The Journal of Laryngology & Otology
March 2005, Vol. **119,** pp. 219–221

Inflammatory pseudotumour of the neck with multifocal sites on positron emission tomography scan imaging

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Abstract
We present a patient with an inflammatory pseudotumour of the neck with multifocal sites in the head and chest responding to steroids. A review of the literature revealed that this is the first case of a pseudotumour with multiple sites in the head and neck as revealed by 2-[18]fluorodeoxyglucose (FDG) PET scan imaging.

Key words: Neoplasms, Muscle Tissue; Neck; Chest; Tomography, Emission, Computed; Neoplasms, Multiple Primary

Introduction

Inflammatory pseudotumours are benign idiopathic tumours commonly mistaken as neoplastic lesions, which most commonly occur in the lung, abdomen and head and neck. There may be more than one site involved but multiple sites are rare and some authors[1] believe that they do not occur, due to the histological benign characteristics of pseudotumours.

Histological appearances of pseudotumours are varied and comprise a range of different cells, including myofibroblasts and fibroblasts together with polymorphic inflammatory cells and plasma cells and lymphocytes. They can be distinguished from lymphomas, sarcomas, rhabdomyosarcomas and histiocytomas based on their histological and immunohistochemical staining.

2–[18] fluorodeoxyglucose (FDG) positron emission tomography is a whole body imaging technique that uses radioactive positrons to detect areas of increased metabolism. FDG is taken up by metabolically active tumour cells, which can be detected even before being seen on magnetic resonance imaging (MRI) or computed tomography (CT). The amount of uptake is expressed as a standardized uptake value (SUV) with a threshold of 2.5–3 being consistent with malignancy.

Case report

A 31-year-old man presented with a five-month history of an intermittently painful swelling on the right side of his neck. This was associated with weight loss, sweats and blurring of the vision in his left eye. He was a smoker and drank 20 units of alcohol per week. Past medical history was unremarkable. Clinical examination demonstrated a firm, right-sided neck mass with no other significant findings. Ultrasound of this area revealed a right suprahyoid mass measuring 4 x 3 x 2 cm, infiltrating the sternocleidomastoid muscle. Fine needle aspiration (FNA) was performed, but the cytological examination showed inflammatory changes only.

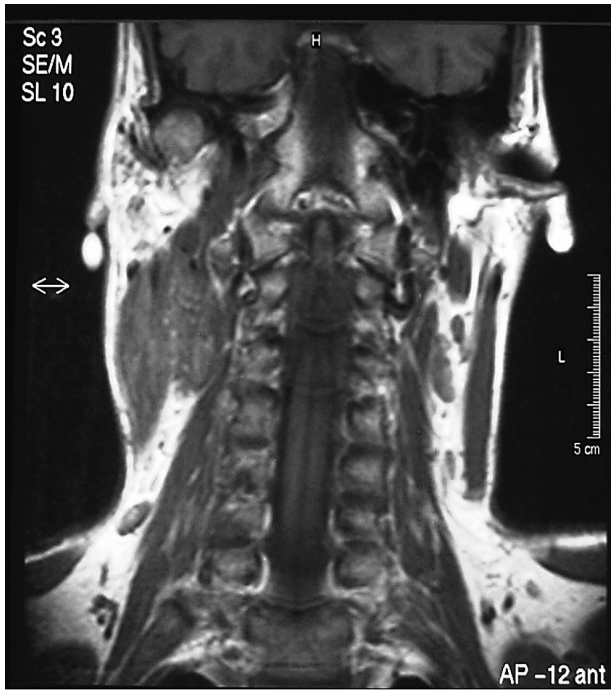

Fig 1.
MRI scan of right neck pseudotumour.

An initial MRI (Figure 1) demonstrated an invading mass in the right side of the neck level II region which directly involved the ipsilateral sternocleidomastoid as well as slightly enlarged left sided level II nodes. An FDG PET scan was then performed within a week of the MRI scan. 330 Mbq 18-FDG were injected intravenously and blood glucose was 4.7 mmol/l. The patient was scanned on a Siemens/CTI EXACT 47 according to whole body protocol. The FDG PET scan showed intense abnormal

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Accepted for publication: 20 November 2004.

https://doi.org/10.1258/0022215053561620 Published online by Cambridge University Press

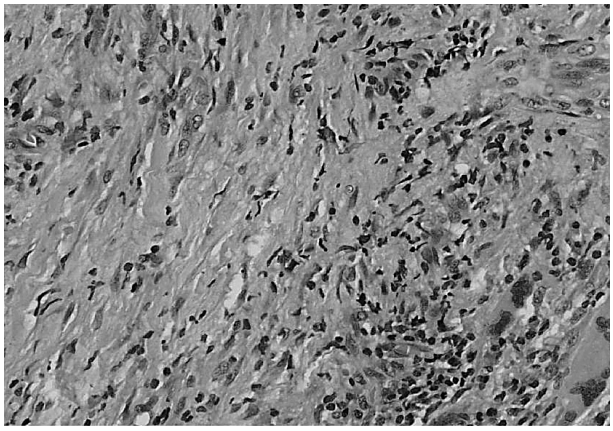

FIG. 2.
Inflammatory pseudotumour from the neck biopsies (H & E; × 400). Proliferating myofibroblasts set in a collagenized stroma containing chronic inflammatory cells.

uptake in the right neck with an SUV(max) of 8.6. However, the scan had two unexpected findings, that of intense abnormal uptake in the left infratemporal fossa (SUV(max) 4.3) as well as an area of abnormal uptake on the left chest wall (SUV(max) 2.2). The SUV(max) obtained from the right neck and left infratemporal fossa masses were consistent with malignancy. A follow-up MRI scan six weeks after the first MRI scan confirmed a new soft tissue mass in the left infratemporal fossa with involvement of the pterygoids and the left masseter as well as some small nodes in this region.

The patient underwent a panendoscopy with multiple biopsies, right tonsillectomy and open neck exploration with biopsies. Histology specimens from the neck biopsies consisted of a florid reactive proliferation of spindle-shaped fibroblasts infiltrating the strap muscles of the neck, with an associated dense inflammatory infiltrate of lymphocytes and plasma cells (Figure 2). Immunohistochemistry demonstrated that the proliferative stromal cells were positive for calponin but negative for desmin, smooth muscle actin, muscle specific actin, myogenin and myo D1, confirming their myofibroblastic origin, thereby excluding the differential of sarcomas and rhabdomyosarcomas which are positive for these markers.[2]

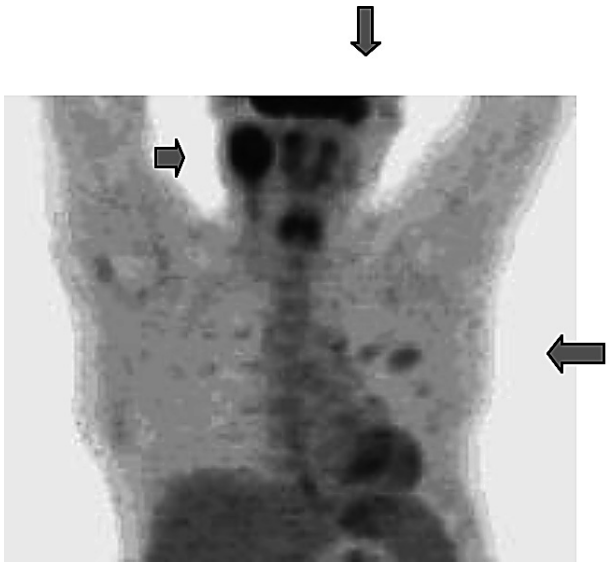

FIG. 3
FDG PET before treatment. Right neck lesion ➧. Left chest lesion ➧. Left infratemporal lesion➧.

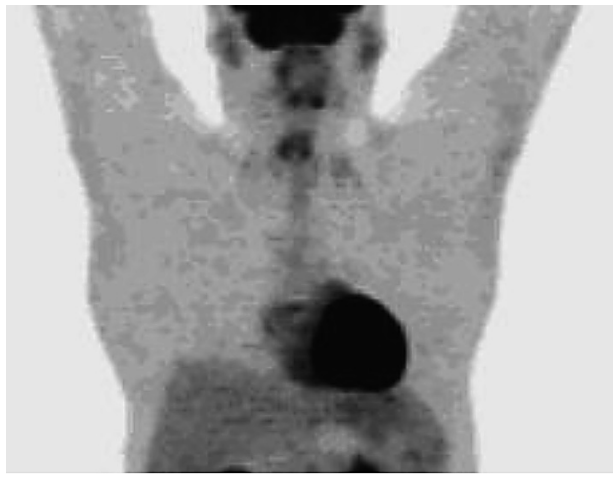

FIG. 4
FDG PET after treatment.

The lymphoid infiltrate appeared reactive with CD20 and CD3 positive B and T-cells. There was also early germinal centre formation containing cells that were CD23 positive. There was no evidence of malignant lymphoma on immunohistochemical analysis as demonstrated by the lack of a monoclonal population of lymphoid cells, which are present in lymphomas.[2] Histology therefore confirmed an infiltrating fibro-inflammatory lesion (pseudotumour).

A second opinion from the Royal Marsden Hospital confirmed a pseudotumour with inflammatory fibroblastic characteristics. The absence of light chain restriction, lack of centrocytic morphology and the identification of other inflammatory cells as well as fibroblastic / myofibroblastic cells within the lesion served to make the diagnosis of inflammatory pseudotumour.

Clinical symptoms evolved over the course of this time and the patient developed severe jaw pain with trismus, and worsening left visual acuity corresponding to radiological findings. Following ophthalmology review, optic disc swelling and engorged retinal veins were seen suggesting possible invasion of the optic nerve. Prednisolone 30 mg daily was commenced. Rapid improvement of symptoms was seen with the steroids within days and the neck swelling and pain resolved and vision returned to normal within weeks. A two year follow-up has continued to show disease resolution and is currently continuing (Figures 3 and 4).

Following treatment with steroids, FDG PET was used to monitor disease resolution showing remarkable response of the pseudotumour to steroids. The left chest wall lesion and left infratemporal fossa lesion on PET scan had disappeared and only low grade uptake was seen in the right neck where previously an intense signal had been seen (Figure 5).

To the authors' knowledge, this case is unique from the imaging point of view for two reasons: there is no documented case of pseudotumour being followed up by PET scans to monitor disease resolution, nor has a head and neck pseudotumour ever been known to have multifocal sites. It does of course raise the interesting possibility that this behaviour may be characteristic of these lesions but that it is the use of PET imaging which has made the authors aware of other focal sites which may otherwise have simply resolved with treatment and never been documented.

https://doi.org/10.1258/0022215053561620 Published online by Cambridge University Press

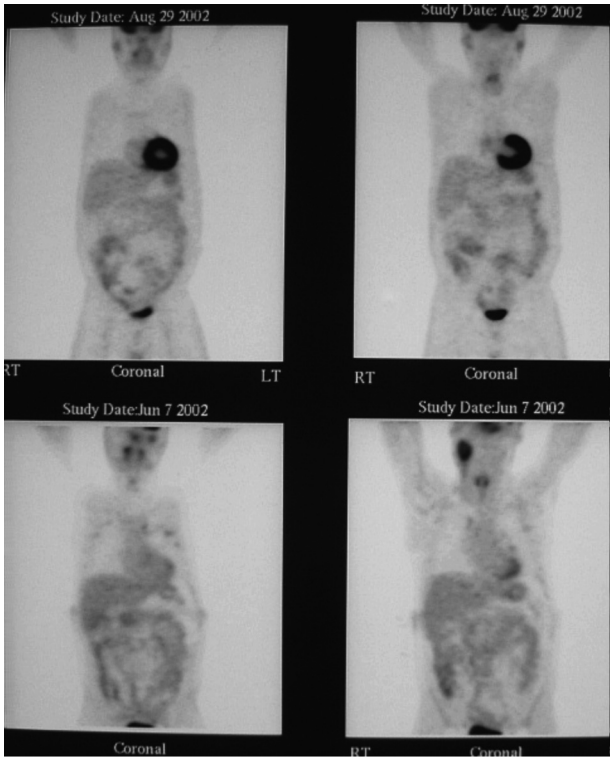

FIG. 5
FDG PET before and after treatment.

Discussion

The diagnosis of an inflammatory pseudotumour is one of exclusion of neoplasia. Open biopsy is necessary for adequate tissue sampling and diagnosis.[1] Treatment remains controversial, steroids, radiotherapy and surgical excision being the main options. Lymphoma can be excluded in this case by immunohistochemical analysis, absence of classical Reed-Sternberg cells or their variants excludes Hodgkin's lymphoma and the polyclonality of the infiltrate excludes non-Hodgkin's lymphoma. Lymphomas may also respond to steroids and are detected by PET scan but their treatment modality is mainly chemotherapy.

This rare and fascinating case has demonstrated a neck pseudotumour with hitherto unreported presence of multifocal sites, detected by FDG PET imaging in the chest wall, jaw and orbit. Batsakis *et al*.[3] have reported metastasis with abdominal pseudotumours and orbital pseudotumours have shown direct extension into the cervico-facial region[4] but no other cases of metastases have been documented. Clinically and radiologically pseudotumours can behave aggressively with cranial nerve palsy and bony destruction but histologically they are benign tumours.

While it is well known that malignant tumours take up FDG PET avidly, this is the first case of pseudotumour with multifocal sites that has been initially assessed, and then followed up by FDG PET scanning. Traditionally, CT and MRI are the imaging modalities used, but there is no doubt that FDG PET scanning has added a new dimension to management of this patient by detecting unsuspected secondary sites.

Metastases are not generally associated with pseudotumours so whole body MRI or spiral CT would not be considered unless suspected by clinical symptoms. In this case the rib lesion would have been undetected as there were no clinical symptoms in this region.

Conclusion

In this case, FDG PET imaging has proved to be an invaluable adjunct to MRI by locating unsuspected multifocal sites, which were previously thought not to occur. Our case demonstrated improved management of the patient by using FDG PET to assess and monitor disease resolution.

- **This is a report of an inflammatory pseudotumour of the neck presenting in multiple sites**
- **The findings on PET scanning are presented**

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Miss Babar-Craig takes responsibility for the integrity of the content of the paper.
Competing interests: None declared