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ADULT GIANT RETROPERITONEAL PLEOMORPHIC RHABDOMYOSARCOMA.

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ABSTRACT

Rhabdomyosarcoma is a malignant tumour of mesenchymal origin. Unlike in childhood and adolescence, rhabdomyosarcoma is an infrequent occurrence in adulthood, with most cases in the adult population affecting the extremities. The prognosis for rhabdomyosarcoma in adults is generally poor due to aggressive tumour behaviour and poor response to adjuvant chemoradiotherapy. We discussed the case of a 38-year-old gentleman presenting with a progressively enlarging abdominal mass and was diagnosed with pleomorphic rhabdomyosarcoma of the psoas muscle on ultrasound guided biopsy of the mass. He underwent complete resection of the primary tumour and two other retroperitoneal secondaries embedded in the sigmoid mesocolon. A third secondary mass was found to be stuck on to the abdominal aorta and unfortunately could only be partially debulked. Post-operatively the patient received adjuvant radiotherapy but refused chemotherapy. He subsequently deteriorated and passed away 3 months after surgery. We also elaborate further on the signs and symptoms of rhabdomyosarcoma, its investigation and various aspects of its treatment.

Keywords: Adult, Case report, Psoas muscle, Pleomorphic Rhabdomyosarcoma, Retroperitoneum.

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ABSTRACT

Rhabdomyosarcoma is a malignant tumour of mesenchymal origin. Unlike in childhood and adolescence, rhabdomyosarcoma is an infrequent occurrence in adulthood, with most cases in the adult population affecting the extremities. The prognosis for rhabdomyosarcoma in adults is generally poor due to aggressive tumour behaviour and poor response to adjuvant chemoradiotherapy. We discussed the case of a 38-year-old gentleman presenting with a progressively enlarging abdominal mass and was diagnosed with pleomorphic rhabdomyosarcoma of the psoas muscle on ultrasound guided biopsy of the mass. He underwent complete resection of the primary tumour and two other retroperitoneal secondaries embedded in the sigmoid mesocolon. A third secondary mass was found to be stuck on to the abdominal aorta and unfortunately could only be partially debulked. Post-operatively the patient received adjuvant radiotherapy but refused chemotherapy. He subsequently deteriorated and passed away 3 months after surgery. We also elaborate further on the signs and symptoms of rhabdomyosarcoma, its investigation and various aspects of its treatment.

Keywords: Adult, Case report, Psoas muscle, Pleomorphic Rhabdomyosarcoma, Retroperitoneum.

INTRODUCTION

Adult rhabdomyosarcoma is a rare occurrence, accounting for less than 1% of solid tumour malignancies in the adult population.[1] The pleomorphic subtype is one of the two most common subtypes encountered in this age bracket (the other one being rhabdomyosarcoma not otherwise specified), unlike the other subtypes of rhabdomyosarcoma such as embryonal and alveolar rhabdomyosarcomata which are more common in the paediatric and adolescent population.[1] In the adult subpopulation, rhabdomyosarcoma usually affects the extremities.[2] This report demonstrates an atypical case of adult retroperitoneal pleomorphic rhabdomyosarcoma of the psoas muscle, and elaborates further on the presentation, investigation and various aspects of management of pleomorphic rhabdomyosarcoma in adults.

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CASE REPORT

A 38-year-old male with no known medical illness presents with a 5-month history of a progressively enlarging abdominal mass with early satiety and weight loss of 30kg. One month prior, he developed intermittent crampy abdominal pain, especially over the left iliac fossa with a pain severity score of 5/10. This was associated with anorexia and

generalized lethargy. He had no significant bowel or urinary symptoms. Abdominal examination revealed a firm to hard abdominal mass occupying the left side, approximately 10 cm x 20 cm in size with a smooth surface and a regular margin. It was mobile in the horizontal plane and non-tender. Rectal examination was unremarkable. There were no associated palpable superficial lymph nodes.

Computed tomography (CT) imaging of the abdomen and pelvis showed a large lobulated heterogeneously enhancing hypodense mass at the left retroperitoneal region measuring 20.5 cm x 23.3 cm x 2.5 cm causing displacement of surrounding structures. The mass arose from the left psoas muscle. It was multi-lobulated and multi-septated with mixed density within. It extended into the peri-aortacaval region, suggesting nodal involvement. The mass also displaced and compressed the descending colon anteriorly and the inferior vena cava (Figure 1). Multiple small well-defined hypodense lesions were seen in liver segments VI and VII measuring approximately 4mm-5mm. The spleen, pancreas, adrenal glands and kidneys were normal. There was no ascites.

Ultrasound-guided biopsy showed fragmented cellular tumour tissue of plump spindle cells arranged in short intersecting fascicles. The spindle cells were moderately pleomorphic. Immunohistochemistry was positive for desmin. The tissue biopsy was consistent with high-grade spindle cell neoplasm. Differential diagnoses include leiomyosarcoma and spindle cell rhabdomyosarcoma.

The patient was presented to a multidisciplinary team discussion involving orthopaedic surgeons, general surgeons, urologists, clinical oncologists, pathologists and radiologists. The collective decision was for complete resection of the large primary tumour, despite the nodal spread of the tumour, for the purpose of immediate symptom resolution. The left ureter was stented to facilitate intraoperative identification. A left extended ilioinguinal incision allowed for partial excision of the left psoas major, following which the patient was repositioned supine and a midline incision done and extended to the left ilioinguinal excision. Peritoneal cavity was entered along the left paramedian plane at the fascia layer to allow for tumour mobilization medially from the paraaortic plane. The large primary tumour was resected entirely. Intraoperatively, three further retroperitoneal masses were identified embedded in the sigmoid mesocolon, two of which were resected entirely. However, the third retroperitoneal mass was located immediately anterior to the descending aorta with no clear

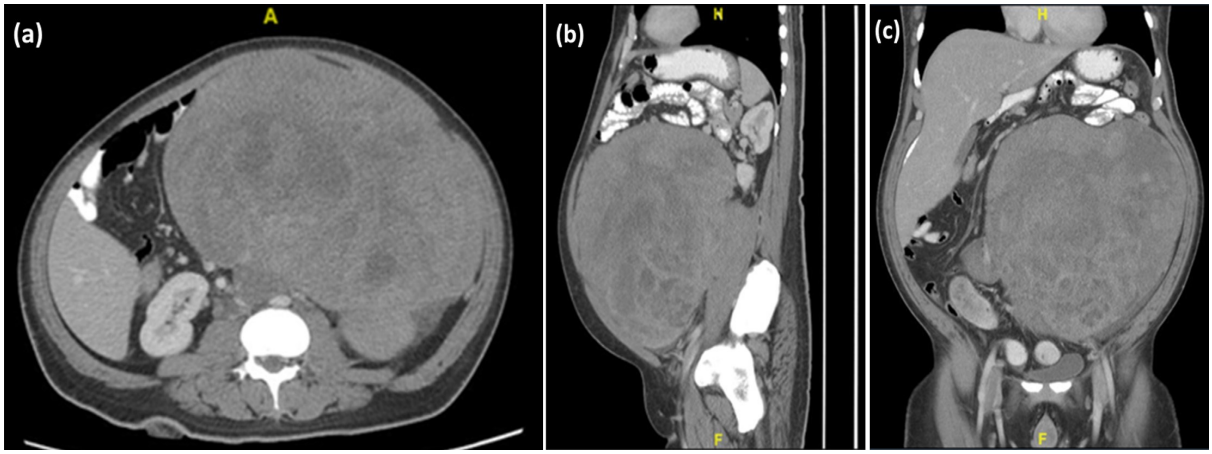

Figure 1: (a) Axial, (b) sagittal and (c) coronal views of the retroperitoneal mass. [(A) Anterior, (H) Head, F (Foot)]. (Click image to enlarge).

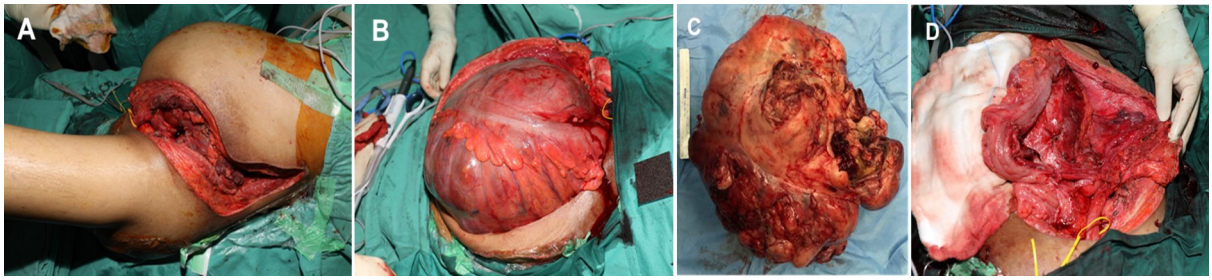

Figure 2: A) Extended Ilioinguinal incision allowing for retroperitoneal dissection of the tumour. B) Entry into the abdominal cavity along the fascial place exposing descending colon and sigmoid closely adherent to the tumour. C) Gross specimen of retroperitoneal rhabdomyosarcoma. D) Defect following debulking of the third retroperitoneal tumour. (Click image to enlarge).

plane visible between the tumour and the aorta, and only debulking of this third retroperitoneal mass could be carried out (Figure 2). The patient had an uneventful post-operative recovery and was discharged home on day seven post-surgery.

Histopathological examination revealed high-grade sarcoma exhibiting large pleomorphic and bizarre cells displaying hyperchromatic nuclei and abundant eosinophilic cytoplasm. Multinucleated giant cells and rhabdomyoblasts were present. These cells showed strong positivity for desmin and myogenin immunohistochemical stains which are the markers for skeletal muscle (Figure 3).

Four weeks post-operatively, he subsequently underwent adjuvant chemoradiotherapy with 16 fractions of radiotherapy followed by Doxorubicin. Follow-up imaging showed poor tumour response to the adjuvant radiotherapy evidenced by the tumour regrowth at the para-aortic region. Unfortunately, he developed neutropenic sepsis and eventually succumbed to the disease progression three months after his surgery.

DISCUSSION

Pleomorphic rhabdomyosarcoma is a rare entity. Despite accounting for a significant fraction of adult rhabdomyosarcoma, the latter itself is uncommon, representing only 3% of adult soft tissue sarcomata; soft tissue sarcoma in turn constitutes less than 1% of adult solid tumour malignancies.[1] In addition, retroperitoneal soft tissue sarcoma specifically makes up for only 12% of soft tissue sarcomas with an incidence of 2.7 per million people.[3] Retroperitoneal sarcoma can be classified into mesodermal (e.g. lipoma, leiomyoma or rhabdomyosarcoma), neurogenic (e.g. neurofibroma) and extragonadal (e.g. teratoma).[4] According to the World Health Organisation (WHO), rhabdomyosarcoma can be divided into 4 subtypes: embryonal, alveolar, pleomorphic and spindle cell/sclerosing.[5] WHO defines pleomorphic rhabdomyosarcoma as a high-grade sarcoma composed of undifferentiated round and spindle cells that display skeletal-muscle differentiation without embryonal or alveolar components.[5] The

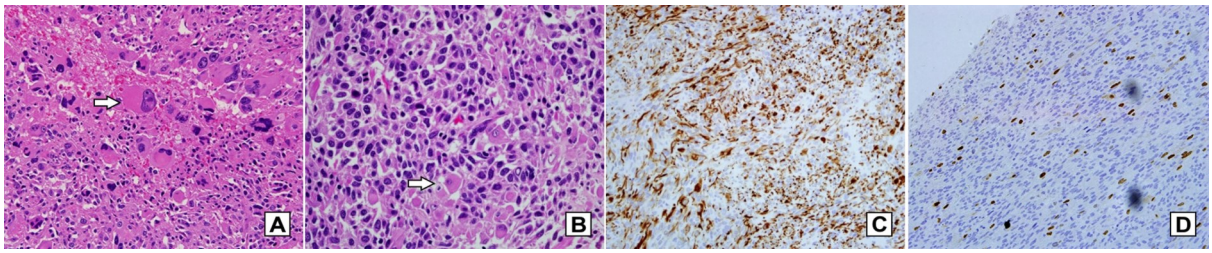

Figure 3: A) and B) Presence of large bizarre and pleomorphic cells and multinucleated cells and typical rhabdomyoblasts (arrowed) (H&E stain X400). C) Immunohistochemical stains (X400) show strong positivity for desmin and D) myogenin. (Click image to enlarge).

overall median age at diagnosis was 71.5 years with a male-to-female predilection ratio of 1.8:1.[6]

Patients with retroperitoneal sarcoma often present late as a result of the large retroperitoneal space available for the tumour to expand before causing symptoms. They also often present with non-specific symptoms.[7] In one study, abdominal pain was the most common presenting symptom in 69 out of 138 patients (50%) with retroperitoneal soft tissue sarcoma, and an abdominal mass was the most common examination finding in 82 out of 138 patients (59%). Sixty-four percent (88 out of 138) of patients presented within 6 months from onset of symptoms.[8]

Given the complexity of retroperitoneal anatomy, proper investigation preoperatively is crucial. It helps to delineate the size of the tumour, other organs or vessels which may be invaded and the organs that require partial resection or reconstruction. Ultrasound, CT scan and ultrasound-guided biopsy (gold standard), as well as immunohistochemical studies, are essential for diagnosis and planning of treatment.[3] In our case, the ultrasound-guided biopsy and the immunohistological studies (positive for desmin) helped narrow down the differential diagnoses to rhabdomyosarcoma of the pleomorphic subtype and leiomyosarcoma.

A study by Lee et al., demonstrated that 53% of retroperitoneal tumours at the time of diagnosis exceeded 10 cm in diameter with a median diameter of 13.7 cm (range: 2.5 - 39 cm).[9] Our patient had a tumour mass with a diameter of 29 cm. Multiple studies have stressed that complete resection of the tumour is of utmost importance, with incomplete resection and even microscopic spread adversely affecting prognosis. Lee reported that completeness of resection, adjuvant therapy and re-resection after recurrence were predictive factors on multivariate analysis of 41 patients with pleomorphic rhabdomyosarcoma with p-value of 0.006, 0.008 and 0.027 respectively.[9] Garcia-Ortega et al., also demonstrated complete resection of retroperitoneal sarcomas as a predictive factor; in their study, only 57.9% of their patients were able to undertake complete resection and the mean survival of patients with complete against incomplete resection was 55.6 against 28.1 months respectively.[10] Management at high volume centres have also demonstrate improved outcomes in multiple studies.[3,4,11] In line with this, as demonstrated in this case, despite complete resection being used for the main primary tumour and two adjacent retroperitoneal masses, debulking of the third retroperitoneal mass due to its proximity to the aorta still eventually resulted in a poor post-operative outcome as the patient passed away 3 months later.

With regards to adjuvant therapy, Noujaim et al., demonstrated the limited success of chemotherapy in adult pleomorphic rhabdomyosarcoma, with only 1 out of 45 patients confirmed to have partial response with paediatric-type chemotherapy regimen vincristine, actinomycin D and cyclophosphamide only.[6] The authors attributed this to the very complex karyotype in the genetic composition of pleomorphic rhabdomyosarcoma in comparison to alveolar and embryonical rhabdomyosarcoma. They also concur that wide surgical resection continues to be the primary predictive factor in patient survival. Their study further demonstrated poor prognosis in adult pleomorphic rhabdomyosarcoma with the median survival for those with localised (32 patients, 71.1% of cohort) and metastatic disease (13 patients, 28.9% of cohort) being 12.8 months and 7.1 months respectively. The relapse rate was 53.8% (4 local and 10 distant relapses).[6] The impact of neoadjuvant chemotherapy also seems equivocal with a scarcity of evidence regarding its potential benefits in treating soft tissue sarco-

mata.[12]

Therefore, preoperative or postoperative radiotherapy with complete surgical resection remains the standard of care for localised disease and improves survival rates in a minority of cases [6,13,14].

CONCLUSION

Rhabdomyosarcoma is a highly malignant sarcoma which has poor prognosis. This case report illustrates how debulking surgery of secondary tumours still resulted in poor postoperative prognosis in spite of complete resection of the primary tumour and multiple other secondary tumours. In addition, preoperative neoadjuvant chemoradiotherapy may assist in downstaging the tumour to allow for complete resection.

FINANCIAL DISCLOSURE OR CONFLICT OF INTEREST

The authors of this manuscript certify that there are no conflict of interest nor any financial interest in the subject matter or materials discussed in this manuscript.

CONSENT

We have acquired consent from patient for all images used in publication purpose.

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