Adult Omental Alveolar Rhabdomyosarcoma: an unusual site. A case report and literature review

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Case Report

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Abstract

Background: Omental alveolar rhabdomyosarcoma in adults is a very hazardous tumor. Its pathogenesis is still under discussion and many studies consider it different from its pediatric counterpart. This tumor can arise from any soft tissue in the body thus it can be found anywhere, but it's found mainly in the head and neck region. Omentum as the primary site was reported only by six studies in the English literature before. In this paper we aim to report the seventh case and perform a review of all paperworks that pertains to this topic.

Case Presentation: we present a case of a 52 years old smoker male. His medical record was not significant. He presented with dull pain in the right iliac fossa. He had suffered from generalized abdominal pain in the past several months but this time the pain was unbearable. On examination he had a palpable mass in the region. CT revealed a mass arising from omentum. A laparotomy was performed and biopsies were taken, stained, examined and led to the diagnosis of ARMS. The patient was put on radio and chemotherapy. He missed some appointments after that. When he was admitted 2 months later he was suffering from neurological symptoms which after performing lumbar puncture turned out to be tumor metastasis. Regrettably the patient died after his second cycle of chemotherapy.

Conclusion: Adult rhabdomyosarcomas are rare tumors that can arise from any soft tissue including omentum and should be taken into consideration when we deal with primary tumors originating from that area. The studies and our understanding for this neoplasm is still so limited and should be expanded widely.

Background

Alveolar Rhabdomyosarcoma (ARMS) is one of the four subtypes of rhabdomyosarcoma that was proposed by Horn et al in 1956 and is the one with the worst prognosis. It rarely affects the adult population. The tumor can originate from any soft tissue. Mostly from the deep soft tissue of the extremities and paraspinal region. Omental origin in adults was only mentioned in six cases and were all alveolar typed.

Until today, this topic is still poorly discussed as all of the previous reviews had discussed RMS as one entity with no in-depth study of the properties and treatment options of each one of its subtypes severally.

Here we report a case of omental ARMS in a 52 years old male which presented with symptoms similar to appendicitis. We also conducted a review of literature on the topic of omental alveolar rhabdomyosarcoma in adults.

Case Presentation

A 52 years old male with no significant medical or surgical history other than being a heavy smoker presented to our department with generalized abdominal pain for several months which became more dull and localized in the right iliac fossa in the last two days. The pain was accompanied with bowel habit changes. He had lost about 20 kilograms in the last couple months although his appetite was normal.

The patient looked uncomfortable, his abdomen was tender and rigid. He had a palpable nodular mass in the right iliac fossa and a single palpable axillary lymph node.

Abdominal US (ultrasonography) showed a 12.2*3.6 cm mass with cecal distention and grade I hydronephrosis in the right kidney. A non contrast CT abdomen showed a small mass measuring 7*4*3 cm (Fig. 1) rising from abdominal wall and adhering to cecum. A colonoscopy showed external compression and a massive ringed ulcer on the cecal area. Biopsies were taken and came negative for colorectal cancer.

The patient underwent a laparotomy and an excisional biopsy 9*6 cm was obtained from the colonic omentum. It looked yellow, soft and rubbery in consistency and attached to many encapsulated nodules. Microscopically, the mass had a fibrous septa enclosing alveolar nests of neoplastic small blue round cells lacking a discriminant cytoplasm with high mitotic rate. In the lumen of the alveoli there were clusters and individual neoplastic cells.

Immunohistology was negative for CD3, CD19, CD20, LCA, CK, CHROMO, SYNAPTO, CD99, DOG1, MYOD1 but it came positive for DESMIN.(Fig. 2)

Based on immunostaining results and the histologic picture Alveolar rhabdomyosarcoma was diagnosed.
Bone scan (Fig. 3) showed Multiple focal areas of increased tracer uptake around the orbit, multiple ribs bilaterally, vertebrae, inferior angle of left scapula, manubrium sterni.

A full body CT was scheduled to show if there were any metastases to other organs. however it was delayed due to the war circumstances.

When the patient was readmitted two months later, the patient developed neurological symptoms including lethargy and anterograde amnesia. LP (lumbar puncture) showed the presence of neoplastic cells in CSF (cerebrospinal fluid). chest, abdomen and brain CT showed no metastases. The patient was put on radiotherapy and chemotherapy consisting of vincristine and cyclophosphamide. sadly the patient died two weeks later after his second chemo cycle.

Discussion And Conclusion

Soft tissue sarcomas (STS) are an unusual finding mainly affecting the pediatrics population\[1\]. Rhabdomyosarcomas account for more than half of them\[2\]. This percentage drops significantly in adults where it doesn't exceed 2–5\%\[3\].

Rhabdomyosarcoma seems to originate from any immature mesenchymal cells regardless of what it was dedicated for\[2\].

Its pathogenesis is still poorly understood. Mutations in macrophage inhibitory factor (MIF) and p53 are responsible for tumor progression\[1\]. Compared to pediatric RMS which has clear risk factors such as prenatal drug exposure and X ray, there are no studies published regarding the etiology of RMS in adults\[4\].

Stout et al in 1946 was the first to classify Rhabdomyosarcoma tumors\[6\]. However, in 1956 Horn et al proposed the widely-used classification which comprises the embryonal, botryoid (a subtype of embryonal), alveolar (20% of cases and has the worst prognosis\[5\]) and pleomorphic types\[2\]. Whereas WHO classifies this tumor into Embryonal, alveolar, pleomorphic, and spindle cell/sclerosing RMS\[1\]. The first well described Alveolar Rhabdomyosarcoma case was done by Riopelle et all in 1956\[7\].

Alveolar RMS, consists 20–30% of RMS in patients whose ages are 15 to 20 years old\[8\]. It arises mostly from the deep soft tissue of the extremities, paraspinal, perineal regions and the paranasal sinuses\[8\].

Omental rhabdomyosarcoma in adults is uncommon and very rare as a primary site\[9\]. All previous cases were alveolar\[10\]. Here we perform a literature review of alveolar rhabdomyosarcoma of omentum. We have excluded two cases because they were not in english.

Consequently, six cases were studied with our case being the seventh one. Five of the seven cases were men. Ages ranged between 21 and 85 years old [Table 1].

Symptoms of omental tumors including ARMS are nonspecific and mass related, most commonly: abdominal discomfort (45.5\%), abdominal mass (34.9\%), and abdominal distention (15.2\%)\[9\]. Other symptoms depending on our review were constipation (42.8\%) followed by nausea and vomiting (28.5\%), weight loss (28.5\%) and pyrexia was an odd complaint in one case\[11\].

Ultrasonography (US) is usually the first performed investigation for abdominal complaints\[12\]. The tumor is reported as a well defined mass with mixed echogenicity in most of the studies reviewed. MRI is the gold standard for RMS in the abdominal region\[13\]. The mass appears hyperintense on T2, hypointense on T1 with heterogeneous enhancement\[13\].

Computed Tomography (CT) was widely used in most reviewed cases\[1\]. RMS was described as a large, well defined and solid mass\[1\]. Although no clear pattern of enhancement was found in the literature, one case mentioned mild enhancement\[2\].

PET scan defines the successfullness of treatment by detecting residual tumor, local recurrence and metastatic spread\[1\].

Metastatic cells most commonly affect lungs\[1\]. But they also spread to bone, subcutaneous tissues, lymph nodes, liver, myocardium, kidney, adrenal glands, and the brain\[14\].

Definitive diagnosis can not be made on the basis of imaging studies as it has no pathognomonic sign and the diagnosis is made on the light of microscopy and immunostaining\[9\]. The differential diagnosis encompass blue small round cell tumors: Ewing Sarcoma or primitive neuroectodermal tumor, Non-Hodgkin lymphoma (particularly B-cell lymphomas), Neuroblastoma, Desmoplastic small round

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cell tumor, Epithelioid sarcoma, poorly differentiated monophasic synovial sarcoma, clear cell sarcoma (previously known as malignant melanoma)[15].

The gross appearance of ARMS is similar to all rhabdomyosarcomas. It appears as well-circumscribed nodular firm masses ranging in consistency and size[2]. Cystic degeneration and areas of necrosis were mentioned in many studies[9][11][14]. V Sennu et al reported increased vascularity within the tumor[11]. Histologically, the tumor appears as irregular alveolar space made of sheets of predominantly small and round cells with abundant eosinophilic cytoplasm, small oval nuclei and prominent nucleoli separated by a fibrous septa[16]. Mitotic figures and spindle cell myoblasts can also be seen.

Cross-striations which are the standard for RMS diagnosis can only be seen in 50–60% of cases[2], but among the 6 cases in the literature regarding the alveolar type only Sanjay Kumar Yadav et al mentioned it[2]. On electron microscopy Z-bands can be identified. Immunohistochemistry reveals positivity for Desmin, Myogenin, MYOD-1, and Vimentin. Monoclonal antibodies against desmin, muscle specific actin, sarcomeric actin and myoglobin have a high specificity and sensitivity for diagnosing the myogenic nature of the tumor[2].

Treatment of RMS should be multidisciplinary according to the Intergroup Rhabdomyosarcoma Study (IRSG)[16]. Complete resection is essential and further excision is indicated if microscopic extensions were found[2]. Concerning chemotherapy, all patients must receive a combination therapy of Vincristine, Actinomycin-d, Cyclophosphamide, Etoposide or Ifosfamide and Irinotecan to attain long term control of the tumor[2]. Chemotherapy also improves survival and should be admitted shortly after diagnosis or after resection to encounter any metastasis[2]. Nonetheless, Hawkins et al recommended against this as they have not seen any increase in survival rate in the adult population[19]. Lastly, radiotherapy which has been used widely for the treatment of RMSs in infants has a significant role in reining residual disease. Doses between (36 and 54) Gy are usually utilized[9].

A follow up should be scheduled every 3 months in the first year, then every 6 months for the next two years and then once every year[9].

Prognostic factors for adult RMS (regardless of subtypes as they were insignificant prognostically) include: age, tumor size, extent of disease, and margin status[18]. On the other hand Nestor F. Esnaola et al in their study which was published in the same year indicated the absence of association between these factors and survival rate in patients who underwent multimodellary treatment[19]. Mostly, adults had significantly worse outcomes than children (5-year overall survival rates, 27%±1.4% and 61%±1.4%, respectively; P < .0001)[20]. In our review, 3 patients out of 7 died shortly after presentation including our case. Bad response to chemotherapy is a predictor of unfavorable outcomes[19].

Limitations:

1. We couldn't perform MRI or full body CT because the patient died before his scheduled date.
2. We couldn't get any image from the laproscopy nor the previous endoscopy.
3. We couldn't conduct more immunostaining examinations due to the shortage of these materials.
4. In our review we excluded two cases: “Alveolar rhabdomyosarcoma originating from the greater omentum” by H Svanholm as it was not in english and we couldn't reach a scanner version and “Alveolar Rhabdomyosarcoma: apropos of a rare location” by Petit ML et al as the tumor arises from mesentery and it was in french.

Abbreviations

ARMS: alveolar rhabdomyosarcoma; STS: soft tissue sarcoma; Us: ultrasonography; CT: computed tomography; MRI: magnetic resonance imaging, LP: lumbar puncture; CSF: cerebrospinal fluid; Gy: gray is the international system of units (SI) equivalent of 100 rads.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication
Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

Availability of data and materials

All the data supporting our findings is contained within the manuscript.

Acknowledgments

We would like to thank

Competing interests

The authors declare that they have no competing interests.

Funding

We have no funding to disclose.

Contributions

MSSH, ABK made major contributions to manuscript writing. MSSH is also the corresponded. ACH diagnose3d the case and performed a critical revision of the manuscript. MN and AM helped with the diagnosis and they provided us with the study materials. RA helped us with pathology and immunostaining. All authors read and approved the final manuscript.

References


Table

Table 1
<table>
<thead>
<tr>
<th>Case Number</th>
<th>Author/Date</th>
<th>Topic</th>
<th>Patient's Age</th>
<th>Patient's Sex</th>
<th>Symptom</th>
<th>Primary Location in the abdomen</th>
<th>Size of tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>V SEENU, M C MISRA, et al 1995</td>
<td>Omental Rhabdomyosarcoma Presenting with Pyrexia</td>
<td>45</td>
<td>Male</td>
<td>Pyrexia, night Sweats, no chills or rigor, no localizing Symptoms and a palpable lump in the lumbar region</td>
<td>behind the urinary bladder</td>
<td>10 cm x 8 cm</td>
</tr>
<tr>
<td>Case 2</td>
<td>Samer H. Dbouk, Hussein Mcheimeche, et al 2020</td>
<td>A Very Aggressive Case of Adult Omental Rhabdomyosarcoma: Case report and Literature Review</td>
<td>54</td>
<td>Female</td>
<td>Acute right upper quadrant abdominal pain, Nausea, postprandial vomiting, constipation and increasing abdominal girth</td>
<td>mid-lower abdomen</td>
<td>12 cm x 9 cm</td>
</tr>
<tr>
<td>Case 3</td>
<td>Priyank Pathak, Mayank Nautiyal, et al 2015</td>
<td>Omental rhabdomyosarcoma (primary rhabdoid tumor of greater omentum): a rare case report</td>
<td>21</td>
<td>Male</td>
<td>Dull abdominal pain with palpable lump in the hypochondriac region extending up to the epigastric</td>
<td>left hypochondrium</td>
<td>9.8 cm x 7.4 cm</td>
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<tr>
<td>Case 4</td>
<td>L. C. J. van Rensburg 1980</td>
<td>ALVEOLAR Rhabdomyosarcoma Of THE GREATER OMENTUM: A CASE REPORT</td>
<td>84</td>
<td>Female</td>
<td>anorexia, loss of weight, lassitude, constipation, constant pain in the epigastrium, and a palpable movable mass above the umbilicus</td>
<td>Epigastric region</td>
<td>Two masses one is 8.0 cm in diameter in the omentum. The second mass is 2.0 cm in diameter present on the lesser curve of the stomach</td>
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<td>Case 5</td>
<td>Sanjay Kumar Yadav &amp; Dipendra Kumar Sinha, et al 2015</td>
<td>Primary Intra-Abdominal Rhabdomyosarcoma in an Adult: an Unusual Presentation and Review of Literature</td>
<td>65</td>
<td>Male</td>
<td>Vomiting, abdominal distention and total constipation</td>
<td>Subhepatic region and extending up to the right iliac fossa</td>
<td>23×20×15 cm</td>
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<tr>
<td>Case 6</td>
<td>ARTHUR PURDY STOUT, J. HENDRY, et al 1963</td>
<td>PRIMARY SOLID TUMORS OF THE GREAT OMENTUM Case NO.23</td>
<td>53</td>
<td>Male</td>
<td>Left upper quadrant and left lumbar pain with occasional radiation</td>
<td>left upper quadrant</td>
<td>Not mentioned</td>
</tr>
<tr>
<td>Case 7</td>
<td>Mohammad Sami Alshutaihi, Ahmad bisher Kelarji, et al</td>
<td>52</td>
<td>Male</td>
<td>Adult Peritoneal Alveolar Rhabdomyosarcoma: an unusual site. A case report and literature review.</td>
<td>Right iliac fossa</td>
<td>7<em>4</em>3 cm</td>
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<td>Case number</td>
<td>Laboratories</td>
<td>Radiology</td>
<td>Histology</td>
<td>Treatment</td>
<td>Prognosis</td>
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<td>Case 1</td>
<td>Not mentioned</td>
<td>U.S. well defined mass with mixed echogenicity</td>
<td>Globular, well defined, smooth, nontender mass extreme vascularity, friability, and gelatinous and cystic degeneration</td>
<td>Complete resection with doxorubicin as adjuvant chemotherapy</td>
<td>No symptoms after 38 months follow-up</td>
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<td>CECT. well defined mass with solid and cystic areas with moderate enhancement and necrotic areas</td>
<td>Mitotic figures were common and ranged from 5-30/10 HPF</td>
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<td>Case 2</td>
<td>Leukocytosis with neutrophilia. C.E.A, CA19-9, AFP, CA 125 were within normal range</td>
<td>U.S. acute calculus cholecystitis</td>
<td>Gross appearance: large, hard and nodular Mass. Immunohistochemistry revealed strong staining for Desmin, Myogenin, CD34 and MYOD-1</td>
<td>Chemotherapy on VAC-IE regimen</td>
<td>Died on the tenth day of chemotherapy</td>
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<td>CT revealed a mass located at the level of left mid-lower abdomen</td>
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<td>Case 3</td>
<td>Normal values</td>
<td>Ultrasound guided FNAC from the left hypochondriac region showed deposits of adenocarcinoma. CT revealed a large well defined mass lesion in the left hypochondrium, causing displacement of the bowel loops</td>
<td>On cross section examination, cut surface was homogenous grayish white, areas of hemorrhage and necrosis. poorly differentiated malignant round cell tumor with metastasis in regional lymph nodes and perinodal extension.</td>
<td>Complete excision via omentectomy. chemotherapy (4 cycles) vincristine, dactinomycin, and ifosfamide regimen</td>
<td>Good, No signs of recurrence.</td>
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<td>Case 4</td>
<td>Normal values</td>
<td>A barium enema showed that the mass had pushed the transverse colon downwards</td>
<td>In the section, the tumor was solid. with a tan color, and showed irregular areas of necrosis, involved the muscle coats of both the large bowel and the stomach. On microscopical examination, alveolar growth pattern. The alveolar spaces were lined by large pleomorphic cells with abundant eosinophilic cytoplasm. No crosstriations could be demonstrated. Up to 6 mitoses per high power field, many of which were atypical. Much of the tumor had undergone necrosis. The reticulin stain showed the alveolar pattern of the tumor clearly.</td>
<td>partial gastrectomy and partial transverse colectomy with a fairly deep removal of the transverse mesentery.</td>
<td>good post-operative recovery</td>
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<td>Case 5</td>
<td>Not mentioned</td>
<td>X ray erect abdomen was done which showed multiple air fluid levels. CECT abdomen showed a large, solid, mildly enhancing mass arising from the subhepatic region and extending up to sheets of predominantly small and round cells with abundant and eccentric eosinophilic cytoplasm, small oval nuclei with prominent nucleolus. Few spindle cell myoblasts with prominent tapered fibrillar eosinophilic cytoplasm and cross striations were also present.</td>
<td>Due to unresectability of the mass, incisional biopsy was taken and ileostomy was made.</td>
<td>Not mentioned</td>
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the right iliac fossa, pushing and compressing the adjacent bowel loops

Highly cellular areas around blood vessels
alternating with parvocellular mucoid regions (resembling normal embryonic myogenesis) were seen. Cells stained for myogenin on immunohistochemistry

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<th>Case 6</th>
<th>Not mentioned</th>
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<td>A roentgenogram showed a mass in the left upper quadrant with displacement of stomach and descending colon. Intravenous and retrograde pyelograms showed distortion of the pelvis and calyces of the left kidney from outside pressure.</td>
<td>Microscopically, the tumor was composed of masses of rather large amorphous cells that were sometimes elongated. They had acidophilic cytoplasm and many were vacuolated. An occasional spider-web cell was found. In the cytoplasm of some cells, elongated fibrils were detected but there were no cross striations. The nuclei were all well-preserved and showed large nucleoli. There was no pyknosis, and mitoses averaged 22 in 50 high power fields.</td>
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<tr>
<th>Case 7</th>
<th>Elevated LDH and Uric acid</th>
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<tr>
<td>U.S. showed a mass with cecal distention and grade I hydronephrosis of the right kidney</td>
<td>Gross appearance: yellow, soft and rubbery in consistency and attached to many encapsulated nodules</td>
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<tr>
<td>CT showed a small mass measuring 7<em>4</em>3 cm rising from abdominal wall and adhering to cecum</td>
<td>Immunohistology was negative for CD3, CD19, CD20, LCA, CK, CHROMO, SYNAPTO, CD99, DOG1, MY0D1 but it came positive for DESMIN</td>
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</table>

**Figures**
Figure 1

A non-contrast MSCT of the abdomen (A) coronal view showing a well-defined irregular mass in the left lower quadrant. (B),(C) show a homogeneous mass with clear boundaries emerging at the expense of the abdominal wall and adhering to cecum.
Figure 2

(A) and (B) **H&E stain of the mass**: The figure exhibits fibrous septa enclosing alveolar nests of neoplastic small blue round cells. Neoplastic cells can be seen in the lumen nests. (C) Desmin immunostaining showing positivity. (D), (E), (F) Immunostaining for ck, MYOD1 and CD99 ordinally showing negativity.
Figure 3

Bone scan with Tc-MDP (A) Anterior view (B) posterior view. Showing increased tracer uptake around the orbit, multiple ribs bilaterally, Vertebrae, inferior angle of left scapula, manubrium sterni with secondary neoplastic skeletal changes (metastasis).