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CASE REPORT

A RARE CASE OF INTESTINAL OBSTRUCTION SECONDARY TO METASTASIS DERMATOFIBROSARCOMA PROTUBERANS

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue sarcoma, which arises from the dermis. It behaves as a low to intermediate-grade malignancy, is locally aggressive, and frequently has local recurrence but rarely metastasize. The most common organ of metastasis reported is the lungs. Here we present a rare case of metastasis dermatofibrosarcoma protuberans (DFSP) that presents with intra-abdominal mass causing intestinal obstruction. Exploratory laparotomy and en-bloc resection of the tumor were done followed by adjuvant chemotherapy.
***Keywords**: Dermatofibrosarcoma protuberans, Metastasis, Intestinal obstruction*

INTRODUCTION

Dermatofibrosarcoma protuberans is a rare slow-growing fibrohistiocytic, intermediate-to low-grade malignancy. It accounts for approximately 0.1% from all cancers and 1-6% of soft tissue sarcoma (1-3). It usually occurs in young to middle-aged individuals and commonly affects trunk, proximal extremities, head, and neck (1,4). Most cases present with slow-growing bluish or brownish erythematous skin nodules. It could also present as a keloid scar (1,5). DFSP typically arises in the dermis, has indolent growth but could be locally aggressive as it spreads into the subcutaneous tissue and muscles (1,2). It rarely has distant metastasis with the lung the most common site of metastasis. Intra-abdominal metastasis is rare (1,2,4,5). Treatment for DFSP is either wide local excision or Mohs surgery. It frequently recurs locally in cases of incomplete excision with a recurrence rate of up to 53% being reported (5,6).

Clinical presentation

We report a 41-year-old female with a previous history of dermatofibrosarcoma protuberans of the left shoulder 1 year before the current presentation. Her initial presentation was a mass over the left shoulder progressively increasing in size over 4 months with core biopsy consistent with DFSP. There were no distant metastases on imaging. She underwent wide local excision of the tumor and histopathology reported tumor margin of less than 3 cm. Adjuvant radiotherapy was planned but she defaulted due to logistic reasons. Her current presentation is an intestinal obstruction for 3 days in December 2018. Examination revealed intra-abdominal mass measuring 15 cm x 15 cm over the left flank. The prior surgical wound over the left shoulder was well-healed with no evidence of local recurrence. Abdominal x-ray showed dilated proximal small bowel.

Contrast-enhanced CT thorax, abdomen, and pelvis showed a well-defined intra-abdominal mass measuring 15x 17x 20 cm causing intestinal obstruction and evidence of lung metastasis (Figure A and B). She subsequently underwent laparotomy and tumor debulking. Intraoperative findings showed a multilobulated soft tissue tumor measuring 20cm x 20cm along the mesenteric plane with extension into the retroperitoneum. Complete excision of the tumor was achieved. The tumor displaced the descending colon and retroperitoneal structure medially and caused external compression on the small bowel. The intestine and its associated vascular trunks were preserved.

Histopathological examination of the tumour showed a fairly circumscribed and unencapsulated tumour composed of fibroblastic spindle-shaped cells arranged in a herringbone pattern, with a mitotic index of 6/10 HPF. Immunostaining was positive for CD 34 (Figure E, F, and G). The histopathology assessment was consistent with metastatic high-grade fibrosarcoma.

Post-surgery she was started with intravenous chemotherapy ifosfamide and doxorubicin for 4 cycles. Her condition did not improve post chemotherapy and she developed tumor recurrence.

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Subsequent CT scan showed a recurrent left intra-abdominal mass measuring 5cm x 7cm x 5cm (Figure C and D). Managed as recurrent metastatic intra-abdominal DFSP, she was planned for second line chemotherapy with gemcitabine and docetaxel. Her condition continued to deteriorate due to disease progression. She was not able to undergo the second line chemotherapy and subsequently succumbed to disease progression.

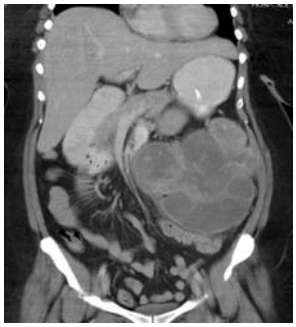

Figure A

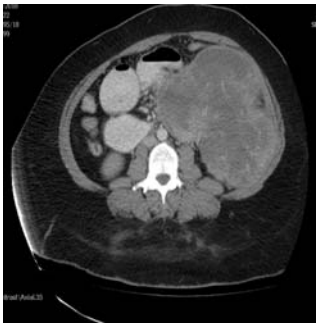

Figure B

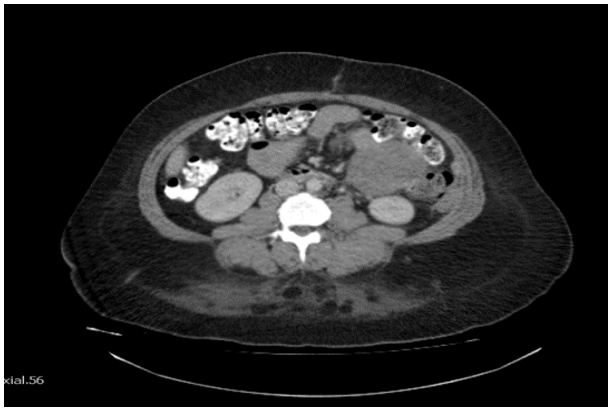

Figure C

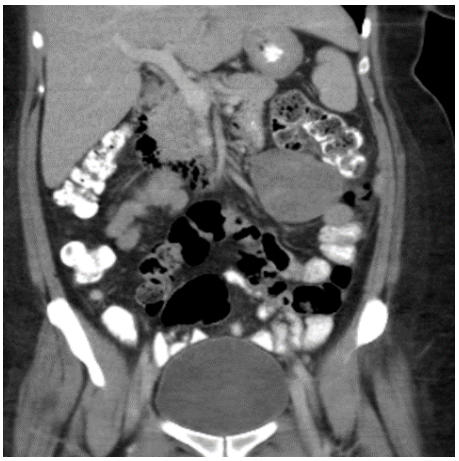

Figure D

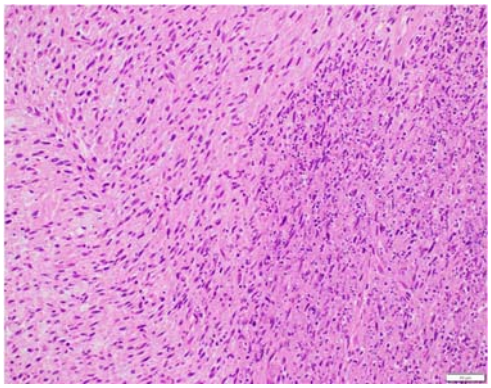

Figure E: Spindle tumor cells arrange in Herring Bone pattern adjacent necrosis (arrow).

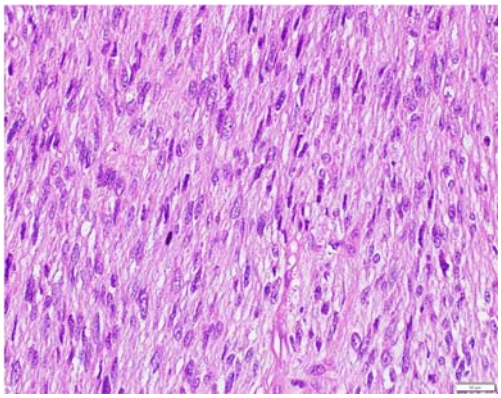

Figure F: The spindle tumor cells are pleomorphic having hyperchromatic nuclei with presence of mitosis, mitotic index of 6/10 HPF(arrow)

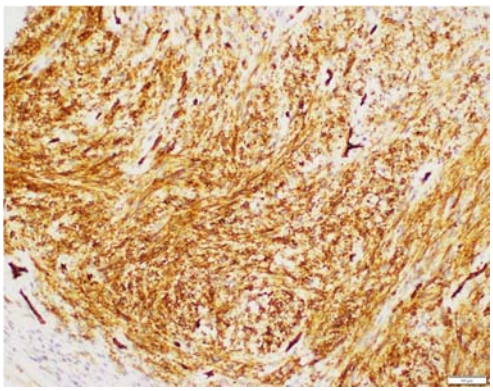

Figure G: CD34 immunostain highlights the tumor cells in brown colour.

Figure A & B: showed heterogenous intra-abdominal mass occupying over the lower left side of the abdomen, that possible arising from proximal jejunum or descending colon.
Figure C & D: Repeated CT showed a recurrent new lesion of intra-abdominal mass over the left side abdomen, possibly the location near the previous lesion. The lesion was smaller in size compared to the previous.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a mesenchymal neoplasm that typically involves both dermis and subcutaneous tissue (1,7). It commonly occurs in the trunk followed by the extremities, head, and neck (3,5). The lesion is usually painless, has indolent growth but is locally invasive with invasion into the underlying fascia, muscles, or bones (1,4,7). Even though this tumour is aggressive locally, distant metastasis is not common with less than 5% reported cases in the literature (6). It has hematogenous spread, typically to the lungs. Cases of metastases to the retroperitoneum, mediastinum, bones, the kidney, brain, omentum, scalp, ovaries, liver, and heart have been reported (4,8).

It is difficult to diagnose DFSP since the early clinical symptoms are non-specific, it is slow growing and mimics another non-malignant tumour such as dermatofibroma. Dermatofibroma appears similar clinically and is distinguished from DFSP by the absence of extension to deeper structure and the size of the lesion (3,5). The standard diagnosis of DFSP is by tissue biopsy with histopathological and immunohistochemical assessment. Imaging is for the assessment of extension to the deeper and surrounding structures as well as for operative planning. Computed tomography and MRI are both acceptable options, but MRI provides a better assessment of the tissue infiltration and depth of involvement. It is also useful for preoperative and post-operative evaluation (5).

The histological features in DFSP are characterized by spindled cells arranged in a distinct herringbone or storiform pattern and immunohistochemical staining positive for CD-34 (1,2,4,7). There are several histological variants of DFSP that have been described including myxoid, pigmented, atrophic, giant cell fibroblastoma (GCF), and DFSP with fibrosarcomatous change (5). Dermatofibrosarcoma protuberans with fibrosarcomatous areas (DFSP-FS) is recognized as a high-grade type of variant, with higher rates of local recurrence and potential for distant metastasis. This case presentation is most consistent with the DFSP-FS subtype (2). The diagnosis of DFSP-FS is based on Enzinger and Weiss's criteria. It includes the presence of fibrosarcomatous changes of more than 5 mitoses/10 HPF, fascicular growth pattern, increased cellularity, and atypia in at least 5% of the tumor tissue (9).

The standard treatment for DFSP is wide local excision with a margin of more than 3 cm. The alternative approach includes Mohs micrographic surgery (MMS) which requires immediate microscopic examination of the margins in order to ensure a tumor-free margin (1,3,5,7). The recurrence rate associated with MMS is less than 2% with no reports of distant metastasis (2,10).

MMS applies systematic horizontal sectioning compared to the traditional method which applies vertical sections which only assess limited tumor margin. In MMS, all sides of tumour are assessed using a frozen section which allows for a complete evaluation of tumour margins (11).

DFSP is a radiosensitive tumour and indication for radiotherapy includes the margin-positive tumour, unresectable tumour, or recurrent tumour (5). Tyrosine kinase inhibitor such as Imatinib, Sunitinib, and Sorafenib has been shown to induce regression of DFSP and has been applied clinically in recurrent, metastatic, or advanced diseases (5,7). The response rate of tyrosine kinase inhibitor in this clinical scenario of distant intra-abdominal metastasis is unknown as the efficacy of tyrosine kinase inhibitor in DFSP is only proven in the adjuvant setting after resection of primary high-risk tumors (12). Conventional chemotherapy has a limited role in the treatment of DFSP and is associated with poor response rates and clinical outcomes (5). Doxorubicin and ifosfamide for five or six cycles are the common regimes that are applicable in DFSP (5). In regards to this case, it is one of the rare cases of aggressive DFSP-FS that presents with intestinal obstruction due to intra-abdominal DFSP-FS metastasis. We, therefore, advocate a close follow-up protocol in all cases of DFSP which not only leads to a higher rate of compliance to adjuvant treatment but also provides a platform for early detection of possible tumor recurrence.

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