CASE REPORT

Recurrent inflammatory myofibroblastic tumour of the lung: FDG PET/CT scan findings

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SUMMARY

Inflammatory myofibroblastic tumour (IMT) is a rare neoplasm, occurring most often in children and young adults. IMTs have intermediate biological behaviour with the chance of local invasion, recurrence and even distant metastasis. Wide range of clinical presentations makes the precise diagnosis of IMT more challenging. The best method for definitive diagnosis is tissue biopsy and newer imaging modalities including fleurodeoxyglucose (FDG) positron emission tomography (PET)/CT are useful tools in detection of disease recurrence or distant metastasis. Complete surgical resection is the best-known treatment for this tumour. Here we are presenting an IMT case in a 12-year-old girl in which her recurrent pulmonary IMT was diagnosed based on FDG PET/CT findings and referred for further salvage treatment. Overall imaging modalities are not specific, but PET/CT scan can be useful tool for evaluation of IMT regarding initial staging and restaging to assess treatment response and recurrence.

Complete surgical resection is the best-known treatment for this tumour.¹⁵ Imaging modalities can be considered as diagnostic tool for detecting IMTs. Considering the hypermetabolic nature of this tumour, fleurodeoxyglucose (FDG) positron emission tomography (PET)/CT can be used for detection of recurrence and distant metastasis.^{16 17} FDG PET/CT findings of IMTs have been discussed in limited number of cases based on our literature review.¹⁶ In this paper, we will present an interesting case of recurrent pulmonary IMT in a 12-year-old girl and discuss her FDG PET/CT findings followed by a quick glance at the literature.

CASE PRESENTATION

A 12-year-old girl was referred to our department (PET/CT) for evaluation of response to treatment.

She was first diagnosed with IMT in September 2015. The patient was diagnosed after 3 weeks of fatigue, nervousness, malaise, decreased appetite and weight loss (30 kg to 24 kg) which was followed by 1 week of uncontrolled chills and fever.

BACKGROUND

Inflammatory myofibroblastic tumour (IMT) is a rare neoplasm which is believed to include less than 1% of soft-tissue sarcomas.¹ The tumour that is also called inflammatory pseudotumour (IPT) or plasma cell granuloma occurs most often in children and young adults and predominantly has a benign nature but in some occasions, it can have more aggressive and malignant behaviour.²

Aetiology of this tumour is not well understood; however, trauma, surgery, some viral infections and autoimmune disorders can be considered as an underlying aetiology.^{3 4}

Lung is the most common site for IMT. However, the variety of extrapulmonary sites including skull base, head and neck, liver, peritoneum, retroperitoneum, gastrointestinal tract, pancreas, genitourinary tract, extremities, breast and even heart can be affected.^{5–11} Depending on location, this tumour may present with different symptoms such as fever, flu-like symptoms, headache, nerve palsy, facial pain, exertional dyspnoea and abdominal pain^{7 12–14} and this wide spectrum of clinical presentations makes the precise diagnosis even more challenging for this entity.⁵

The best method for definitive diagnosis is tissue biopsy. Typical pathological finding in the biopsy specimen is presence of mesenchymal spindle cells in the context of inflammatory cells.¹⁵

INVESTIGATIONS

Initial investigations

She was admitted to children hospital for further workup. Initial laboratory findings demonstrated anaemia, leucocytosis and evidence of a 92×82×61 mm pulmonary mass in the posteromedial segment of the right lower lobe.

The mass was biopsied under CT scan guidance and the pathology report stated pulmonary tissue involved by uniform neoplastic cells with fascicular pattern and scattered mononuclear leucocytes and peripheral lymphoplasmacytosis (figure 1). Immunohistochemical studies of the biopsy specimen showed a positive reaction for vimentin and smooth muscle and negative reaction for S-100, CD34 and ALK-1 and about 5% of tumour cells demonstrated positive reaction for Ki-67. Finally, the diagnosis of the IMT was established.

DIFFERENTIAL DIAGNOSIS

The main differential diagnosis for an IMT presenting in the lung is IgG4-related IPT which is an inflammatory reactive lesion in which the expression of ALK-1 in spindle cells helps to distinguish IMTs from IgG4-related IPTs.¹⁸ Additional differential diagnosis includes pulmonary desmoid tumours and malignant lung masses such as primary bronchogenic carcinoma.

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Figure 1 Myofibroblastic and fibroblastic spindle cells with inflammatory infiltrate of lymphocytes, plasma cells, eosinophils and histiocytes.

TREATMENT

After initial workup, she was referred for further evaluation and resection of the tumour. The tumour was successfully resected followed by referral to an oncologist for systemic treatment. Chemotherapy was initiated with chemotherapy courses at 25-day intervals.

OUTCOME AND FOLLOW-UP

Recurrence

Six months after completion of chemotherapy and during routine surveillance, recurrent mass in right pulmonary lower lobe was detected on CT-scan. Subsequently, she underwent the second surgery in March 2016 followed by another course of chemotherapy. Almost 4 months later, another mass in left lower lobe was found on CT-scan and the third subsequent surgery was performed in July 2016.

Secondary investigation

During routine follow-up, after detection of a single 9.5 mm nodule in superior segment of left lower lobe and three additional small nodules (2.5–3 mm) in left upper lobe and also a suspicious bone lesion in T11 found on the whole body bone scan, the patient was referred to PET/CT department for recurrence evaluation.

Sixty minutes after injection of 4.5 MBq/kg of ^{18}F -FDG, the PET/CT imaging was acquired using a Discovery 690 VCT (General Electric (GE) Healthcare, Milwaukee, USA) that is equipped with 64-slice CT scan (Light Speed VCT) from head to mid-thighs. PET/CT scan revealed a hypermetabolic left lung nodule measuring 15×17 mm (SUVmax=5.8) (figure 2), a hypermetabolic right pleural focus adjacent to right 11th rib (SUVmax=4.1) and right pleural effusion and thickening with mild metabolic activity (figure 3), sclerotic lesion in 11th vertebra with sclerotic border and no abnormal metabolic activity and also diffuse increased metabolic activity of spleen compared with the background liver parenchyma.

With respect to these findings, the patient was referred for the fourth surgery and subsequent chemotherapy (salvage treatment).

Discussion

Since its first introduction in 1939 by Brunn in the lung, our knowledge about IMT has changed dramatically.¹⁹

Numerous reports stated that it can be found in different locations from head and neck to thorax and abdomen and even extremities. Therefore, the variety of signs and symptoms can be presented by this tumour. Pang *et al*¹¹ reported a case of the right ventricular IMT in an 11-month-old infant presented with febrile illness. Oh *et al*²⁰ described an IMT in the appendix of an 85-year-old man with the history of gastric cancer. Another interesting site for this tumour reported by Lyon *et al*²¹ in carina of a 7-year-old girl who presented with cough, wheezing and pneumonia with evidence of near-total obstruction of the trachea.

Figure 2 Hypermetabolic left lung nodule measuring 15×17 mm (SUVmax=5.8).

Figure 3 Hypermetabolic right pleural focus adjacent to right 11th rib (SUVmax=4.1) and right pleural effusion and thickening with mild metabolic activity.

At the initial diagnosis, majority of these tumours are large, ranging from 3 to 10cm (average 7cm)).²² This rare tumour which is frequently found in children and young adults has the female gender preference.

It is predominantly benign, but has the potential to infiltrate to adjacent organs which mimic malignant tumours^{23 24} and sometimes it can have malignant behaviour or transform into a malignant tumour.²⁵ It frequently recurs but metastasis is a rare finding, so it is believed that it has an intermediate biological behaviour which is also confirmed in the histological classification of soft tissue tumours by WHO.^{12 26}

Wang *et al*²⁷ reported a case of recurrent retroperitoneal IMT in a 74-year-old woman, who was presented with 10cm mass found incidentally during cholecystectomy. The tumour recurred 30 months after first surgery and was successfully retreated. Two cases of recurrent IMTs were found by Li *et al*²⁸ in the right upper extremity of a Chinese woman and the right thigh of a Chinese man. Both tumours recurred less than a year after initial surgery. Corsi *et al*²⁹ also reported a case of recurrent IMT in anterior wall of the glottis. Pascual-Gallego *et al*³⁰ presented a rare case of meningeal IMT which was found in a woman with a history of retroocular pain and recurrent headache.

Gorolay *et al*³¹ found a 9.7×6×8.6cm tumour in mediastinum of a 49-year-old man with oesophageal and bronchial invasion who was evaluated for progressive dyspnoea and dysphagia. Serial imaging with CT scan revealed a large subcarinal mass with dilation of upper oesophagus and main stem bronchus and lobar obstruction. PET/CT was performed for staging and revealed an area of intense metabolic activity in mediastinum with no evidence of distant metastasis. Inoue *et al*³² described a case of the IMT with multiple organ involvements in a 16-year-old girl with the history of tonic-clonic seizure. The patient was worked up and at least three tumours were found in the right cerebral hemisphere. With a high level of suspicious for malignancy, chest and abdominal CT scans were performed revealing multiple lung nodules, a breast mass and also at least four pancreatic masses.

The histopathological feature of this tumour includes myofibroblastic mesenchymal spindle cells accompanied by inflammatory cells including plasma cells, lymphocytes and eosinophils in a collagenous or myxoid stroma.³

Overall on CT scan, IMTs usually appear as solitary, well-circumscribed peripheral lung masses, with predominance for the lower lobes. Calcification of the masses is unusual (about 15%). It is not frequent, but they can also be multiple. On CT with intravenous contrast, they present a variable heterogeneous or homogeneous degree of enhancement pattern. It has been described an aggressive behaviour with invasion of the adjacent structures. On MRI, they present similar radiological findings as in other locations. Because of their similar radiological appearance to malignant lung masses, biopsy is necessary.³³

PET/CT is a well-known non-invasive hybrid imaging modality (both anatomical and functional) that has gained lots of attention during past decades. 18F-FDG is the main PET radio-tracer which is used in oncology. Many malignant and benign neoplasms and also inflammatory processes demonstrate uptake of 18F-FDG relative to the rate of tissue metabolism.¹⁷ Providing physiological information makes the PET/CT scanning a unique imaging tool in the evaluation of oncology patients in terms of response assessment, recurrence evaluation, tumour staging and radiotherapy planning.^{17 34} There are several reports in the literature that describes 18F-FDG avid IMTs. Ilic *et al*¹⁷ described a pulmonary IMT in a 14-year-old girl with the history of Hodgkin's lymphoma which showed very intense uptake on 18F-FDG PET/CT. Kuo *et al*³⁵ reported a case of the metastatic IMT with the origin in left hemithorax. The 18F-FDG PET/CT revealed areas of higher activity in jejunum and body of the pancreas. Yoon *et al*³⁶ described a large IMT in the right paratracheal space of a 25-year-old man with was incidentally found in X-ray which showed heterogeneous 18F-FDG uptake. The PET/CT scan findings in our case were consistent with the recurrent/residual disease after multiple treatments and more importantly, PET/CT was able to show metabolic activity in the right pleural effusion and hypermetabolic focus in the right pleura which were almost impossible to be detected by means of other imaging modalities.

Conclusion

IMTs are rare tumours with different origins and different signs and symptoms. Imaging modalities are not specific, but PET/CT scan can be a useful imaging tool for evaluation of IMT regarding

initial staging and restaging to assess treatment response and recurrence.

Learning points

- Inflammatory myofibroblastic tumours (IMTs) can occur almost anywhere in the body and mimic other diseases. Hence, clinicians should consider this condition in differential diagnosis.
- Although imaging findings of IMTs are not specific, 18F-fluorodeoxyglucose positron emission tomography/CT can be considered as a diagnostic guide.
- IMTs have intermediate biological behaviour with the chance of local invasion, recurrence and even distant metastasis.
- Complete surgical resection of the tumour is the best-known treatment.

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