Gastric ulcer that turned out to be metastasis of a synovial sarcoma: A case report and literature review

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SUMMARY

Primary gastrointestinal synovial sarcoma or its metastases to the gastrointestinal tract is rare. Here we present a case of 56-year-old gentleman with left thigh synovial sarcoma and gastric metastases along with the literature review.

KEY WORDS:
Gastric ulcer, synovial sarcoma, stomach, metastasis

INTRODUCTION

Synovial sarcoma is a malignant soft tissue neoplasm that may present itself from a variety of sites in a human body commonly near large joints or tendons (80%). The nomenclature however is misleading as there is no evidence of differentiation towards synovium and this tumour can actually occur in any part of the body.[1] Most synovial sarcomas of the monophasic type are composed entirely of spindle cells and can hardly be associated with synovium in terms of morphology.[1] Gastrointestinal synovial sarcomas are rare but more and more cases are being reported each year increasing awareness about the potential misdiagnosis between the monophasic synovial sarcoma and KIT-negative gastrointestinal stromal tumour (GIST).[2] To our knowledge only about 27 cases of stomach related synovial sarcoma are previously reported. Here we present a case of a metastatic gastric synovial sarcoma, the first reported in Malaysia.

CASE REPORT

A 56-year-old gentleman presented with a mass over the left posterior upper thigh for three months. Initially small but progressively increasing in size (20x15cm) with no skin changes, mobile and non-tender on palpation. Magnetic resonance imaging showed a large heterogeneous left upper thigh mass with malignant features (capsular breach and intramuscular extension). Biopsy of mass showed to be synovial sarcoma as and it was resected completely with clear margins. Histopathological examination reported a 9.5cm tumour and initial Contrast Enhanced Computed Tomography (CECT) staging showed no enlarged lymph nodes or any distant metastasis (T2bNoMo). Patient later underwent radiotherapy 66Gy for 33#. Patient did not have any active complaint post adjuvant radiotherapy however repeated CECT Thorax Abdomen and Pelvis one-year post resection showed lung metastases and focal mucosal thickening of the stomach. Patient was then planned for an Oesophagogastroduodenoscopy (OGDS). OGDS done and revealed a malignant looking ulcer with rolled up edges and fungating in nature at the incisura (Figure 1). Ulcer edge biopsied and result of biopsy was metastasis of synovial sarcoma. Immunohistochemical staining done and it was stained positive with vimentin, cytokeratin, epithelial membrane antigen (EMA) and BCL2 antibodies but were negative for KIT, CD34, CD99, desmin, DOG 1 and SMA (Figure 2). No molecular genetic study was done as it is unavailable in our centre. He was given six cycles of Doxorubicin however repeated CECT post chemotherapy showed increasing size of bilateral lung nodules and stomach mass along with new liver lesion suspicious of liver metastasis. Currently patient was advised for second line palliative chemotherapy but still undecided as he is asymptomatic.

DISCUSSION

Synovial sarcomas are malignant mesenchymal tumours that commonly arise in the limbs1 and sometimes in other internal organs. The typical synovial sarcoma has a biphasic growth pattern and commonly occurs near joints thus it was mistakenly deemed as a tumour of synovial differentiation. However monophasic fibrous synovial sarcomas are composed exclusively of spindle cells and this may result in misdiagnosis if inadequate immunohistochemical and/or genetic studies are carried out. Immunohistochemically synovial sarcomas are often focal reactive to cytokeratins and epithelial membrane antigen.[1] This proves presence of epithelial differentiation rather than synovial differentiation.

When a spindle cell gastrointestinal tract tumour is encountered in the lab the common preliminary diagnosis would be a GIST. However, the pathologist can differentiate a GIST from synovial sarcoma as DOG 1 antibody is sensitive to GIST. Leiomyosarcomas and malignant spindle cell melanomas are characterised by a higher degree of pleomorphism and a panel of smooth muscle markers and melanocytic markers may exclude these diagnoses. Sarcomatoid carcinoma exhibits conspicuous pleomorphism a higher level of epithelial markers and is usually accompanied by conventional carcinoma. The common mimickers of synovial carcinomas are malignant peripheral nerve sheath tumour and fibrosarcoma need to be ruled out as well. The synovial sarcoma is also known to mimic an Ewing's sarcoma or malignant solitary fibrous tissue.[1] In brief the morphology and immunoprofile can often differentiate these mimickers from synovial sarcomas but at times

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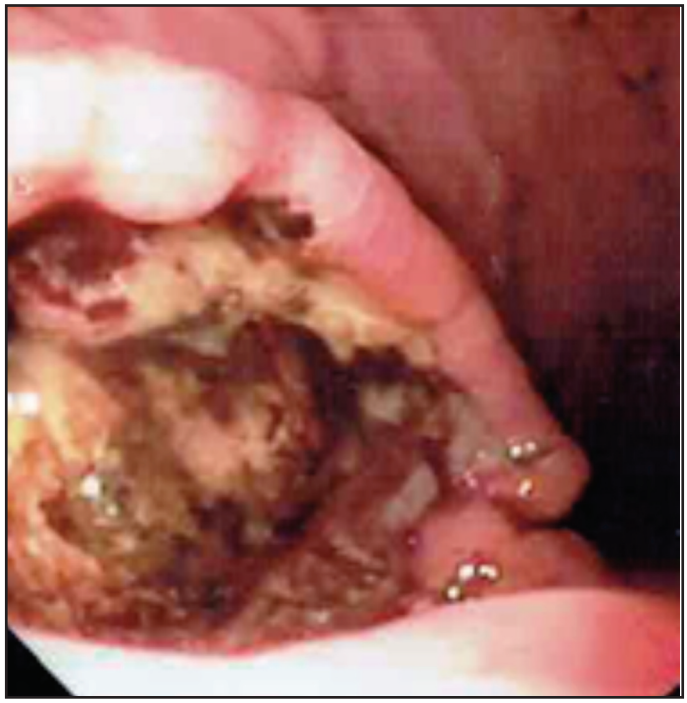

Fig. 1: Malignant looking ulcer with rolled up edges and fungating in nature at the incisura.

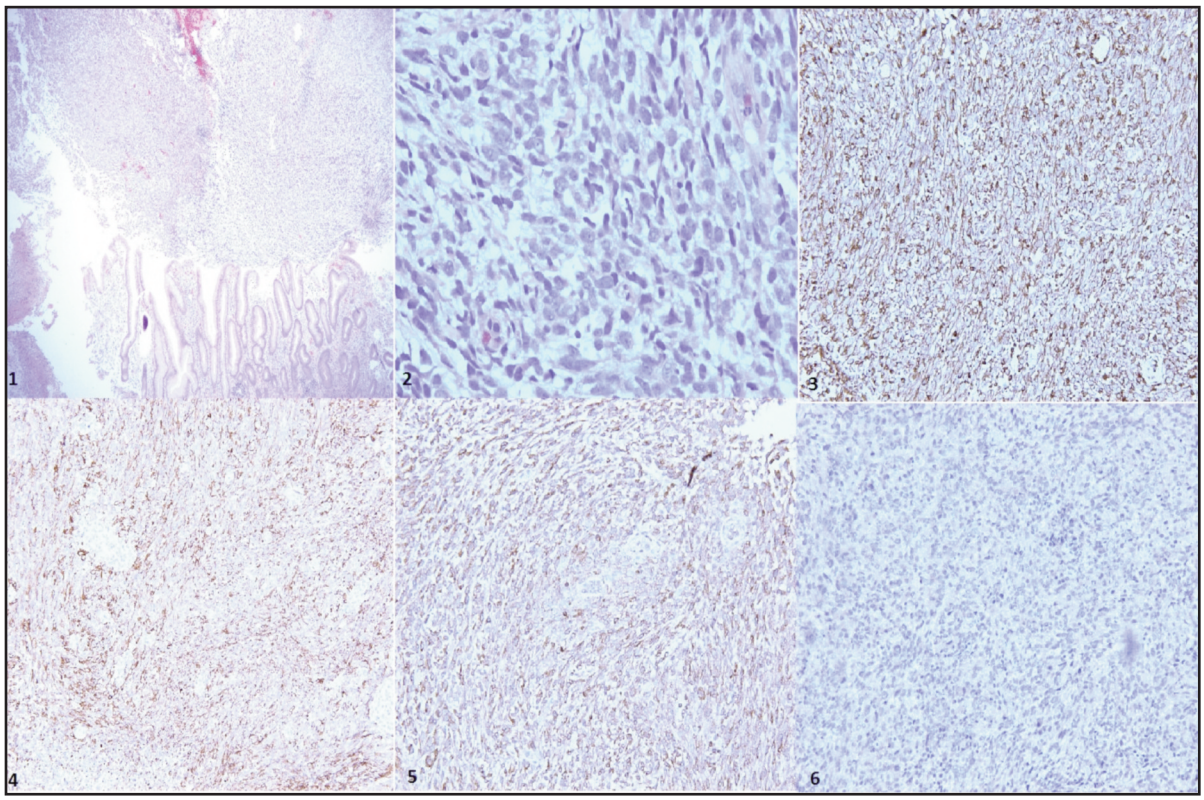

Fig. 2: Histopathological examination: 1. Low power microscopic picture (12.5x) noted spindle cells; 2. High power microscopic picture (40x) noted pleomorphic cells; 3. Vimentin – tumor cells shown positive immunoreactivity by giving a brown appearance that indicate mesenchymally derived cells undergoing an epithelial-mesenchymal transition usually during metastatic progression; 4. EMA – tumor cells shown positive (brown) stain with epithelial membrane antigen proving presence of epithelial differentiation; 5. BCL 2 – tumor cells exhibit overexpression of bcl-2 protein by staining positive (brown) with BCL2 antibody; 6. DOG 1 – negative stain rules out GIST as possible diagnosis.

molecular genetic study (SYT-SSX2 or SYT-SSX1) may be required for confirmation in difficult cases. This proves a challenge for developing countries with limited resources and may lead to misdiagnosis.

The main treatment for synovial sarcoma is surgical resection with/without radiotherapy to enhance local control. Chemotherapy (typically Doxorubicin and/or Ifosfamide) might be recommended in the treatment especially in advanced/metastatic disease.[2] Due to the rarity of this malignant tumour there is still no consensus from experts on how much the role of chemotherapy plays in preventing metastases and improving survival.

The prognostic factors in a synovial sarcoma patient are influenced by the quality of surgery (clean margins) and the characteristics of the disease (size of tumour, local invasion, histology subtype, presence of metastases and lymph node involvement). There is excellent prognosis for patients with small tumour that managed to be resected with clear margins. Patients with tumours more than 5 cm tend to have higher risk in developing distant metastases.[3] Patients with poorly differentiated subtype and those already having metastases tend to have a poor prognosis.

CONCLUSION

Synovial sarcoma in the digestive tract is rare and prone to misdiagnosis. The accurate and early diagnosis is crucial for effective and appropriate therapy. The potential clinical benefits for a patient with metastatic synovial sarcoma to undergo another resection for gastric metastases are still unclear. More cases should be reported in order to study its disease pattern and prevalence as only then clinical practice and management guideline for this malignant disease may be established.

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