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Case Report

# A Very Aggressive Case of Adult Omental Rhabdomyosarcoma: Case report and Literature Review

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## Abstract

The greater omentum is a common location for metastatic tumors. On the other hand, the primary tumors of the omentum are rare. Intra-abdominal rhabdomyosarcoma (RMS) is extremely rare in adults. Few cases have been reported in the literature and the most of them were in the pediatric population. Omental rhabdomyosarcoma (RMS) in an adult, is an even rarer case that deserve the presentation. We are reporting a case of intra-abdominal rhabdomyosarcoma in a 54-year-old lady who presented with constipation and increasing abdominal girth. Investigations revealed disseminated omental rhabdomyosarcoma. We decided to start chemotherapy, but unfortunately she passed away on the tenth day of treatment.

## Introduction

Rhabdomyosarcoma (RMS) is a malignant soft tissue tumor. It constitutes more than 50% of soft tissue sarcomas (STS) in children [1]. Whereas, it is extremely rare in adults and it accounts for less than 1% of all malignancies [2]. It can originate in any site of the body, but the most common locations are the head, neck and genitourinary tract [3]. In any age, intra- abdominal RMS and especially the omental involvement, is very rare, and only few cases have been reported in the literature [4]. Here, we are reporting a very rare case of omental rhabdomyosarcoma in adult.

## Case presentation

A 54-year-old lady, smoker, with no known food and

drug allergies, nor any past medical or surgical history, presented to the emergency department for acute onset of right upper quadrant abdominal pain associated with nausea and postprandial vomiting. Abdominal examination was only remarkable for positive Murphy sign. Laboratory studies revealed mild leukocytosis (WBC 12 x $10^3$/µL) with neutrophil shift (PMN 85%). Abdominal ultrasonography was done and showed acute calculus cholecystitis for which she was scheduled to undergo laparoscopic cholecystectomy.

Under general anesthesia, the abdomen was insufflated and 2 trocars were inserted. Exploration revealed mild amount of clear peritoneal fluids in addition to adhesions between the omentum, liver and the anterior abdominal wall. The gallbladder was found to be severely inflamed. Peritoneal fluids were taken for bacterial culture and cytology examination. Due to the technical difficulty related to the adhesions, the operation was converted to open and cholecystectomy was then done. The post-operative course was uneventful and the patient was discharged home on the second postoperative day in good condition. The pathological examination of the gallbladder revealed chronic cholecystitis without malignancy. Unfortunately, the cytological examination of the peritoneal fluids demonstrated clusters of dysplastic epithelial cells consistent with carcinoma. The patient was then contacted and advised to come back for further workup but she was reticent because of the corona outbreak.

She presented after one month for evaluation of constipation and increasing abdominal girth. On physical examination: Her abdomen was distended with mild diffuse tenderness and dullness on percussion. Laboratory tests were normal except for mild leukocytosis (WBC 13 x $10^3$/µL). A CT scan of the abdomen and pelvis revealed a moderate amount of ascites with a large intra-abdominal mass located at the level of left mid-lower abdomen, measuring 12 x 9 cms, causing displacement of the adjacent bowel loops (Figure 1). Tumor markers including C.E.A, CA19-9, AFP, CA 125 were within normal range.

Faced with this presentation of ascites and intra-abdominal masses and the inability to identify the origin of the tumor, we decided to perform a diagnostic laparoscopy. Exploration revealed a large amount of clear peritoneal fluids with diffuse omental thickening and adhesions, in addition to a large, hard and nodular mass located within the greater omentum. It was adherent to several small bowel loops and the transverse colon. The liver, spleen and ovaries appeared normal. As the mass was unrespectable, biopsies were taken.

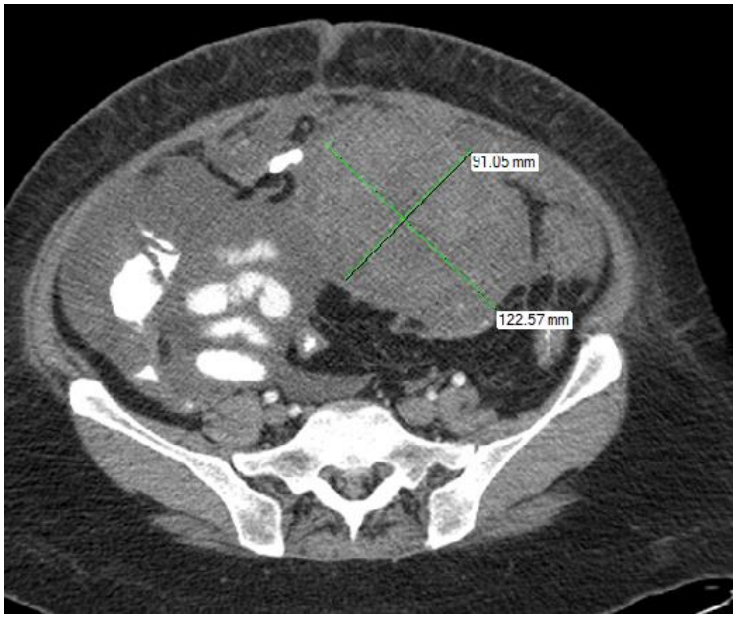


**Figure 1:** Showing a large, well defined intra-abdominal mass, measuring 12 x 9 cms

The histopathological examination revealed fibro-adipose tissues with multiple foci of infiltrating poorly differentiated neoplasm, consists of spindle mesenchymal cells with multiple better differentiated rhabdoid and muscle fiber cells, consistent with rhabdomyosarcoma (Figure 2).

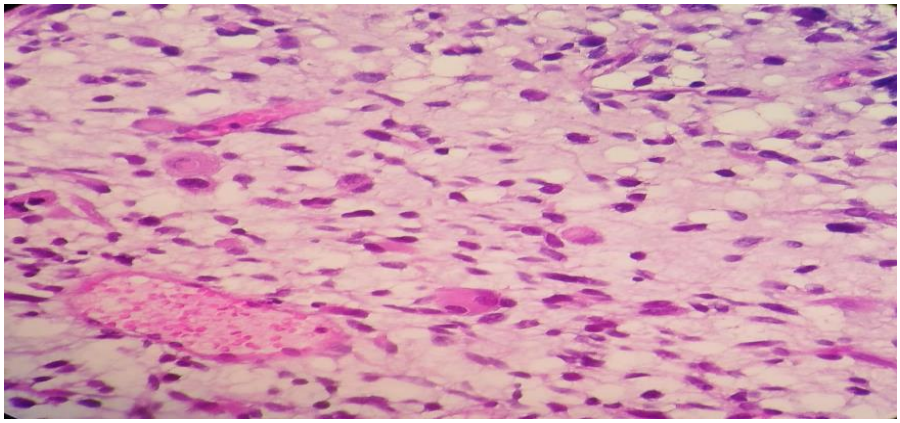

**Figure 2:** H and E; 40x40 X showing mixture of spindle cells, small round to ovoid cells and prominent rhabdoid cells having an eosinophilic cytoplasm and eccentric nuclei

Immunohistochemistry revealed strong staining for Desmin, Myogenin, CD34 and MYOD-1, therefore confirming the diagnosis of RMS (Figures 3, 4 and 5).

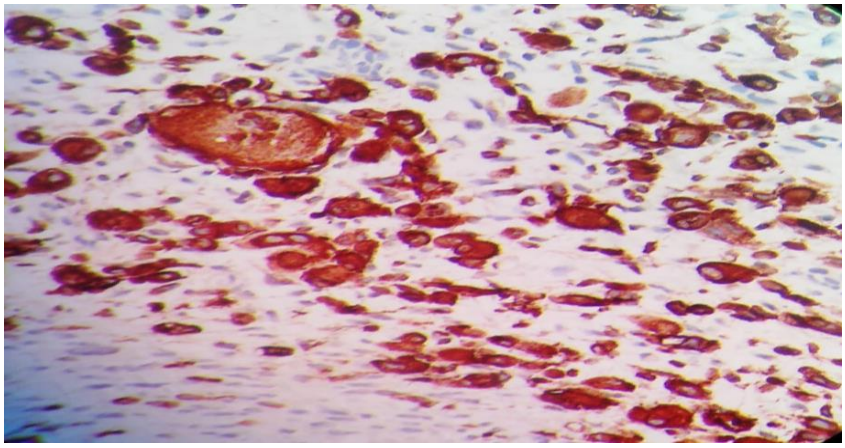

**Figure 3:** Immunohistochemistry, 40 × 40 X shows tumor cells showing desmin positivity

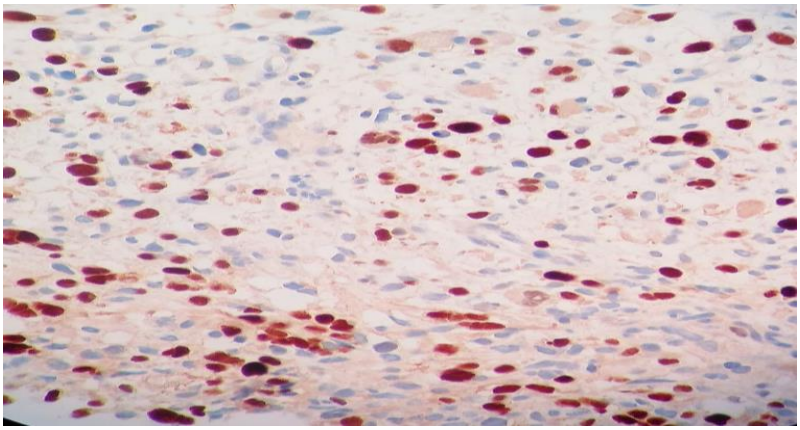

**Figure 4:** Immunohistochemistry, 40 × 40 X shows tumor cells showing Myogenin positivity

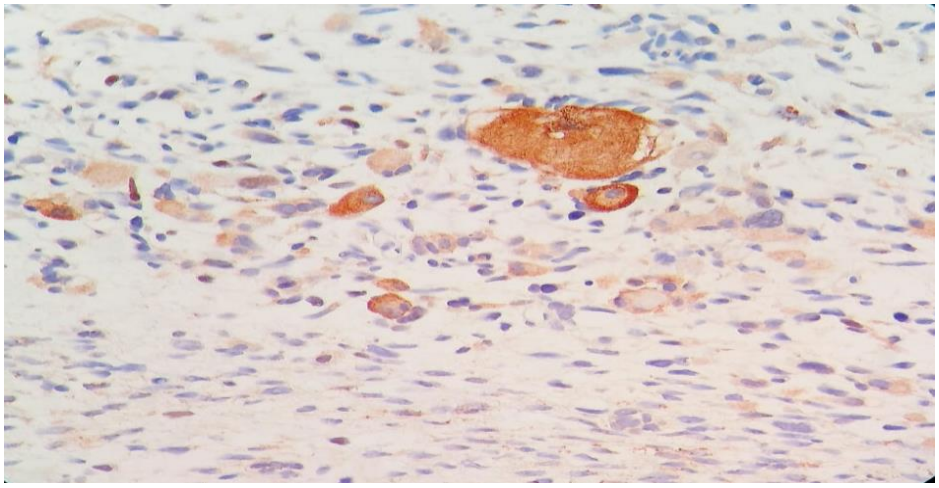

**Figure 5:** Immunohistochemistry, 40 × 40 X shows tumor cells showing MYOD-1 positivity

The patient was then started on VAC-IE regimen (vincristine, doxorubicin, cyclophosphamide alternating with ifosfamide and etoposide). Unfortunately, her condition began to deteriorate rapidly after the tenth day of chemotherapy until she was admitted to the intensive care unit and passed away.

## Discussion

RMS is the most common soft tissue sarcoma in children. Omental RMS is an extremely rare malignancy. Only few cases have been reported in the literature and almost all of them were in the pediatric population [4]. Its pathogenesis is still poorly understood [7]. Some evidence suggests that RMS can arise from aberrant development of non-myogenic cells [8]. And mutations in macrophage inhibitory factor (MIF) and p53 are responsible for tumor progression [7]. The most common symptoms of intra-abdominal RMS –including omental RMS- are abdominal discomfort, abdominal mass and distention [9].

The imaging of choice for intra- abdominal RMS is Magnetic Resonance Imaging (MRI), which can determine the tumor origin and the relationship between the tumor and the adjacent organs. RMS is usually hyperintense on T2 and hypointense on T1 with heterogenous enhancement [6]. Computed Tomography CT scan, in the other hand, is important for staging and for assessing spread to lungs, as lung is the most common site of metastasis [8]. We note that recently PET-CT and PET -MRI are gaining a huge role in the management of RMS, from assessing response after neoadjuvant treatment to detecting residual tumor to local recurrence, metastatic spread and even have a role in the diagnosis of RMS. They give an insight on the tumor biology [10].

The world health organization (WHO) classifies this disease into 4 categories which differ by their histomorphology, most frequent localization, prevalence, IHC, genetics: Embryonal, alveolar, pleomorphic, and spindle cell/sclerosing RMS [11]. Pleomorphic subtype RMS is the most common subtype in adults [12]. Concerning the histological staining in helping to diagnose RMS, the myogenin and MyoD1, contrary to desmin, are highly sensitive and specific myogenic nuclear transcription factor for RMS and present early in the skeletal muscle differentiation [13].

Due to its rarity, the optimal treatment of adult RMS is uncertain [14]. Currently, the management of adult RMS is in line with the multidisciplinary treatment protocol which is proposed by the Intergroup Rhabdomyosarcoma Study (IRSG) group for treating children with RMS and includes complete surgical resection of the tumor with free margins, radiation and chemotherapy. Surgery and radiotherapy are used for the treatment of primary tumor site; however, chemotherapy is used to prevent tumor spread [15].

IRSG recommends that chemotherapy should be given to all patients with RMS and it improves survival. The recommended combination of chemotherapy includes vincristine, actinomycin-d, etoposide or ifosfamide and cyclophosphamide. Irnotecan, a topoisomerase I inhibitors is also recommended. Concerning radiotherapy, all patients must receive radiotherapy to ablate the microscopic residual tumor and to obtain long term control [16].

Chemo-radiotherapy can also be used as neo-adjuvant therapy that results in down staging of the disease. Thus, it allows complete excision of the tumor [17].

The prognosis is related to the presence of metastasis at presentation and the response to chemotherapy [18]. Adult RMS is very aggressive tumor and has a low 5-year survival rate (27%) compared to that observed in pediatric population [19].

Since RMS in children is a different entity from that of adults in terms of sensitivity to chemo-and radiotherapy, some studies showed that the prognosis in adults can be similar to that observed in children, if they are treated aggressively by using the pediatric protocol [18].

## Conclusion

Adult omental RMS is an extremely rare and highly malignant neoplasm with a poor prognosis, even after surgical and chemotherapy interventions. Early diagnosis, complete resection, and appropriate radiotherapy and chemotherapy are the keys to managing this disease. Further studies are needed to develop the optimal treatment protocol for improving the prognosis of patients with this rare but deadly cancer.

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