Sultan Qaboos University Med J, August 2012, Vol. 12, Iss. 3, pp. 371-374, Epub. 15th Jul 12
Submitted 30th Jun 11
Revision Req. 1st Nov 11, Revisions recd. 7th Dec 11th & 27th Mar 12
Accepted 11th Apr 12

INTERESTING MEDICAL IMAGE

Recurrent Dermatofibrosarcoma Protuberans of the Shoulder with Rare Distant Abdominal Metastasis detected by Fluorodeoxyglucose-Positron Emission Tomography/Computed Tomography (FDG-PET/CT)

*Ammad Al-Tamimi,[1] Sumbul Zaheer,[1] Chow Kah Hoe Pierce,[2] Saabry Osmany,[3] Kesavan Sittampalam[4]

ساركوما ليفية جلدية متكررة في الكتف مع انتشار نادر للبطن اكتشف عن طريق التصوير المقطعي بالاصدار البوزيتروني

عماد التميمي، سمبل زاهر، بييرس تشاو، صبري عثماني، كيسافان سيتمبالام

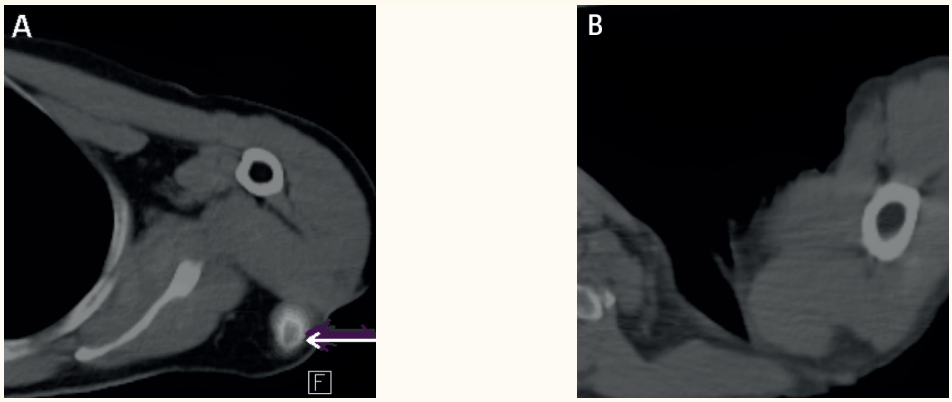

Figure 1: Fluorodeoxyglucose **(**FDG) positron emission tomography (PET)/computed tomography (CT) scans at the same trans-axial levels, showing mildly FDG-avid local recurrence in the shoulder (**A**), with no lesion subsequently identified in the same region (**B**).

A 56-year-old male patient with a rare *dermatofibrosarcoma protuberans* and fibrosarcomatous change (DFSP-FS) presented with a solitary fluorodeoxyglucose (FDG)-avid tumour with no evidence of metastasis in the left upper extremity [Figure 1A]. This initial presentation was treated with wide excision, and then again at its recurrence 8 months later. At admission, the patient had symptoms of gastritis and underwent an oesophago-gastro-duodenoscopy, which was normal. Almost 3 years after the initial presentation, the patient presented with a retroperitoneal mass measuring 1.4 x 0.8 cm. Investigation revealed the tumour, which was avid on FDG-PET/CT (standardised uptake value [SUV] max 11.3) and new compared to the previous study [Figure 2B]. Mild FDG uptake was also noted in a new pulmonary nodule (SUV max 1.9) and

Departments of [1]Nuclear Medicine & PET, [2]General Surgery, [4]Pathology, Singapore General Hospital, Singapore; [3]Radlink PET & Cardiac Imaging, Paragon Medical, Singapore.
**Corresponding Author e-mail: he77@hotmail.com*

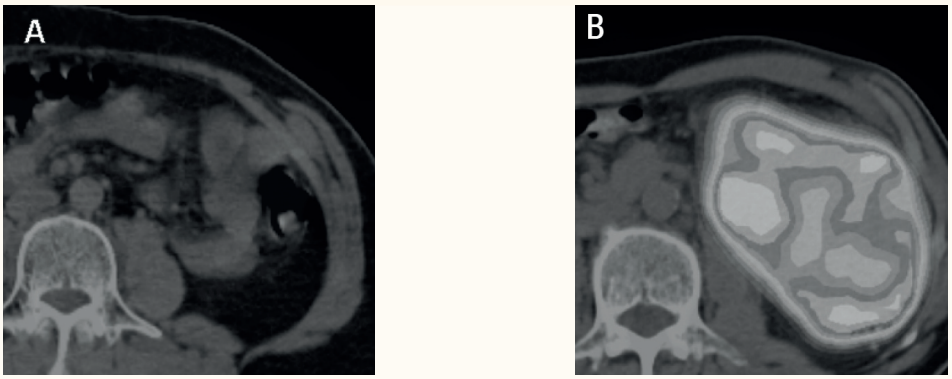

Figure 2: Fluorodeoxyglucose (FDG) positron emission tomography (PET)/ computed tomography (CT) scan at the same trans-axial level comparing the two scans at the time of local recurrence in 2006, when there was no retro-peritoneal lesion (**A**) and subsequently at the time of 1.4 x 0.8 cm heterogeneous, left retro-peritoneal distant metastasis in 2009 (**B**).

appeared malignant [Figure 3B]. Figure 4 shows the maximum intensity projection (MIP) images from the two studies. The retroperitoneal tumour was found to be *dermatofibrosarcoma protuberans* with extensive fibrosarcomatous transformation, histology grade 3 [Figures 5A and 5B].

DFSP is a rare cutaneous tumour of low malignant grade characterised by a pattern of slow, infiltrative growth and a marked tendency to recur locally after surgical excision.[1] DFSP is commonly found on the trunk (42–72%), occasionally in the proximal extremities (16–30%), and infrequently above the neck (10–16%).[7] Rare distant metastases (in less than 5% of cases) may occur many years after the onset of disease and are limited mostly to the lungs, followed by regional lymph nodes, while the visceral organs and bones are rarely affected.[2] Metastases are more likely with repeated incomplete surgical excisions and by tumour de-differentiation to higher grades.[6] Metastases are associated with local recurrence and poor prognosis. The 5-year survival rate is estimated at 99.2%.[7]

DFSP is a fibrosarcoma originating from dermal fibroblasts.[6] The genetic abnormalities in DFSP include a supernumerary ring chromosome related to the low amplification of sequences of chromosomes 17 and 22 which is a form of balanced reciprocal translocation.[8] This rearrangement will cause fusion of alpha chain type A (COL1A1) located on 17q22 to the platelet-derived growth factor beta (PDGFB) located on 22q13. As a result, the COL1A1-PDGFB gene formation will result in up regulation of PDGFB expression, resulting in continuous autocrine activation of PDGF receptor beta (PDGFR-B) and propagation of the mitotic signal by formation of autocrine and paracrine loops.[9] These transformed cells are inhibited by the tyrosine kinase inhibitor imatinib mesylate with

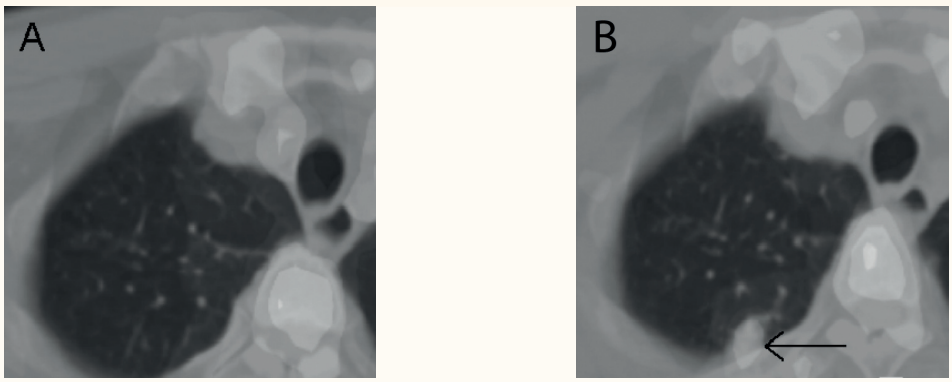

Figure 3: Fluorodeoxyglucose **(**FDG) positron emission tomography (PET)/computed tomography (CT) scan at the same trans-axial level comparing the two scans at the time of local recurrence in 2006 when there was no pulmonary lesion (**A**) and subsequently at the time of retro-peritoneal distant metastasis in 2009, when a mildly FDG-avid pulmonary nodule was detected (**B**).

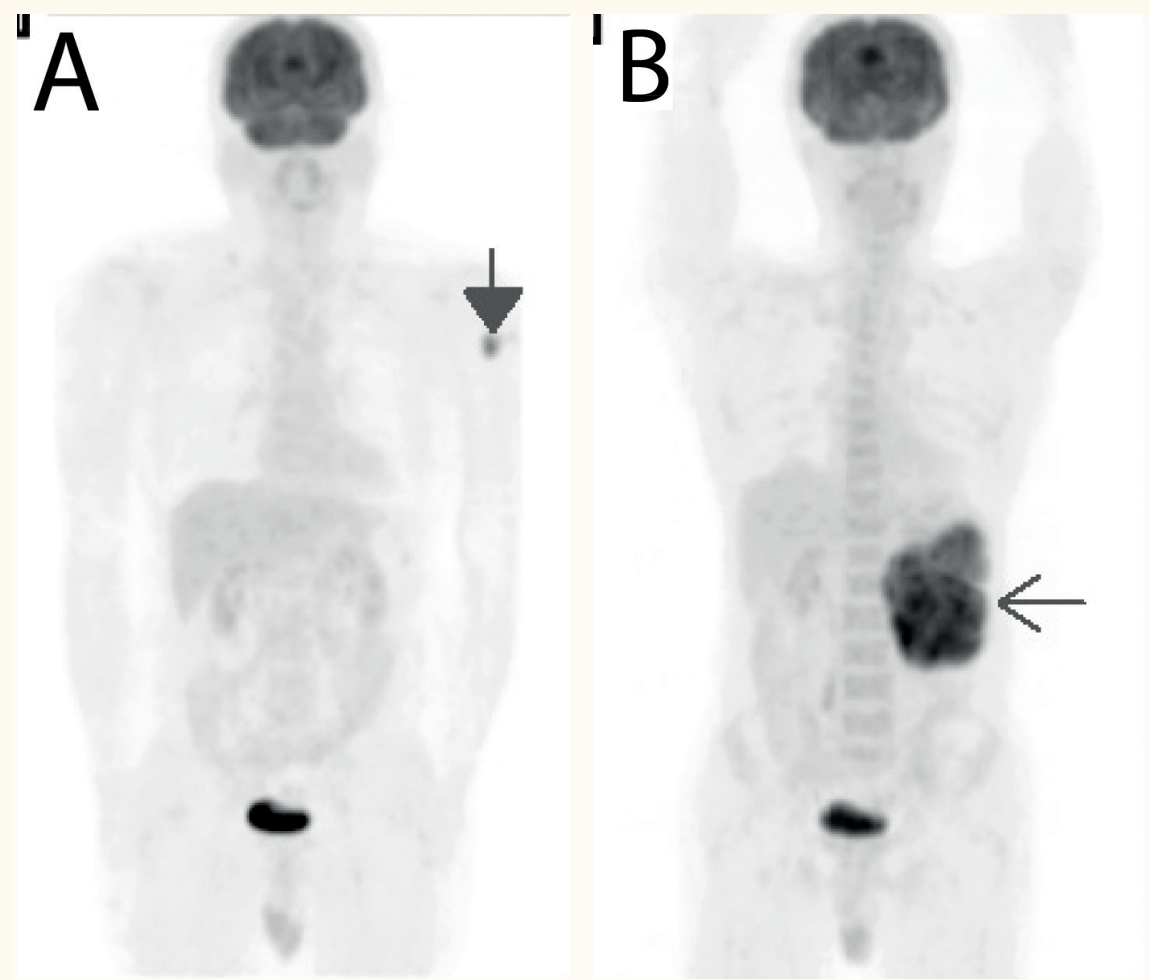

Figure 4: Maximum intensity projection of the fluorodeoxyglucose (FDG) positron emission tomography (PET)/computed tomography (CT) scans at the time of the first presentation in 2006 with (**A)** a mildly FDG-avid local recurrence, and (**B)** subsequently, in 2009, with an intensely FDG-avid retro-peritoneal metastasis.

reported cases of response often documented with FDG.[10]

The surgical objective in DFSP is complete tumour excision with maximal normal tissue preservation. Hence, a wide local excision with lateral margins of at least 3 cm and including the underlying fascia is recommended. In addition, micrographic controlled excision (MCE) showed favourable outcomes in treating DFSP.[6] FDG-PET/CT findings in a case of recurrence of dermatofibrosarcoma in the surgical scar have been reported.[3]

FDG-PET/CT has also been used to monitor response to imatinib mesylate in 2 reported cases of unresectable metastatic DFSP, with a decrease in tumour uptake of FDG after treatment documented in both cases.[4–5] In the former case, this decrease in uptake coincided with clinical improvement as early as 2 weeks after commencement of the therapy. [4]

In contrast to our case, where a FDG-avid pulmonary lesion was noted on PET/CT, a previous case reported a pulmonary nodule which was noted on CT but not on FDG-PET. It was thought this may have been due to the size (8 mm). A case has also been reported where DFSP response to imatinib was noted on FDG-PET but not on CT.[10] This is similar to findings in other malignancies.

DFSP is a rare cutaneous tumour of low malignant grade. FDG-PET/CT has been reported to be useful in DFSP imaging for delineating disease extent, both in staging and recurrence, which can be useful in planning surgery. FDG has also been documented as assessing DFSP response to imatinib, showing response when none was noted on CT. Given the rarity of the disease there are no large studies to date but the existing data appears promising. We suggest that FDG PET/CT be considered as an important component of the treatment plan of patients with DFSP.

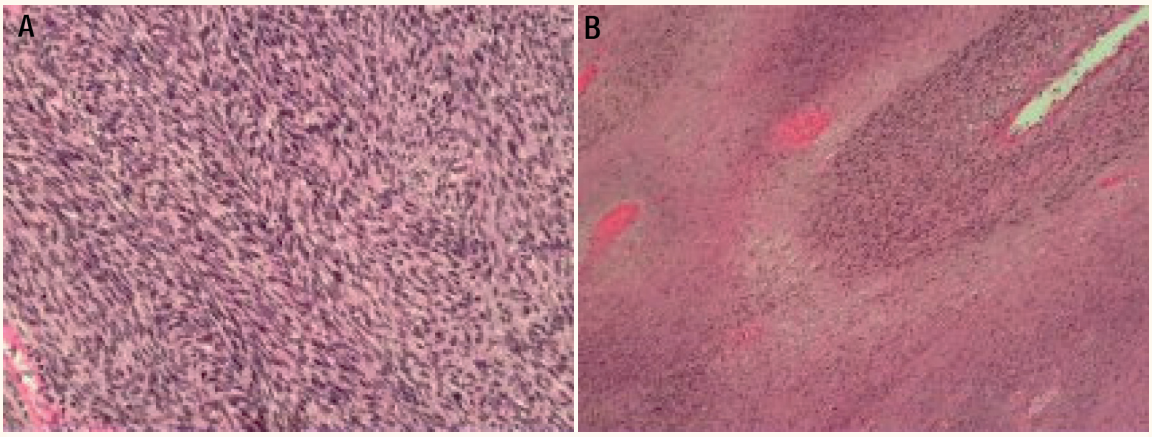

Figure 5: (A) Pathology slide of low-powered view of fibrosarcoma with herring-bone pattern; **(B)** Pathology slide showing areas of viable tumor fascicles with adjacent necrosis

References

1. McArthur G. Dermatofibrosarcoma protuberans: Recent clinical progress. Ann Surg Oncol 2007; 14:2876–86.
2. Yilmaz A, Cenesizoglu E, Egilmez E, Onel S, Mustu M, Cennet A. Dermatofibrosarcoma protuberans: A case report of a rare, bulky tumor that was managed with surgical therapy. Int J Shoulder Surg 2009; 3:16–20.
3. Basu S, Baghel NS. Recurrence of dermatofibrosarcoma protuberans in post-surgical scar detected by (18) F-FDG-PET imaging. Hell J Nucl Med 2009; 12:68.
4. Rubin BP, Schuetze SM, Eary JF, Norwood TH, Mirza S, Conrad EU, et al. Molecular targeting of platelet-derived growth factor B by imatinib mesylate in a patient with metastatic dermatofibrosarcoma protuberans. J Clin Oncol 2002; 20:3586–91.
5. Mizutani K, Tamada Y, Hara K, Tsuzuki T, Saeki H, Tamaki K. Imatinib mesylate inhibits the growth of metastatic lung lesions in a patient with dermatofibrosarcoma protuberance. Brit J Dermatol 2004; 151:232–57.
6. Ah-Weng A, Marsden JR, Sanders DSA, Waters R. Dermatofibrosarcoma protuberans treated by micrographic surgery. Brit J Cancer 2002; 87:1386–9.
7. Dragoumis DM, Katsohi LA, Amplianitis IK, Tsiftsoglou AP. Late local recurrence of dermatofibrosarcoma protuberans in the skin of female breast. World J Surg Oncol 2010; 8:48.
8. Cottier O, Fiche M, Meuwly J-Y, Delaloye J-F. Dermatofibrosarcoma presenting as a nodule in the breast of a 75-year-old woman: A case report. J Med Case Reports 2011; 5:503.
9. Rutkowski P, Wozniak A, Switaj T. Advances in molecular characterization and targeted therapy in dermatofibrosarcoma protuberans. Sarcoma 2011; 959132.
10. Labropoulos SV, Razis ED. Imatinib in the treatment of dermatofibrosarcoma protuberans, Biologics 2007; 1:347–53.