on physiotherapy, analgesia, steroids. 3 weeks post operatively she showed signs of gaining power in the left upper limb. She however died while awaiting second stage of the procedure where we were planning to do neck dissection for excision of the extra-spinal component of the tumor.

Histopathology

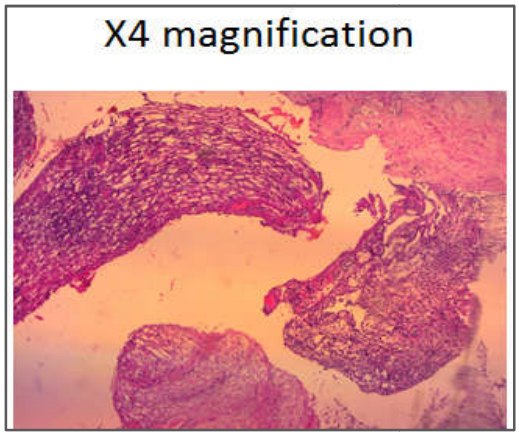

Fig. 12

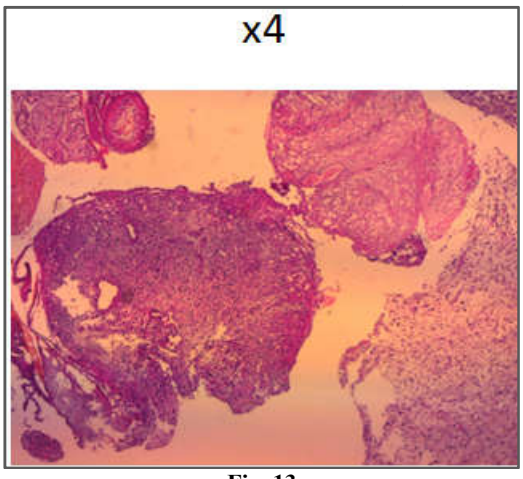

Fig. 13

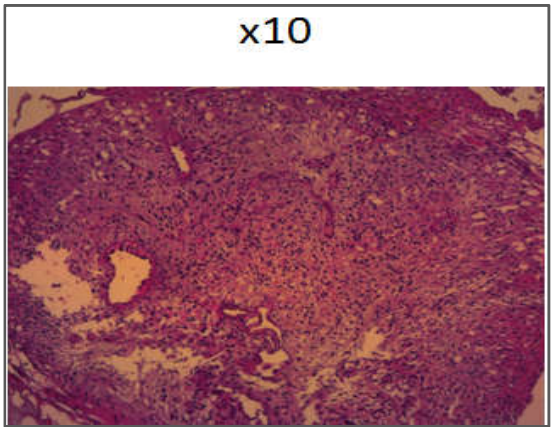

Fig. 14

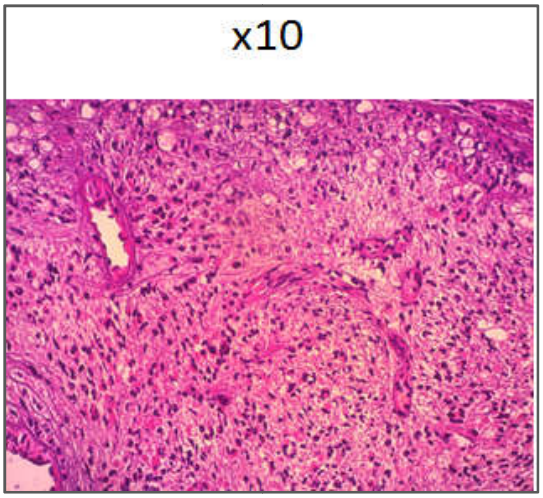

Fig. 15

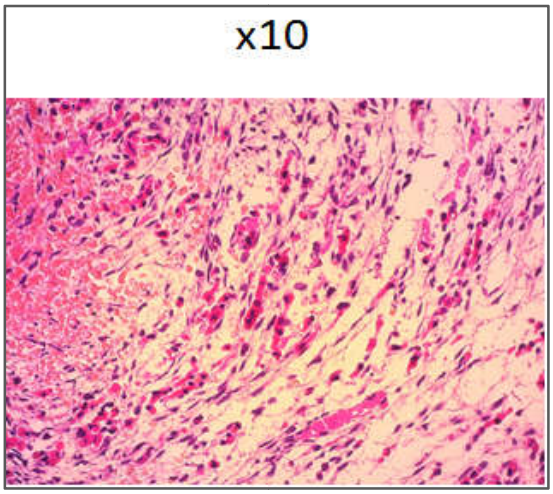

Fig. 16

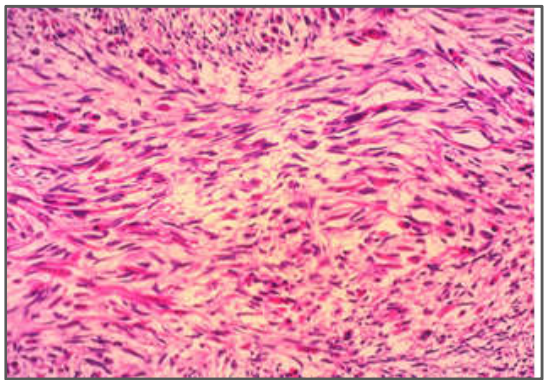

Fig. 17

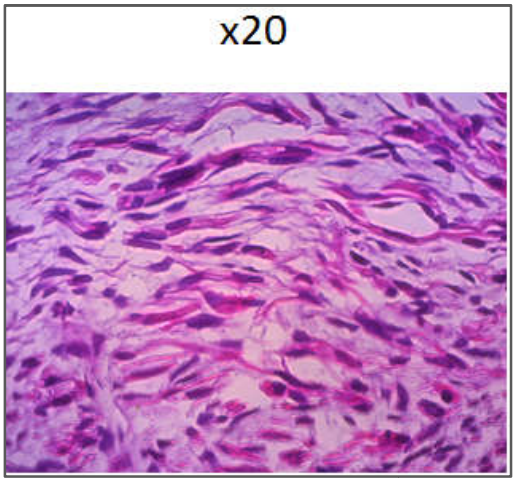

Fig. 18

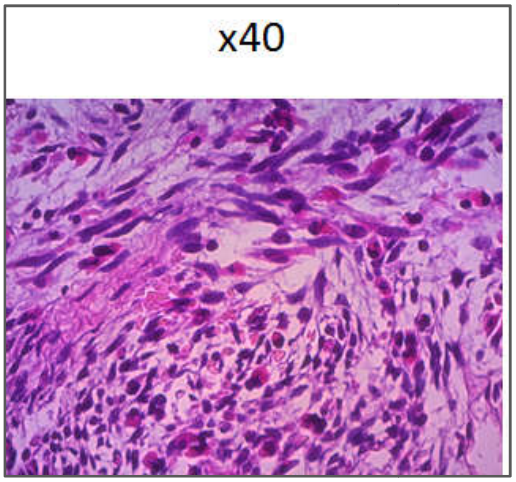

Fig. 19

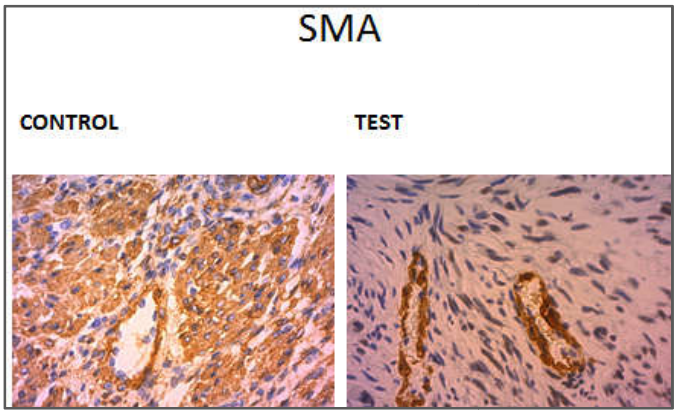

Fig.20

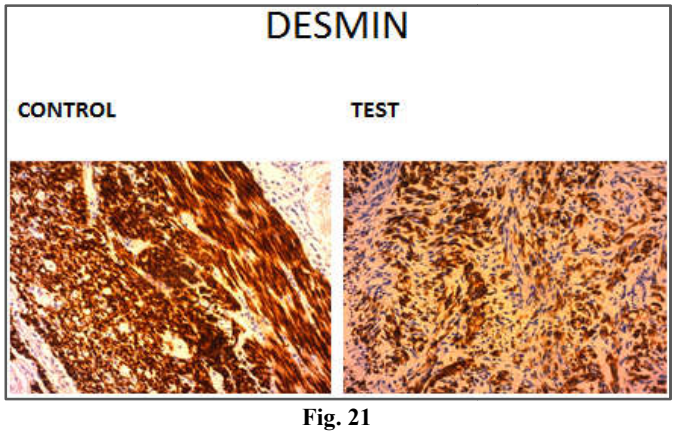

Fig. 21

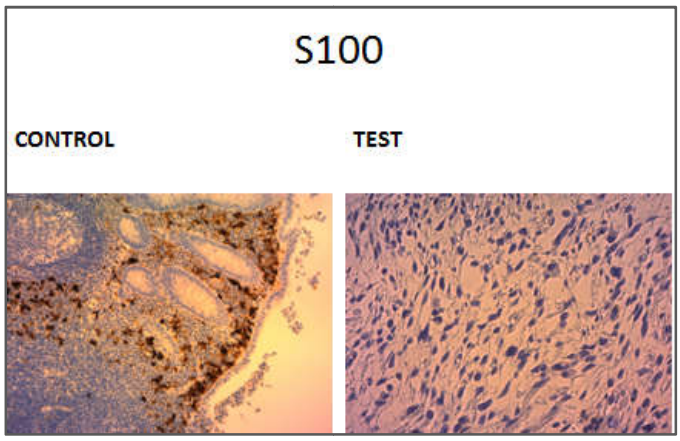

Fig.22.

Fig.12-22.The sections show a tumor composed of spindled and stellate cells in a myxoid stroma. The cells show eosinophilic cytoplasma. There is positivity for the immunomarkersdesmin and smooth muscle actin. The appearances are those of a rhabdomyosarcoma

DISCUSSION

Rhabdomyosarcoma is the most common soft tissue sarcoma of the pediatric age group accounting for 5-8% of childhood cancer (Kim *et al.,* 2000). 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 (Miller *et al.,* 1995). This is different to our case where presentation was in an adult. There is a slight male to female predominance (5:3) and no known predilection for race (Shields and Shields, 2003). Forty percent of RMS arise from the head and neck region; around 15% are genitourinary (GU) non-bladder prostate (BP) tumors (i.e., para-testicular, vaginal and uterine tumors), 10% are BP tumors, 15% occur in the limbs, and 20% occur in other sites (i.e., thoracic or abdominal tumors).RMS occurs sporadically and no predisposing or risk factors have been recognized in the majority of cases (Xu *et al.,* 2000). Rhabdomyosarcoma is a fast-growing, primitive, high-grade, malignant mesenchymal tumor. It is thought that RMS arises as a consequence of regulatory disruption of skeletal muscle progenitor cell growth and differentiation (Merlino and Helman, 1999). The disease has been associated with familial syndromes, including Li-Fraumeni syndrome, neurofibromatosis type 1, and hypo-melanosis of Ito (Fountas *et al.,* 2005). RMS has also been reported to occur as a congenital tumor. Congenital dysrrhaphic malformation of the spine may predispose children to intra-lessional development of RMS (Wagner and Koch, 2004). 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 (Van Rijn *et al.,* 2008). Alveolar RMS is associated with a significantly higher risk of relapse and a much higher risk of metastasis (Van Rijn *et al.,* 2008). It presents in a wide range of histologic types. There are various patterns of spread of tumor. Histologically, it contains a mixture of rhabdomyoblasts, which are recognized by their typical cross-striations, and undifferentiated cells. Most extremity tumors are alveolar or undifferentiated histologic types, unlike the embryonal or botryoid types found in the face and neck and GU system. Prognosis is bad for patients with

RMS of the extremities than for those with tumors arising from the GU system or the head and neck region. In the extremities, the tumors are deep and tend to spread along fascial planes. Immuno-histochemical detection of muscle-specific protein expression, including staining for a-actin, myosin heavy chain, desmin, myoglobin, Z-band protein and MyoD, is useful in confirming the diagnosis. These tumors present with a wide range of symptoms, depending upon the location of the primary tumor, but in general presenting symptoms often are indolent, with nonspecific symptoms. Limb and trunk tumors often present with a painless swelling and/or with enlarged regional lymph nodes. In patients presenting with metastases more general symptoms of fatigue, weight loss, and low blood counts are noted (Van Rijn *et al.,* 2008). Imaging of the primary site with radiography, ultrasound(US), MRI or computed tomography (CT) scan is required in all patients as an initial investigation (Brisse *et al.,* 2005). Once the diagnosis has been confirmed histologically, the most frequently involved metastatic sites should be investigated; CT of the lungs, imaging of regional lymph nodes, and a technetium bone scan are recommended in every patient. This malignant tumor invades local structures and metastasizes to remote sites by lymphatic and hematogenous spread. Children with metastatic RMS have poor survival rates (Sandler *et al.,* 2001). The most common metastatic sites from primary rhabdomyosarcoma are lung, bone, bone marrow and liver.

Imaging findings

Radiography does play a significant role in its diagnosis. Localized bony erosion adjacent to the primary site can be seen. This area may show increased uptake on technetium-99 m methylene diphosphonate (99 mTc-MDP) bone scan in the absence of metastatic disease in the skeleton (Sebire *et al.,* 2006) USG is usually the first imaging investigation that is done in children with soft-tissue masses due to its easy availability. Lesion characterization can be done; its extent and vascularity of a mass can be assessed. On US, RMS in general is seen as a well-defined, slightly hypoechoic inhomogeneous mass that can show significantly increased flow. In all other RMS locations, additional imaging using CT or MRI is needed. US can be useful in performing image guided biopsies (Kim *et al.,* 2000). MRI is the imaging modality of choice for diagnosis of RMS due to its excellent soft tissue contrast resolution. The imaging characteristics of RMS are generally nonspecific. They have intermediate signal intensity onT1WI.

On T2WI, they tend to be of intermmediate-to-high signal intensity. If the tumor contains a high number of septa, it may present as a lobulated lesion. These tumors show strong enhancement on post-contrast MRI imaging due to their highly vascular nature. In very rare instances, the tumor may show a predominantly cystic appearance (Sebire *et al.,* 2006). Dynamic series are useful in order to assess tumor vascularity and to differentiate between post-chemotherapy/surgery residual disease and fibrosis (Enneking *et al.,* 2003). Vascular involvement is considered to be absent if there is a normal tissue plane visible between the tumor and the vessel, or if the tumor has a <180° circumferential relationship. If the tumor surrounds the vessel for more than 180°, it is considered to be encased (Kim *et al.,* 2000). Positron emission tomography-CT (PETCT) evaluates uptake in metabolically active malignant cells, with the help of fluorine-18 fluoro-deoxyglucose. The combination of PET with CT has a higher accuracy for the depiction of pulmonary metastases (McHugh *et al.,* 2005). Differentiating RMS from other malignant tumors is difficult. Few differentials that should be considered are Ewing sarcoma, peripheral neuroectodermal tumors (PNET), infantile fibrosarcoma, and desmoplastic small round-cell tumors soft-tissue tumors in childhood. 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 Meningioma and immunohistochemical study is mandatory for a definitive diagnosis (Ozawa *et al.,* 2007). 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 (Hayes-Jordan and Andrassy, 2009). Using this multimodality approach, the cure rates for RMS have steadily increased from only 25% in the 1970s to 70% in the 1990s. Goal of surgery is excision of the epidural tumor with preservation of neurological function (Van Rijn *et al.,* 2008). Prognosis depends on the age of the patient, extent of the tumor, tumor histology, and presence or absence of metastasis (Pappo *et al.,* 1995).

Conclusion

Primary spinal epidural RMS is an extremely rare tumor especially in adults 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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