CASE REPORT

Pigmented villonodular synovitis: dedicated PET imaging findings

Ian Blake Amber,¹ Brian J Clark,¹ Gary Stuart Greene^{1,2}

¹Department of Radiology, The Pennsylvania Hospital, University of Penn Health System, Philadelphia, Pennsylvania, USA

²Department of Nuclear Medicine, The Pennsylvania Hospital, University of Penn Health System, Philadelphia, Pennsylvania, USA

Correspondence to
Dr Gary Greene,
greene@gpahosp.com

SUMMARY

Pigmented villonodular synovitis (PVNS) is an uncommon entity, which has the potential to cause severe pain. The gold standard for evaluation is MRI, and previous PET findings associated with PVNS have only been documented in the setting of concurrent malignancy. In the setting of recurrent disease, PET is being used to evaluate prebiological and postbiological treatment responses. Recurrent PVNS demonstrates greater hypermetabolic activity than previously documented, supporting the case as a potential mimic of malignant/metastatic disease. Post-treatment evaluations demonstrate decreased metabolic activity, which suggests response to treatment. This behaviour further supports the contention that there is a neoplastic origin to PVNS.

BACKGROUND

Pigmented villonodular synovitis (PVNS) is an uncommon benign hypertrophic process that leads to villous, nodular or villonodular proliferations of the joint synovium.¹ The exact aetiology of PVNS is unknown and has historically been attributed to one of the following: an inflammatory process; repeated haemorrhage into the joint from an occult synovial haemangioma or repetitive mild trauma; or a disorder of lipid metabolism. Though typically thought of as a benign lesion, progressive cytological anomalies and rare cases of sarcomatous transformation have been documented suggesting a neoplastic origin.²

Pathologically, PVNS can be classified into: intra-articular diffuse PVNS (23%), intra-articular focal PVNS (6%) and extra-articular variations (71%).¹ Extra-articular PVNS most commonly involves the volar hand or wrist (65–89%), followed by the foot and ankle (5–15%).¹ MRI is the preferred technique for identifying and characterising the extent of PVNS and its variants in order to guide surgical excision.

Positron emission tomography (PET) is not used in the traditional workup of PVNS. However, PVNS has been identified on PET imaging performed in the setting of a known malignancy, with a previously documented maximum standard uptake value (SUV) as high as 11.3.^{3–4} This is of particular importance in the workup of both sarcomas and melanomas, as PVNS can serve as a potential mimicker of musculoskeletal metastatic disease. As PET/CT is not traditionally used in the workup of PVNS, the PET imaging features of PVNS have only previously been described in the presence of a concurrent malignancy.

Currently, there is no standardised treatment for recurrent PVNS. Our patient is involved in phase 1 clinical trial of PLX3397, a selective inhibitor of FMS, Kit and oncogenic Flt3 activity. As part of the study protocol, the patient is evaluated by PET/CT as a marker of treatment response. This study allows a unique opportunity to evaluate PVNS with PET imaging in the absence of concurrent malignancy as well as offering a chance to correlate the PET images with established MRI findings.

CASE PRESENTATION

The patient is a 52-year-old African-American woman. She initially presented with a left foot mass in June 2010, which was biopsied and shown to be PVNS. The mass was surgically excised at an outside institution. In May 2011, she presented with similar symptoms. Further evaluation with MRI demonstrated recurrence of disease. The mass extended across the medial and lateral aspects of the foot, and encased segments of the second through fourth metatarsal shafts (figure 1 A–C). There was bony oedema, and it was unclear if this was due to bony involvement by tumour or stress reaction. The patient underwent surgical removal of the tumour in June 2011. The postoperative course was unremarkable. In January 2012, the patient presented with further recurrence of the soft tissue mass. Over multiple check-ups, the mass continued to enlarge and cause painful symptoms. The patient was advised to consider enrolling in a trial for a new drug to potentially treat PVNS. As per study protocol, she was to have a pretrial PET/CT. This was done in August 2012, and demonstrated marked uptake in the left foot (SUV maximum of 17.1 (figure 2)) in the region of prior surgery. These findings are consistent with recurrent PVNS. Follow-up PET/CT in October 2012 showed a decrease in uptake, with a maximum SUV of 6.1 (figure 3).

TREATMENT

Our patient is participating in a phase I clinical trial of PLX3397 for control of her recurrent PVNS. PLX3397 is an oral therapy, which functions as a selective inhibitor of FMS, Kit and oncogenic Flt3 activity.

OUTCOME AND FOLLOW-UP

Our patient demonstrated a decrease in metabolic activity of her PVNS, and an improvement in symptoms with the use of biological therapy. However, the clinical trial for PLX3397 is underway, and full results are not yet available.

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Figure 1 (A–D) MRI of the left foot.

DISCUSSION

PVNS is an incompletely studied disease entity, with no standardised treatment beyond initial surgical excision. Currently, MRI is considered the gold standard for evaluation of PVNS, and aside from clinical trials, PET/CT is not indicated. Therefore, the only previously published cases regarding the PET appearance of PVNS have occurred in the setting of concurrent malignancy. Though PVNS was documented to have hypermetabolic activity, the imaging characteristics on PET are not well defined as PVNS was considered an incidental finding on these scans.

With the advent of biological therapy there is likely to be an increase in the number of cases of PVNS imaged with PET. PVNS appears to behave similarly to a malignant/metastatic entity, demonstrating markedly hypermetabolic activity, with an appropriate decrease in activity in the setting of targeted treatments. These findings support previous case reports showing that PVNS can successfully mimic malignant/metastatic disease. Additionally, the apparent response to biological treatment suggests that there is a neoplastic component to the aetiology of PVNS.

Figure 2 Positron emission tomography/CT prior to intervention.

Figure 3 Positron emission tomography/CT after intervention.

Learning points

- ▶ Pigmented villonodular synovitis (PVNS) demonstrates hypermetabolic properties typically associated with malignant and/or metastatic disease on positron emission tomography (PET)/CT.
- ▶ PVNS responds to biological therapy with decreasing standard uptake value, similar to the way malignant and/or metastatic disease would.
- ▶ With the advent of biological agents, there will likely be an increase in the role of PET imaging to characterise the activity and treatment response of PVNS.
- ▶ There is likely a neoplastic component to the aetiology of PVNS.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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