Hemolymphangioma involving bones and bladder detected on $^{68}$Ga-NEB PET/CT

A rare case report

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Abstract

Rationale: Hemolymphangioma is a rare developmental defect of combined vasal and lymphatic vasculature. It is very rare that hemolymphangioma affects the bones and bladder simultaneously, and this condition has never been reported in PubMed.

Patient concerns: A 12-year-old male has a history of hospitalization for recurrent episodes of chyluria of 8 years duration and progressively worsening gross hematuria with right hip pain for 6 months.

Diagnosis: Chylous test of urine fluid was positive. There was no organic disease in the kidneys. $^{68}$Ga-NOTA-Evans Blue (NEB) positron emission tomography/computed tomography (PET/CT) images demonstrated clearly several round and strip-shaped low-density shadows with mildly increased radioactive uptake in both bladder wall and pelvis, including sacral, pubic, and ischial bones. Histopathological analysis of biopsy on pubic and ischial bones confirmed the diagnosis of hemolymphangioma.

Interventions: The patient received treatment with traditional Chinese medicine.

Outcomes: At the 6-month follow-up visit, the patient’s symptoms of chyluria, hematuria, and pain were all mitigated.

Lessons: Hemolymphangioma is a rare benign disease. $^{68}$Ga-NEB PET/CT is a specific method for the lymphatic system, and it might provide more accurate and comprehensive information about the disorder of the lymphatic system compared with CT and magnetic resonance imaging. When patients suffer from suspected lesions of the lymphatic system, $^{68}$Ga-NEB PET/CT might be recommended.

Abbreviations: MIP = maximum intensity projection, MRI = magnetic resonance imaging, NEB = NOTA-Evans Blue, PET/CT = positron emission tomography/computed tomography.

Keywords: $^{68}$Ga-NOTA-Evans Blue, bladder, bone, hemolymphangioma, positron emission tomography/computed tomography

1. Introduction

Hemolymphangioma is a congenital and benign malformation of both lymphatic and blood vessel.[1] It is preferentially situated in the head and neck, seldom in the spleen, small intestine, orbit, pancreas, vermiform appendix, or omentum.[2,3] Hemolymphangioma rarely involves bones and bladder simultaneously. At present, there are about 70 cases of lymphangioma that have been reported. This is the first case of lymphangioma involving both bladder and bones simultaneously. $^{68}$Ga-NOTA-Evans Blue (NEB) positron emission tomography/computed tomography (PET/CT) is a new lymphoscintigraphy technique,[4] which could detect the anatomy and function of the lymphatic system. $^{68}$Ga-NEB PET/CT might evaluate lymphatic disorders more accurately than $^{99m}$Tc-SC lymphoscintigraphy.[4]

In this report, we present a very rare case of a 12-year-old male hospitalized in our hospital for recurrent episodes of chyluria for 8 years and progressively worsening gross hematuria with right hip pain for 6 months. Finally, this patient was diagnosed hemolymphangioma by histopathological analysis of biopsy on pubic and ischial bones.

2. Case report

A 12-year-old male was hospitalized for recurrent episodes of chyluria for 8 years and progressively worsening gross hematuria with right hip pain for 6 months. $^{68}$Ga-NEB is a new PET tracer to evaluate the disorder of the lymphatic system.[4] Several studies have reported the utilization of $^{68}$Ga-NEB in lymphatic related
endothelium. The patient’s histopathologic immunostaining for CD34 and CD31 are specific in vascular endothelium.

The application of traditional radiological examinations, such as CT and magnetic resonance imaging (MRI), alone can be a challenge in making the diagnosis of hemolymphangioma. The radiological appearance of the hemolymphangioma on CT or MRI is not specific, because it is a mixture of multiple tissues. For those lesions mainly composed of water-based substance, such as hemolymphangioma, cyst, and abscess, CT and MRI provide only limited information instead of the origin of the fluid. The cyst component of the hemolymphangioma will not show enhancement on contrast-enhanced CT, and the capsule wall might demonstrate no, mild, or uneven enhancement. The presentations of hemolymphangioma in enhanced CT were variable, thus making the diagnosis difficult. Hemolymphangioma might present low or high signal intensity on MRI, which mainly depends on the proportion of lymphatic and vascular vessels. Nevertheless, 68Ga-NEB PET/CT is a specific modality for assessing the lymphatic system because it could provide comprehensive information about lymphangial positions and accurate guidance for biopsy. Hemolymphangioma invading bladder should be taken into consideration in those patients with chyluria after ruling out kidney disease. According to the literature, it might take patients less time for 68Ga-NEB PET/CT lymphoscintigraphy than 99mTc-SC lymphoscintigraphy, and 68Ga-NEB PET/CT lymphoscintigraphy could provide more information about the anatomy and disorder of lymphatic system than 99mTc-SC lymphoscintigraphy, and the images of 68Ga-NEB PET/CT lymphoscintigraphy are more revealing than 99mTc-SC lymphoscintigraphy. Besides the lymphatic system, 68Ga-NEB PET/CT lymphoscintigraphy also reveals distributions of radioactivity in the liver, kidneys, spleen, and cardiac blood pool. Some lymphatic drainage diseases have been reported to be detected by 68Ga-NEB PET, such as lymphedema, lymphangioma, lymphangioleiomyomatosis, lymphatic cyst, chylothorax, and chylolymphoedema.

**Figure 1.** The 68Ga-NEB PET/CT images (A, MIP; B–D, transaxial PET, CT, and fusion; E–G, transaxial PET, CT, and fusion; H–J, transaxial PET, CT, and fusion). 68Ga-NEB PET/CT results show several round and strip-shaped low-density shadows with mildly increased radioactive uptake in both bladder (B–D, blue arrows) and pelvis, including sacral, pubic, and ischial bones (E–J, blue arrows). CT = computed tomography, MIP = maximum intensity projection, NEB = NOTA-Evans Blue, PET = positron emission tomography.
dose CT, which has relatively lower spatial resolution and
definition in soft tissue compared with high-resolution CT or
enhanced CT. Compared with MRI, CT could only provide
limited information about soft tissue. The symptoms of chyluria
and hematuria disappeared and pain mitigated after the
treatment of Chinese traditional medicine 6 months ago, but
the specific mechanism of Chinese traditional medicine mitigating
the disease is still not clear.

Although hemangiolympangioima is a benign tumor, it still
has the potential to invade the ambient organs and relapse after
therapy. The surgery therapy was considered the most
effective treatment for hemangiolympangioima, and the excision
extension should contain the surrounding tissue suspected of
being invaded. It was reported that the lesions would recur
after complete excision in the percentage of 10% to 27%, while
the recurrence rate was 50% to 100% for part resection. The
nonsurgical therapy was less effective than operation, such as
radiotherapy, laser treatment, the injection of curing agent, and
cryotherapy. Hemolympangioima presents a developmental
defect of combined vasal and lymphatic vasculature. 68Ga-NEB
PET/CT is a specific method for the lymphatic system, which
might provide more accurate and comprehensive information
about the disorder of the lymphatic system compared to CT and
MRI. When patients suffering from lymphatic related disease are
suspected, 68Ga-NEB PET/CT should be recommended.

**Author contributions**

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