Ann Nucl Med (2008) 22:715–717
DOI 10.1007/s12149-008-0175-7

CASE REPORT

Histiocytic sarcoma: PET–CT evaluation of a rare entity

**Emel Yaman · Banu Ozturk · Ozlem Erdem
Nahide Gokcora · Ugur Coskun · Omer Uluoglu
Mustafa Benekli**

Received: 30 October 2007 / Accepted: 24 January 2008
© The Japanese Society of Nuclear Medicine 2008

Abstract Histiocytic sarcoma is a rare malignancy of hematopoietic origin. Lymph nodes, skin, and extranodal sites, especially gastrointestinal tract, are commonly involved. Some cases reported in the past as non-Hodgkin's lymphoma are now classified as histiocytic sarcoma by detailed immunohistochemical studies. Patients with clinically localized disease have a good prognosis whereas those with lymphatic involvement have an aggressive course. In our case, histiocytic sarcoma was detected, originating from the skin over the left shoulder associated with disseminated lymphadenopathy. A positron emission tomography/computed tomography examination was done for evaluating the extent of the disease which showed pathologic increased 18F-fluorodeoxyglucose uptake in the lymph nodes, indicating widespread disease. The pertinent literature is reviewed.

Keywords Histiocytic sarcoma · PET–CT · Disseminated disease

E. Yaman · B. Ozturk · U. Coskun · M. Benekli (✉)
Department of Medical Oncology, Gazi University Faculty of Medicine, Besevler, Ankara 06500, Turkey
e-mail: mbenekli@gmail.com

O. Erdem · O. Uluoglu
Department of Pathology, Gazi University Faculty of Medicine, Ankara, Turkey

N. Gokcora
Department of Nuclear Medicine, Gazi University Faculty of Medicine, Ankara, Turkey

Introduction

Histiocytic sarcoma is a rare malignancy of hematopoietic origin, composed of cells morphologically and immunophenotypically similar to that of mature histiocytes [1]. It mainly involves men in their mid-40s. With the developments in immunohistochemical (IHC) techniques, histiocytic sarcoma cases are defined by the expression of CD68 and lysosome, and lack of CD1a and CD21/CD35. Most of the patients described before the widespread use of IHC probably had other types of non-Hodgkin's lymphomas [2–4]. Most cases present with extranodal involvement in advanced stages and have a poor prognosis [5–7].

Positron emission tomography (PET) scan is a new metabolic imaging tool, employed in the staging of lymphomas [8]. PET scan is commonly used to evaluate the response to therapy. It may also be used for guiding treatment and deciding on possible sites of biopsy for tissue diagnosis. The use and characteristics of PET scan in histiocytic sarcoma have not been described. We report a patient presenting with a cutaneous histiocytic sarcoma that later disseminated to involve regional lymph nodes. PET scan was used to detect lymphatic dissemination.

Case report

A 64-year-old Caucasian man presented with a nodular lesion 1 cm in diameter on his left shoulder. He had a history of 6 kg weight loss and fever. On physical examination, there were no other abnormalities. Excisional biopsy of the lesion was performed and was initially reported as undifferentiated squamous cell carcinoma.

However, the tumor had positive surgical margins which necessitated a re-excision following referral to our hospital. Both pathological specimens were reviewed again, and histiocytic sarcoma was diagnosed. On pathological examination, tumor cells were stained strongly and diffusely with vimentin, alpha 1 antitrypsin and CD68 and heterogeneously with kappa and lambda stains.

One month later, axillary lymphadenopathy was palpated on physical examination. Ultrasonography revealed multiple lymphadenopathies. To evaluate the extent of the disease, a PET–computed tomography (CT) scan was performed on a GE Discovery ST PET–CT scanner. After an overnight fast, 12 mCi fluorodeoxyglucose (FDG) was injected and with a waiting time of 2 h and 30 min, PET scanning was done which showed pathologically increased 18F-FDG uptake in the bilateral superior jugular, left inferior jugular, and deep and superficial axillary lymph nodes (Fig. 1). The SUV maximum value of the most prominent lesions was 12.7.

Left axillary lymph node dissection was performed. Histiocytic sarcoma infiltration was detected in 4 of 11 dissected lymph nodes with extension to the perinodal soft tissue. A chemotherapy regimen consisting of cyclophosphamide, adriamycin, vincristine, and prednisolone (CHOP) failed to induce a clinical response. Radiotherapy to the involved lymph nodes was done. Although a clinical response to radiotherapy was obtained, the patient died of lobar pneumonia shortly after completion of radiotherapy.

Discussion

Histiocytic sarcoma is a rare hematopoietic malignancy of histiocytic origin. The largest series reported to date comprises only 18 patients [5]. Patients usually present in advanced stages with extranodal involvement. The natural course of the disease is usually aggressive, and the prognosis is poor. Incorrect diagnoses, mostly lymphomas, were frequent prior to the era of IHC. In our case, the diagnosis of histiocytic sarcoma was confirmed by positive CD68, vimentin, and alpha 1 antitrypsin on IHC.

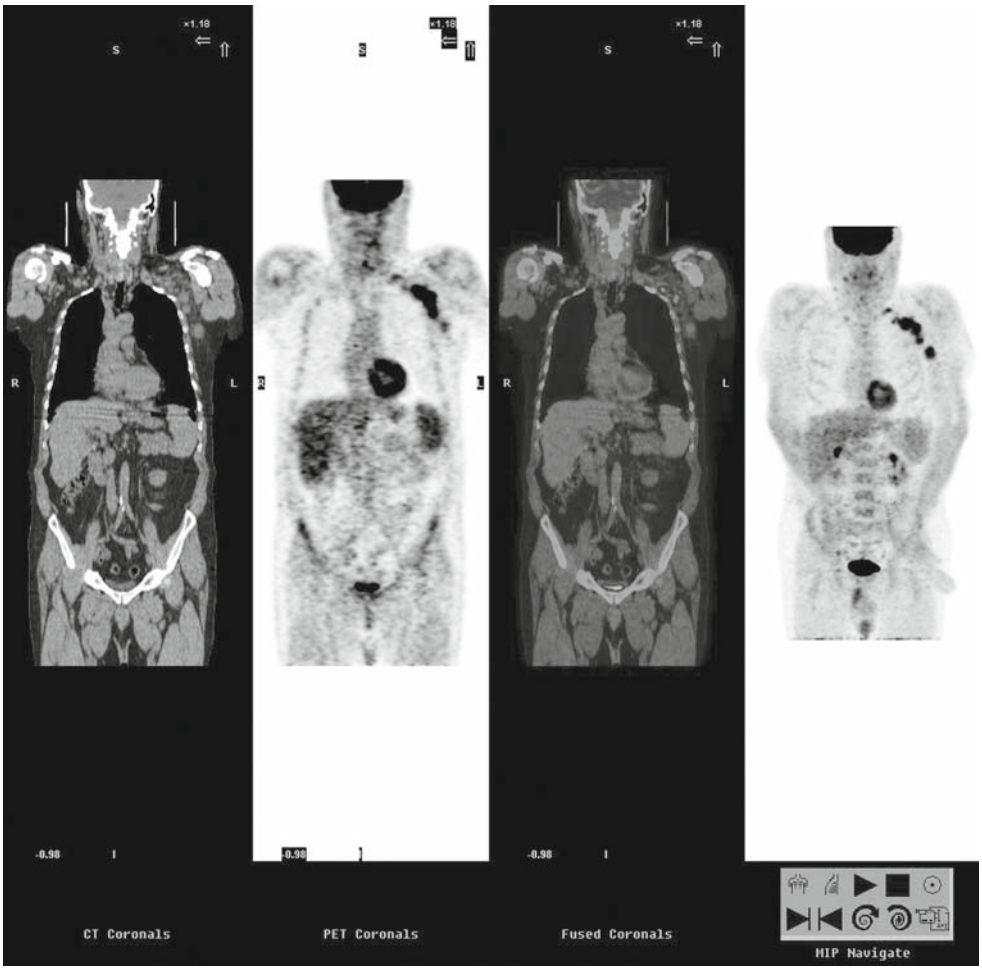

Fig. 1 Positron emission tomography/computed tomography fusion images showing disseminated disease

Positron emission tomography is a metabolic imaging modality widely used in the evaluation of patients with non-Hodgkin's lymphoma. Hybrid PET imaging together with CT provides a precise anatomic localization combined with functional characterization of tissues. FDG-PET might not accurately reflect the malignant potential of all tumors, but rather might implicate cellular components included in the lesions. A high accumulation of FDG can be observed in histiocytic, fibroblastic, and some neurogenic lesions, regardless of whether they are benign or malignant. There are case reports indicating the possible role of PET scan in histiocytic lineage disorders, such as Langerhans cell histiocytosis (LCH) [9–11]. PET–CT was reported to show dissemination of disease more accurately than CT images in LCH. In addition to giving additional information about the extent of disease, it was also helpful in the follow-up, evaluation of response, and detection of early recurrences. PET scan can be also useful as a guide for local treatment or for the identification of possible biopsy sites. However, a literature search revealed only one report of PET scan used in histiocytic sarcoma [7]. PET scan confirmed localized disease to pulmonary nodules which directed targeted delivery of radiation to these lesions in a 3-year-old boy. Similarly, our patient had pathologically increased 18F-FDG uptake in several lymphadenopathies including jugular and axillary lymph nodes, not detected with other imaging modalities. The treatment of the patient was tailored accordingly leading to axillary lymph node dissection. Our case further illustrates the utility of PET scan in diagnosis, staging, and directing treatment in histiocytic sarcoma.

In conclusion, histiocytic sarcomas are very rare hematopoietic tumors of histiocytic origin. Because of the rarity, there is no established role of PET scan in the management of histiocytic sarcoma. Our case illustrates that PET scan is very valuable in the evaluation of disease dissemination and tailoring treatment. Routine incorporation of PET–CT into the management of histiocytic sarcoma is warranted.

References

1. Jaffe ES, Harris NL, Stein H, Vardiman JW. World Health Organization classification of tumors: pathology and genetics of tumours of hematopoietic and lymphoid tissues. Lyon: IARC; 2001.
2. Mirchandani I, Shah I, Palutke M, Varadachari C, Tabaczka P, Franklin R, et al. True histiocytic lymphoma: a report of four cases. Cancer 1983;52:1911–8.
3. Van der Valk P, Van Oostveen JW, Stel HV, Van der Kwast TH, Melief CJ, Meijer CJ. Phenotypic and genotypic analysis of large-cell lymphomas, formerly classified as true histiocytic lymphoma: identification of an unusual group of tumors. Leuk Res 1990;14:337–46.
4. Van Heerde P, Feltkamp CA, Hart AA, Somers R, Van Unnik JA, Vroom TM. Malignant histiocytosis and related tumors: a clinicopathologic study of 42 cases using cytological, histochemical and ultrastructural parameters. Hematol Oncol 1984;2:13–32.
5. Pileri SA, Grogan TM, Harris NL, Banks P, Campo E, Chan JK, et al. Tumours of histiocytes and accessory dendritic cells: an immunohistochemical approach to classification from the International Lymphoma Study Group based on 61 cases. Histopathology 2002;41:1–29.
6. Hornick JL, Jaffe ES, Fletcher CDM. Extranodal histiocytic sarcoma: clinicopathologic analysis of 14 cases of a rare epithelioid malignancy. Am J Surg Pathol 2004;28:1133–44.
7. Buonocore S, Valente AL, Nightingale D, Bogart J, Souid A. Histiocytic sarcoma in a 3-year-old male: a case report. Pediatrics 2005;116:e322–5.
8. Juweid ME, Stroobants S, Hoekstra OS, Mottaghy FM, Dietlein M, Guermazi A, et al. Use of positron emission tomography for response assessment of lymphoma: consensus of the Imaging Subcommittee of International Harmonization Project in Lymphoma. J Clin Oncol 2007;25:571–8.
9. Blum R, Seymour JF, Hickks RJ. Role of FDG-positron emission tomography scanning in the management of histiocytosis. Leuk Lymphoma 2002;43:2155–7.
10. Kaste SC, Rodriguez-Galindo C, McCraville ME, Shulkin BL. PET–CT in pediatric Langerhans cell histiocytosis. Pediatr Radiol 2007;37:615–22.
11. Weiss SE, O'Connor L, Welsh JS. Refinement of radiation therapy based on PET data in adults with Langerhans cell histiocytosis of soft tissues. Clin Adv Hematol Oncol 2006; 4:290–4.