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Short Communication

FDG PET Scanning in Hemophagocytic Lymphohistiocytosis

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Introduction

Hemophagocytic Lymphohistiocytosis (HLH) is a rare disorder. It progresses rapidly and can be life-threatening. Prompt initiation of treatment is critical for survival. Untreated, patients survive only a few months. These patients have multisystem involvement and may develop multi-organ failure. The goal of therapy for patients with HLH is to suppress life-threatening inflammation. After induction, patients who are recovering are weaned off therapy, while those who are not improving are continued on therapy as a potential bridge to stem cell transplantation. Monitoring treatment response is critical as treatment escalation may be warranted if the patient is not improving. The distinction between chemotherapy toxicity and worsening disease may be difficult to make clinically. Quite a variety of clinical, biochemical and immunological biomarkers are used at diagnosis and to assess treatment response. The response to initial therapy is a major factor in determining the need for additional therapy including Hematopoietic Cell Transplant (HCT). Response to induction therapy is monitored by assessing the patient clinically and using HLH disease-specific markers. Although this approach has some success, it is sometimes confounded by a new infection or treatment toxicity. Imaging can provide an additional layer of accuracy in the initial evaluation of HLH patients as well as their monitoring and follow-up. 18F-FDG PET scans are highly sensitive and can be quite helpful in the management of these patients (Figure 1).

Discussion

FDG PET-CT is an established management tool in a variety of cancers, however in noncancerous entities clinicians are more hesitant to use. That said FDG PET has been described as having a high impact in the care of HLH patients. Several authors have described the value of FDG PET scans in delineating the extent of disease in HLH as well as determining the severity/aggressiveness of disease involvement [1-6]. Wang et al, described the added value of FDG PET scans in delineating lymphoma lesions and managing lymphoma associated HLH [7]. On the other hand Yang et al, raised the concern of using FDG PET as a tool to assess bone marrow involvement by lymphoma in lymphoma associated HLH as HLH related bone marrow changes may frequently be a confounding factor [8]. Diffuse bone marrow uptake in HLH has also been described by several other authors [1, 8-11].

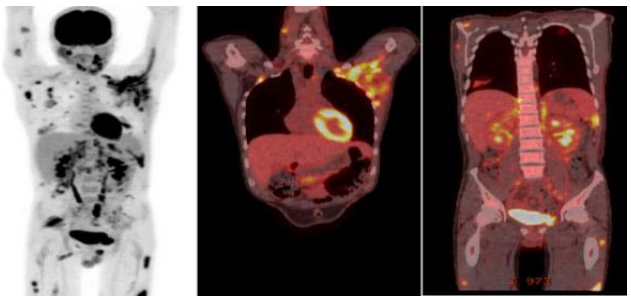

Figure 1: 20 yo gentleman with HLH. FDG PET scan showing multiple FDG avid lesions noted involving muscles and subcutaneous tissues in the face, arms, chest, abdomen and lower extremities. FDG avid lung nodules and mild diffuse bone marrow uptake is also noted. Ferritin levels elevated at 13,500. Patient passed away within a few weeks of initial diagnosis.

Additionally, HLH associated cancers can be detected by FDG PET scans and guide biopsies [4]. In 45 HLH patients Jigang described the distribution and pattern of uptake in secondary HLH cases including rheumatological systemic diseases, infections, and lymphoma [5,12]. Considering HLH is even less common in children and early diagnosis is critical in management and improving morbidity and mortality one should consider using FDG PET in cases of fever of unknown origin or other nonspecific presentations including in pediatrics. Adequate, early assessment of these patients is critical [2,13,14]. FDG PET can detect disease early and is a better predictor of prognosis and outcomes than clinical or laboratory biomarkers [1,6]. Therefore, one would also postulate that it would be a beneficial tool in treatment monitoring.

Conclusion

HLH is a serious life-threatening disease. FDG PET can play a major role in early disease assessment, prognosis, biopsy guidance, the evaluation of secondary causes and treatment monitoring. Larger multicenter studies are required for better guidance in this rare and fatal disease.

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