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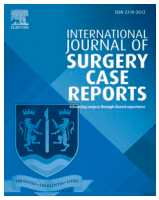

Case report

Clear cell sarcoma at D12-L1 paraspinal region: A reported case and review of the literature

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ABSTRACT

Background: Clear cell sarcoma is a very rare malignant tumor originating from neural crest cells, the tumor most commonly affects the lower limbs but there have been few cases reported to affect the trunk and para spinal area. Patients with Para spinal sarcoma have a variety of clinical presentations and a variable prognosis depending on several prognostic factors including tumor size.

Case presentation: A 14-year-old male patient presented with paraplegia for one month duration and he was diagnosed later on to have a paraspinal clear cell sarcoma at D12-L1 level. A wide local excision was made and patient was referred for oncology treatment. However, the patient came back to the same hospital after two months due to the recurrence of the tumor at the same site.

Conclusion: Clear cell sarcoma, although rare, can affect the spine and present with weakness, paraplegia and backache. It also affects people from all age groups and can have a high metastatic rate and a high recurrence rate.

1. Introduction

Clear cell sarcoma is a very rare malignant tumor originating from neural crest cells which is often described as 'malignant melanoma of soft tissue' due to the similar histology and lack of cutaneous melanoma pigmentation [1]. It is estimated that clear cell sarcoma accounts for 1% of soft tissue sarcomas [2,3] and the average age of diagnosis is 39 years with an equal distribution among males and females [2] but seems to be higher in Caucasians than in black people [2].

The tumor has a higher tendency for lower extremities soft tissues near the fascia, tendon or aponeuroses [4] with 15% of patients presenting with Metastasis most commonly affecting the lungs with an overall 5 years survival estimated to be 50% [2].

Herein, we report a rare case of a 14 years adolescent with a clear cell sarcoma originating from the D12-L1 region of the paraspinal area.

To the best of our knowledge, this is the first case report for clear cell sarcoma presented at the left D12-L1 region that has been documented in English literature worldwide.

2. Case presentation

A 14-year-old male patient presented to Al-Makassed Hospital, Jerusalem complaining of paraplegia for one month prior to admission. The patient's history started two months prior to admission when he started to complain of low back pain that radiates to his left lower limb.

After that, the patient suddenly became paraplegic according to the family. There is a history for urinary and stool retention that started few years ago according to the mother. However, past medical and surgical history is otherwise unremarkable.

On physical examination, the patient was conscious, alert, oriented to place, time and person. Furthermore, the patient had intact cranial nerves with 5/5 for power in the upper limbs, intact sensation and proprioception and lower limb paraplegia.

Whole spine MRI (Fig. 1a, b) showed a huge left D12-L1 space occupying lesion extending to the left D12-L1 foramen and left paraspinal area.

The differential diagnosis and possible treatment were discussed with the patient and his family including the surgery complications,

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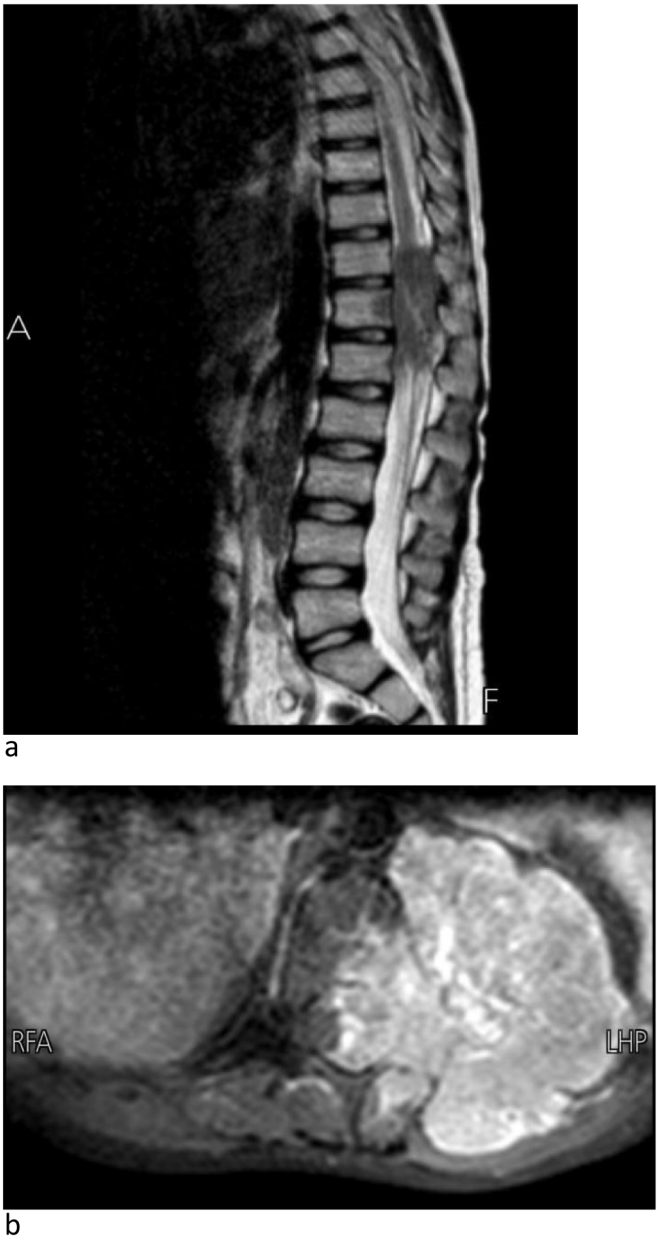

Fig. 1. a: T2 sagittal view MRI of dorso-lumbar spine showing a hypointense lesion involving the spinal area of D12-L1 mainly.
b: T1 axial view MRI at the same level as the above sagittal view presented in a showing a huge paraspinal lesion causing complete cord syndrome extending to the retro-peritoneal cavity and invading the pleural cavity.

spinal instability and the possible need for stabilization.

Next day, the patient underwent microscopic left D12-L1 hemi-laminectomy for resection of left D12-L1 spinal tumor that extended to left D12-L1 foramen and left paraspinal area. The tumor was highly vascular. Good cord and root recompression was done and surgery was enough to remove the entire lesion.

The surgery went well with no intraoperative complications. The patient had stable vital signs post operation and was moving his upper limbs freely. However, lower limbs were paraplegic with intact sensation and proprioception.

Dorso-lumbar CT-scan and MRI second day post operation showed complete resection of the tumor at level D12-L1 with decompression of the cord and minimal left side effusion (Fig. 2a, b).

Physical examination on the 3rd day post-operation revealed abdominal distention and was diagnosed by pediatric surgeons to have

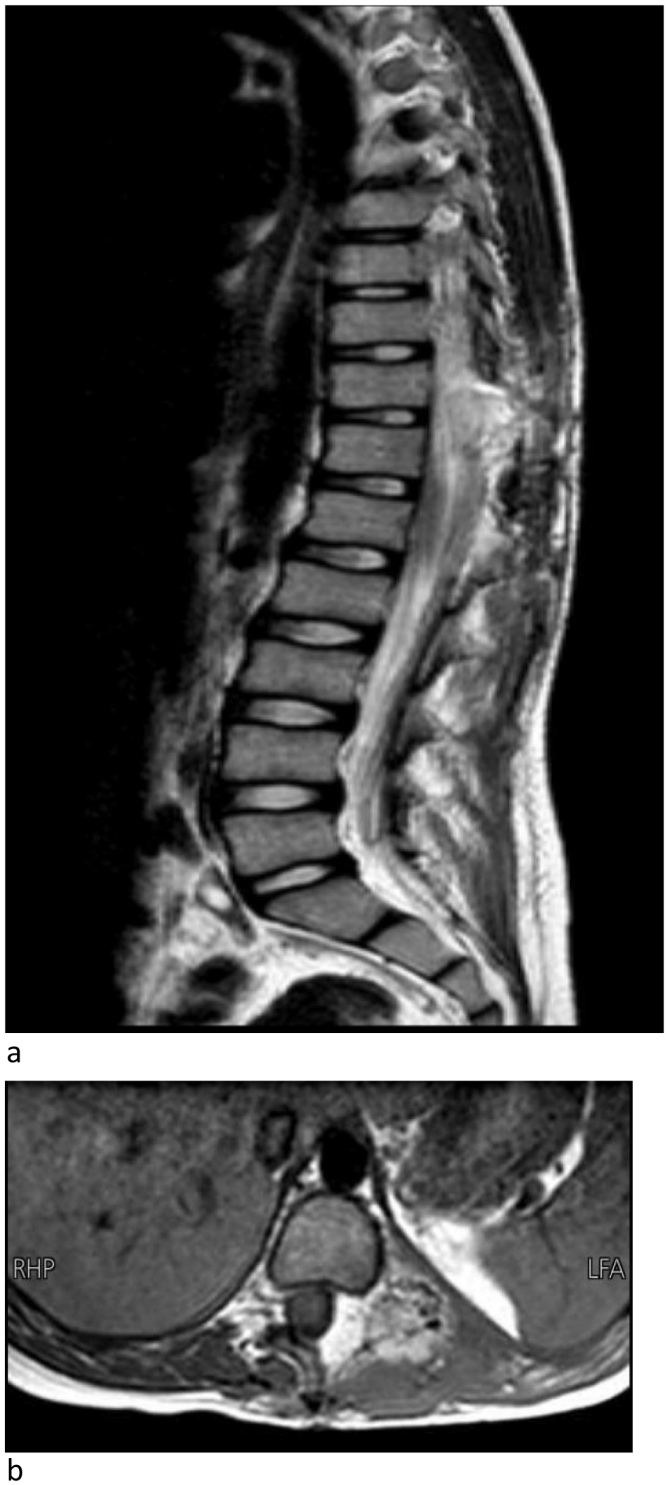

Fig. 2. a: T2 sagittal view MRI for the dorso-lumbar spinal area showing complete resection of the tumor at level D12-L1 with decompression of the cord.
b: T1 axial view MRI at the same level seen in a showing a total excision of the tumor with spinal cord release.

paralytic ileus and was kept in NPO (nil per os), However, glycerin supplements were added regularly and the patient passed stool after that and his abdominal distention was relieved. At that time, the patient was seen pale during the routine follow up at our unit. Complete blood count was done and revealed a hemoglobin value of 7.9 g/dL, so he was given two units of packed red blood cells and the patient improved upon that.

On the 7th day post operation, the patient complained of lower abdominal pain, an ultrasound was done and showed an over distended bladder with over 500 cm^3 volume therefore a Foley catheter was inserted and the abdominal pain improved.

On the 8th day, whole spine MRI showed an enhancing area at the left spinal canal involving D12 with displacement of the spinal canal to the right.

After 2 weeks of hospital stay, the patient was discharged to continue oncology management at a highly specialized center with a thoracolumar corset prescribed.

2.1. Histopathology

The tumor shows infiltration into the tendons and aponeuroses. The tumor is highly infiltrative and organized in a haphazard arrangement into small compact nests and fascicles of uniform neoplastic cells dissecting along the dense fibrous connective tissue of tendons, fascia, and aponeuroses. The stroma may be barely visible, fibrotic, or hyalinized.

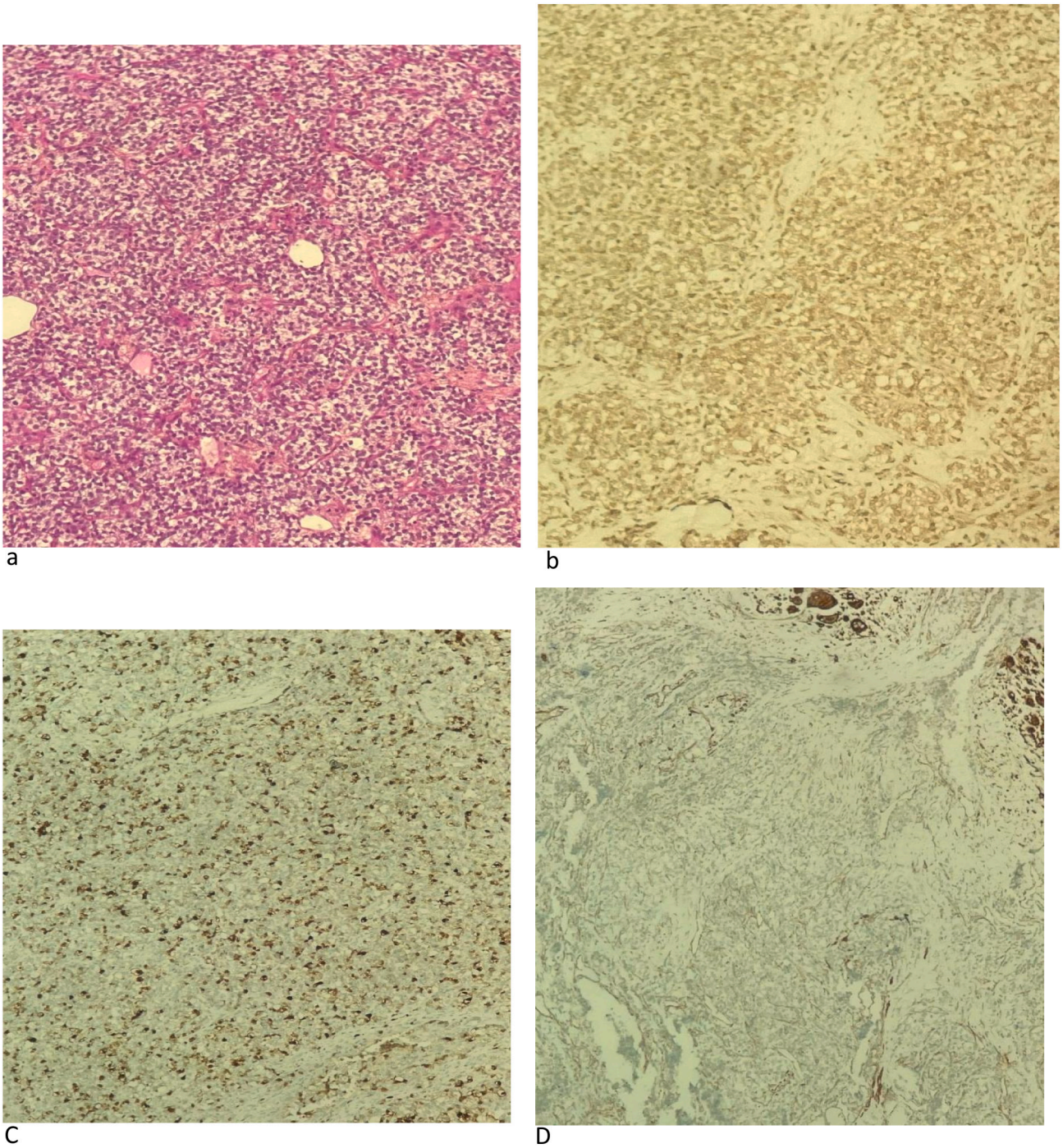

Fig. 3. a: Hematoxylin and eosin stain shows a tumor with nested growth pattern with mixture of spindle, epithelioid and tumor giant cells. The tumor cells have lightly eosinophilic or clear cytoplasm. Cytoplasmic melanin is focally noted.
b: Shows positive HMB45 immunostain.
c: S100 positive immunostaining.
d: Shows negative desmin immunostaining.

These cellular clusters or aggregates are divided into lobules by a fine collagenous framework of variable thickness that can be highlighted with a Masson trichrome stain. The neoplastic cells are polygonal to fusiform with clear or pale eosinophilic cytoplasm and centrally located round to ovoid vesicular nuclei that show prominent basophilic nucleoli (Fig. 3a). The clear cell appearance is due to the accumulation of glycogen as can be demonstrated with periodic acid–Schiff (PAS) and PAS with diastase (PAS-D) stains. The neoplastic cells show no or minimal pleomorphism. There is a paucity of mitotic figures that is in concordance with the slow-growing behavior of the tumor, although the course of this tumor is aggressive. Scattered multinucleated tumor giant cells are commonly present and areas of focal necrosis may be noted. Variants have been described including a case of myxoid clear cell sarcoma, a case with an alveolar pattern of growth because of loss of cellular cohesion, and 2 cases with a microcystic pattern. There should be no evidence of biphasic differentiation.

Immunohistochemical studies of clear cell sarcoma show that the tumor cells express antigens associated with melanin synthesis including diffuse cytoplasmic immunoreactivity with HMB-45 (Fig. 3b), nuclear and cytoplasmic immunoreactivity to S100 protein (Fig. 3c). Cytokeratin (CK), epithelial membrane antigen, carcinoembryonic antigen, desmin, and smooth muscle actin are negative (Fig. 3d).

2.2. Follow-up

The patient was discharged from our hospital going back to his town in a good and stable condition. He started rehabilitation and physiotherapy immediately and mild, but noticeable, improvement has been encountered as the patient and his mother claim. The improvement was in the form of moving the toes bilaterally spontaneously with retained sensations. Moreover, there was an improvement in the defecation process as the patient was not in need to use suppositories for a further time. However, due to experiencing some clashes and events after the patient came back to his town and inability to start the oncology management soon after the surgery, he started to complain of mid back pain two-months later with deterioration in the movement of his toes along with sensory loss and stool impaction. Then, dorso-lumbar MRI scan was done and showed a huge recurrent tumor almost at the same site as the first time with some extension into L2 level.

The patient was therefore referred to our hospital and excision of the tumor was successfully done. However, the surgery was complicated with blood loss of about 2000 mL. 500 cm^3 of blood were given intra operatively, and hemovac and lumbar drains were inserted. Then, the patient was moved into the neurosurgical intensive care unit (ICU) and kept under close observation. His vital signs were stable except for tachycardia, complete blood count was ordered and hemoglobin was seen 6. Therefore, 2 units of PRBCs were transfused. His vital signs improved after that, and hemoglobin approached 10.5.

The patient has shown improvement on a daily basis. He was transferred out of ICU to the ward after three days. The drains were removed and he was tolerating oral feeding. The wound at the site of surgery was seen dry and clean. However, none has been noticed regarding motor or sensory functions in the lower limbs during his stay at our unit.

After 6 days of the surgery, the patient was immediately transferred to a nearby hospital to schedule regular sessions of radiotherapy soon.

The patient was upset about the recurrence of tumor but expressed his gratitude to the medical team.

3. Discussion

Clear cell Sarcoma is a rare tumor affecting most commonly Caucasians in their third or fourth decade of life [2,3,5].

In clear cell sarcoma, the translocation t (12;22) (q13; q12) has been reported to present in most of the cases. Thus molecular studies for the EWS/ATF1 or EWS/CREB1 fusions are used for diagnosis [3]. Moreover, polysomy of chromosome 8 was also reported to present with clear cell sarcoma [3].

The tumor affects most commonly the lower limbs [2,3] and presents with a slow-growing mass that may be accompanied by pain and tenderness in the region in around half of the cases [2,6]. Several unusual sites were affected by the tumor such as chest wall and scapula [7,8].

However, there are six cases reported to originate in the para spinal region [9]. Reported cases of tumor in the para spinal region presented with tingling and numbness in the mid upper thoracic spine, severe pain in the left subcostal region radiating to the left upper quadrant, painless upper thoracic back mass or even backache with progressive weakness of bilateral lower limbs [9]. The mean age of cases reported to have a paraspinal clear cell sarcoma is 25 [9] and most of them are young and less than 40 years. Here however, our patient is a 14 years old Arab adolescent presenting with a paraplegia of the lower limbs which makes it the first case to be reported to present as a paraplegia.

Reported cases of paraspinal CCS affected most commonly D4 level (which was reported in three cases) and the levels reported range from D3 to D6 [9]. Thus, our case has a new and different level of tumor location (D12-L1).

Clear cell sarcoma appears hypointense on T2 weighted image MRI and hyperintense on T1 weighted image MRI since the tumor contains melanin which has paramagnetic properties [10] which was also noted in the case. Furthermore, positive HMB-45 has a significant effect on variant signal intensities on T1 weighted image [10]. Our case had HMB-45 positive immunostaining.

Immunohistochemical studies of clear cell sarcoma show that the tumor cells express antigens associated with melanin synthesis including HMB-45 and S100 protein which are helpful to distinguish clear cell sarcoma from other tumors such as synovial sarcoma and epithelial tumors. Our case, however, was stained positive for both HMB-45 and S100.

Differential diagnosis for clear cell sarcoma includes melanoma, malignant blue nevus, perivascular epithelioid cell tumor, epithelioid leiomyosarcoma, paraganglioma-like dermal melanocytic tumor, malignant peripheral nerve sheath tumor. Actin and desmin are negative in clear cell sarcoma which helps in distinguishing it from PEComa and epithelioid leiomyosarcoma. In addition, clear cell sarcoma has infrequent mitosis and nuclear pleomorphism in contrast to melanoma and malignant peripheral nerve sheath tumor.

The mainstay of treatment of clear cell sarcoma is wide surgical excision with or without radiotherapy [3,9].

Clear cell sarcoma has high rate of local recurrence ranging up to 84% with a high rate or late metastasis which can occur even after 29 years [3]. Several prognostic factors have been established including the site and size of the tumor, presence of necrosis, resected margins and DNA content [3]. Tumors more than 5 cm are more likely to metastasize [3].

Our patient had local recurrence two months after the wide local excision.

4. Conclusion

Herein we present the first case of CCS in the thoracolumbar region (D12-L1) and the sixth case of paraspinal CCS. It has been reported that CCS can affect different parts of the body. CCS has a bad prognosis and a high recurrence rate.

5. Methods

This case report has been reported in line with SCARE 2020 guidelines [11].

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Ethical approval

The study is exempt from ethnical approval in our institution.

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Declaration of competing interest

The authors have no conflict of interests to declare.

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