

# Giant Primary Omental Leiomyosarcoma Mimicking Ovarian Malignancy

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## ABSTRACT

Primary omental leiomyosarcoma is a rare omental malignancy. Its usual manifestations are abdominal pain, abdominal mass and abdominal distension with or without constitutional symptoms. Primary omental leiomyosarcoma varies in its dimensions from being a small-sized leiomyosarcoma to a giant one and can mimic any huge intra-abdominal and pelvic malignancy. We are reporting a case of a 20-year old unmarried female who presented with a giant intra-abdominal mass occupying nearly entire abdomen. Clinically, it was difficult to comment on its site of origination. Abdominal

ultrasonography and CT scan suggested that this heterogeneous tumor was arising from the left ovary. On exploratory laparotomy, the mass was found to be a giant primary omental tumor with no synchronous lesion. The tumor was successfully excised with total omentectomy. Histopathology and immunohistochemistry of the specimen was consistent with omental leiomyosarcoma. Owing to the absence of any synchronous intra-abdominal lesion, the tumor was labeled as primary omental leiomyosarcoma.

**Keywords:** Primary Omental Leiomyosarcoma; Leiomyosarcoma of Gastrocolic Omentum; Omental Tumors

## INTRODUCTION

Although secondary omental leiomyosarcomas from secondary deposits of leiomyosarcomas originates from mesenchymal tissue of the gastrointestinal tract, uterus and retroperitoneum, primary omental leiomyosarcoma (POL) is an extremely rare omental tumor as evident from its limited citations in the biomedical literature [1]. Although no age is exempt, POL usually afflicts the middle-aged population in their 5<sup>th</sup> and 6<sup>th</sup> decades of life with no predominant sex predilection [1-3]. Because of its extreme rarity and scarcity of long-term statistical data, prevalence of POL is not yet precisely known [2, 3]. POL usually presents as a solid abdominal mass of varying sizes. Occasionally, it may achieve enormous dimensions and masquerade intraperitoneal and retroperitoneal soft tissue sarcomas or a huge ovarian malignancy like that of our patient. The conundrum of its extreme rarity and varied morphology of POL can pose perplexing diagnostic challenges even to experienced clinicians in differentiating it from abdominal, retroperitoneal and pelvic tumors.

Even high-tech imaging modalities fail to determine the exact preoperative nature and site of origination of huge POL. Accurate diagnosis of POL can only be arrived after painstaking efforts with histological and histochemical evaluation of the surgically resected specimen. The prime rationale of reporting this case scenario is to acquaint the health-care professionals with clinical-biological behaviour, diagnostic work-up, and therapeutic strategies of this rare omental malignancy.

## CASE REPORT

A 20-year old unmarried female presented with gradually enlarging painless abdominal mass in association with abdominal discomfort and distention of one year duration. She denied alterations in her bowel habits or any genitourinary symptoms. Her general physical and systemic examination revealed marked anemia, asthenia, dehydration, malnutrition, and cachexia to the extent that she was unable to walk without support of two persons. Her abdomen was grossly distended. On palpation,

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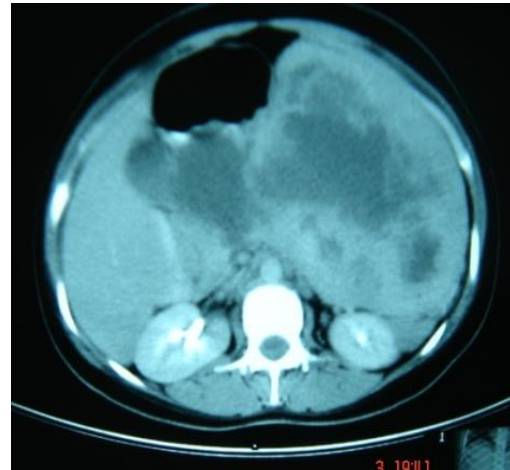
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there was a huge intra-abdominal solid mass that was filling nearly whole of the peritoneal and pelvic cavities. Her digital anorectal examination was normal. Because of its prodigious proportions, it was clinically well-nigh impossible to comment on its site of origination. Laboratory work-up showed hypochromic and microcytic anemia and normal biochemical profile. Her serum CA-125 level was within normal range. Abdominal sonography and CT scan revealed a huge heterogeneous intraperitoneal tumor consisting of hyperattenuating solid areas interspersed with hypoattenuated cystic spaces. Because of its colossal size, it was altogether difficult to measure its precise dimensions. Sonographic and tomographic evaluation imparted the impression of a heterogeneous giant left ovarian tumor having solid and cystic components with no ascites, mesenteric lymphadenopathy or hepatic metastases. Intravenous urography revealed moderate degree of left-sided hydronephrosis and hydronephrosis. Chest X-ray was devoid of any cardiopulmonary pathology or metastases. Based on the indeterminate results of clinical, radiological, sonographic and tomographic evaluation, the decision was taken in favor of open exploration through a midline laparotomy incision. On exploration, we found a massive fleshy multi-nodular tumor of the gastro-colic omentum having solid and cystic spaces filled with straw-colored fluid. The mass was filling almost whole of the peritoneal cavity and was encroaching even the pouch of Douglas. The tumor was extremely vascular and was being fed with dilated feeding vessels emanating from the gastroepiploic vascular arcade. The tumor was abutting the greater curvature of the stomach. Resultantly, a strip of greater curvature was excised to ensure curative resection of the tumor. Histopathological evaluation of the tumor was compatible with omental leiomyosarcoma. Because of absence of synchronous intraabdominal lesion, the tumor was labeled as POL. On immunohistochemistry, the tumor histologic sections displayed striking immunoreactivity for smooth muscle actin (SMA) and desmin and negatively for c-kit (CD117) and CD34. Postoperatively, she received adjuvant chemotherapy (adriamycin, ifosfamide, dacarbazine and mesna) and was doing well one year after surgery.

## DISCUSSION

Exact aetio-pathogenesis of POL is debatable.

**Figure 1:** CT features of POL



**Figure 2:** Intraoperative features of POL

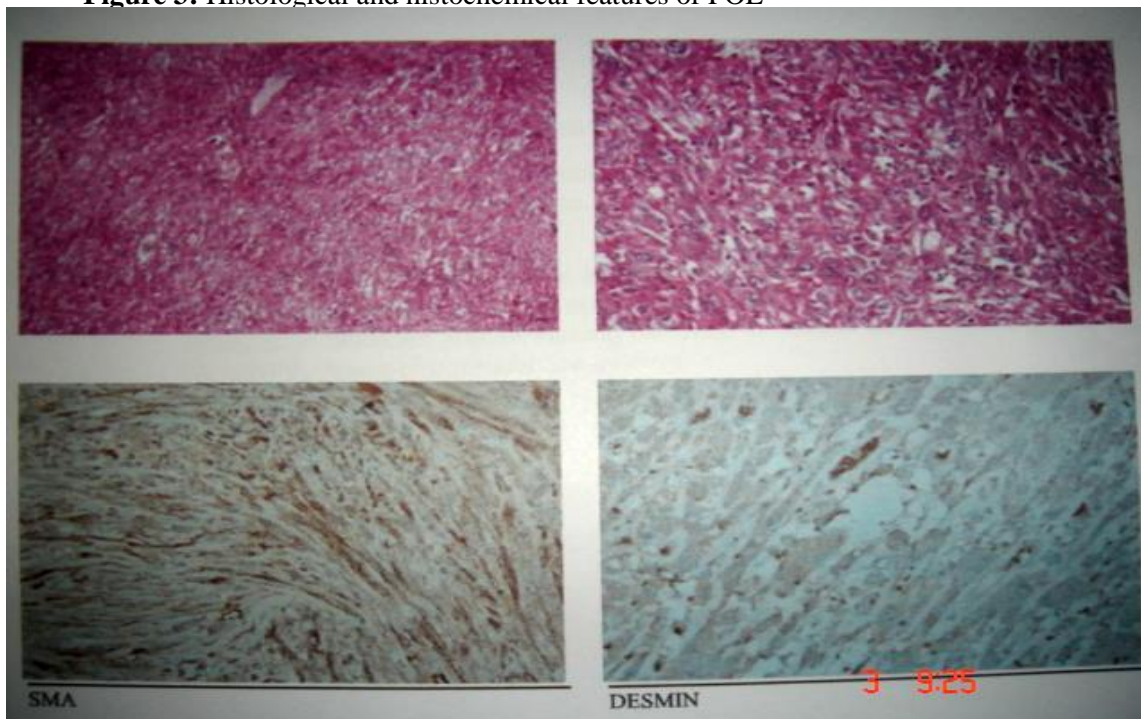


Histologically, the greater omentum is a double-layered structure consisting of trabecular network of connective tissue containing adipose, vascular and lymphatic elements in addition to mesothelial and smooth muscle cells, fibroblasts, pericytes and lipocytes. Accordingly, a wide-spectrum of primary omental tumors like fibroma, lipoma, leiomyoma, liposarcoma, neurofibroma, fibrosarcoma, liposarcoma, leiomyosarcoma, mesothelioma, haemangiopericytoma and gastrointestinal stromal tumours exists [4-6]. Clinical manifestations of POL are entirely non-specific and non-pathognomonic and provide no clue(s) whatsoever to clinch its preoperative diagnosis. In vast majority of cases, POL often remains asymptomatic and is discovered incidentally during abdominal imaging for an unrelated gastrointestinal problem. Common manifestations of POL manifests are abdominal mass (35%), abdominal discomfort (35%) and abdominal distention (15%) with or without constitutional symptoms such as nausea, vomit-

-ing, anorexia, anemia, asthenia, and cachexia. On abdominal palpation, a palpable POL appears as a fleshy, multinodular, solid mass. A giant POL may mimic a huge intraperitoneal or retroperitoneal soft-tissue sarcoma or a massive ovarian malignancy. Abdominal ultrasonography (USG) is the first non-invasive imaging technique commonly used for evaluation of the abdominal, pelvic or retroperitoneal mass(s). USG displays POL as a heterogeneous mass with hypochoic, anechoic and hyperechoic areas. Nonetheless, CT scan abdomen is considered as gold-standard imaging for evaluation of such lesions. The CT features of POL include heterogeneous, multi-lobulated tumor having hyper-attenuating solid areas intermingled with hypo-attenuated cystic spaces. The preoperative role of fine-needle aspiration cytology or core biopsy in establishing tissue diagnosis of POL is not yet established. The exact diagnosis of POL can only be achieved on immaculate histological evaluation of the resected specimen followed by immunohistochemistry. On immunohistochemical staining, POL displays strong positivity for smooth muscle actin (SMA) and desmin and negativity for c-kit (CD117) and CD34 [7-9]. Complete surgical excision (total omentectomy) is the mainstay of treatment of POL. Even in the presence of secondary peritoneal implants, total omentectomy still

improves the survival figures. Now-a-days, laparoscopic resection of POL is being carried out in advanced laparoscopic centres. Undoubtedly, laparoscopic resection has proven supremacy over its conventional counterpart; not only in being minimally invasive with speedy recovery and excellent cosmesis but also in providing golden opportunity for scrupulous visualization of the whole celomic cavity for exclusion and simultaneous management of any synchronous lesion(s) detected in the index surgical session [10]. POL is usually non-responsive to chemo-radiotherapy. However, combination of anti-cancerous chemotherapy, such as MAID (Mesna, Adriamycin, Ifosfamide, and Dacarbazine), is being tried with conflicting results. According to the global literature, POL has been associated with dismal prognosis [8-10]. In conclusion, POL is an uncommon omental malignancy that can pose diagnostic challenges in differentiating it from other intra-abdominal or retroperitoneal soft tissue sarcomas or uterine and ovarian tumors. Exact preoperative diagnosis of POL is hardly possible even with sophisticated imaging modalities. An irrefutable diagnosis of POL is only possible after its complete surgical excision followed by painstaking efforts during histopathological and histochemical evaluation of the resected specimen.

**Figure 3:** Histological and histochemical features of POL



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