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Epithelioid Leiomyosarcoma of Greater Omentum- Case Report of a Rare Tumour

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Abstract

A 50 year old man presented with a large, circumscribed mass in the abdomen. With ultrasonography, computed tomography, endoscopy and FNAC, preoperative diagnosis could not be made. Complete resection of the mass was performed; the tumor weighed 10.5 kg. Immuno-histochemical study confirmed the diagnosis of epithelioid leiomyosarcoma (ELMS). These are generally solid tumours commonly arising from the stomach. It is uncommon in the greater omentum.

Key Words: Leiomyosarcoma; Leimyoblastoma; Greater omentum

Introduction

ELMS is a rare tumour derived from smooth muscle cells. In 1960, Martin *et* al, described a new gastric smooth muscle tumour [1]; in 1976 a series of 69 cases of similar tumour was published by Stout[2] and they were called as leiomyoblastoma. Appelman *et al* reclassified them as epithelioid leiomyomas and epithelioid leiomyosarcomas according to their histological appearance and clinical behaviour[3]. ELMS occur commonly in the stomach [4], rarely in the small bowel, colon, uterus and retroperitoneum. They are multinodular, polypoidal and solid masses [5,6].This is a rare case of epithelioid leiomyosarcoma arising from the greater omentum, weighing 10.5 kg , the largest reported till now; correct diagnosis was made only after the immunohistochemical studies.

Case report

A 50 year old man presented with distension of the abdomen and repeated vomiting since 2 months; Patient was anemic (Hb-6gm%). Abdomen was grossly distended due to a large mass occupying all the quadrants; it was firm in the right flank and cystic in the left flank. Sonography revealed a large intra abdominal mass measuring 36.8 x 30.2 cm, with solid and cystic areas. CT abdomen was reported as 31.3 x 26.0 19.5 cm mass with solid and cystic components (fig-1) suggestive of Mucinous cystic neoplasm of the pancreas. Endoscopy revealed a bulge in posterior wall of stomach, due to extra gastric mass. FNAC was negative for malignant cells.

On laparotomy, a dark brown coloured large tumor, with solid and cystic areas, arising from the greater omentum was found. Dilated veins were running over the surface of the mass. The tumor was easily separated from the surrounding viscera. A branch of the right gastroepiploic artery was feeding the tumor.There were no metastasis in the liver, peritoneum and lymph nodes. 6 litres of dark brown colour fluid was aspirated from the cystic portion of the tumour. The tumor was Completely excised. Post operative recovery was uneventful.

The tumor measured 35.0 x 28.0 x 20.0 cm and weighed 4.5 kg. (fig-2). The surface was bosselated; cut section revealed nodular greyish white solid tumor with large cystic areas. Microscopy revealed neoplasm consisting of round to oval cells having pink cytoplasm and centrally placed hyperchromatic nuclei. Immuno histochemical studies revealed that the tumor cells expressed vimentin, desmin and SMA, suggestive of Epithelioid leiomyosarcoma.

Figure 1: CT image of the tumour

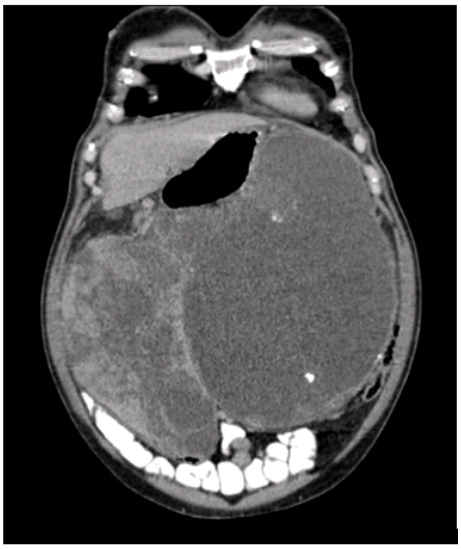
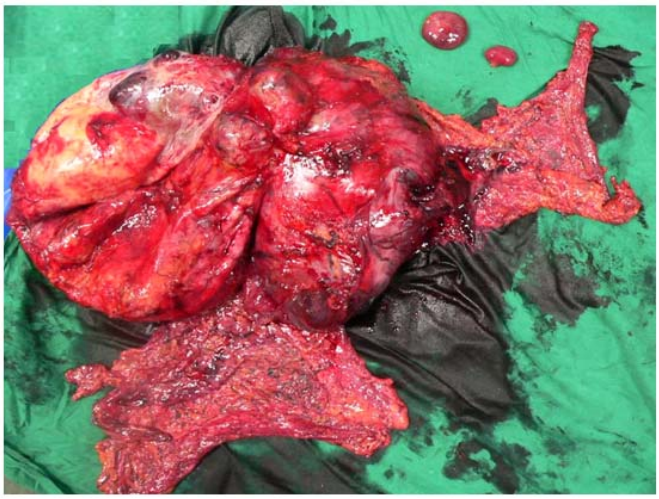


Figure 2: Tumour Specimen arising from greater omentum



Discussion

ELMS are more common in males than females [7,8]. ELMS of the greater omentum is a rare occurence. They are usually large and whitish grey masses with areas of haemorrhage and cyst formation. The patient may present with an abdominal mass, weight loss, fever nausea and vomiting. Intraperitoneal haemorrhage can be a rare mode of presentation[3,8]. The tumours usually grow to a large size, ranging from 6 x 3 x 2 cm to 30.0 x 20.0 x 13.0 cm. In this case , the tumour measured 35.0 x 28.0 x 20.0 cms and weighed 10.5 kg (4.5 kg solid tumour mass + 6 L aspirated fluid); this is perhaps the largest ELMS reported in current english literature till now [9].

ELMS are common in the stomach (90%); they may develop in any part of the gastrointestinal-tract , uterus, retro peritoneum, skin, vulva and neck [8].They should be differentiated from schwannomas, neurofibrosarcomas, liposarcomas, malignant fibrous histiocytomas and adrenal cortical carcinomas. The large, round or polygonal cells with acidophilic, vacuolated cytoplasm, known as epithelioid cells is the characteriastic finding of this tumour [8].

Ultrasound, computed tomography (CT] and endoscopy are useful to define the morbid anatomy[3].However ,it is difficult to make an accurate preoperative diagnosis. The tumour is malignant. Metastasis occur in 63% of the ELMS in two years [3]. Three fourth of them have metastases at the time of the surgery [8] Mitotic figures (> 10 / 50 HPF means 100 % metastasis) and size (> 6 cm) of tumour usually determines the malignant potential[8]. The liver and peritoneum are the most common sites of metastases. The lymph node and distant metastasis are rare. ELMS patients may have a long term survival when a successful resection is performed even when a metastasis is present. A case of gastric ELMS with multiple liver metastasis at the time of diagnosis, has survived for 36 years after gastrectomy[6].Another case of ELMS with lymph nodes and liver metastasis is reported with a follow-up of 18 years[10].

Although some authors advocate chemotherapy and / or radiotherapy, curative surgical resection is the mainstay of the treatment [4,8].

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