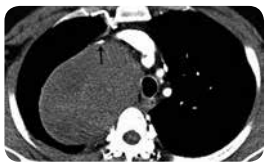

THE VALUE OF 18F-FDG PET/TC IN THE ASSESSMENT OF PERIPHERAL NERVE SHEATH TUMORS

Utilidad de la 18F-FDG PET/TC en la valoración de tumores de la vaina nerviosa

Mónica Vidal[1]
Juan Esteban López[2]
Ana María Alvarado[3]
Alejandro Velez[4]

Key words (MeSH)

Malignant peripheral nerve sheath tumor
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Palabras clave (DeCS)

Tumor de nervio periférico
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Neurofibromatosis
Nervios espinales

Summary

18F-FDG PET/CT is a useful imaging modality in the diagnosis and follow-up of peripheral nerve sheath tumors, especially in the assessment of tumor grade and biopsy guidance. The case of a 34-years-old woman diagnosed with peripheral nerve sheath tumor located in the posterior mediastinum that generated superior vena cava syndrome and Horner syndrome is presented. 18F-FDG PET/TC was performed to assess the possibility of malignancy. An 18F-FDG PET/CT was performed to determine whether it was benign or malignant, a heterogeneous mass with hypermetabolic areas with a maximum standardized uptake value (SUVmax) of 8.5 was found, and suggested malignancy with multiple grades of differentiation. A tumor biopsy from the region of higher metabolism was recommended with pathology result of malignant peripheral nerve sheath tumor.

Resumen

La 18F-FDG PET/TC tiene un papel importante en la evaluación de los tumores de la vaina nerviosa periférica, especialmente para determinar la posibilidad de malignidad y el sitio idóneo para la toma de biopsia. Se expone el caso de una mujer de 34 años de edad con diagnóstico de tumor de vaina nerviosa periférica, localizado en el mediastino posterior, que generó síndrome de vena cava superior y síndrome de Horner. Se realizó 18F-FDG PET/TC para hacer el diagnóstico diferencial entre benignidad y malignidad. Se encontró masa heterogénea con áreas hipermetabólicas que alcanzaban un SUV_{max} (valor de captación estándar máximo) de 8,5, hallazgos que sugerían origen maligno con diferentes grados de diferenciación. La biopsia de los lugares con mayor metabolismo arrojó el resultado de tumor maligno de vaina nerviosa periférica.

[1]Nuclear Doctor, Hospital Pablo Tobón Uribe. Medellín, Colombia.

[2]Radiologist, Hospital Pablo Tobón Uribe. Medellín, Colombia.

[3]Resident of Radiology, CES University. Medellín, Colombia.

[4]Pathologist, Hospital Pablo Tobón Uribe. Medellín, Colombia.

Introduction

Peripheral nerve sheath tumors are those that originate in Schwan's cells. They can be benign or malignant. Malignant tumors are very aggressive, with early metastases and low survival rates. Early surgical treatment with wide resection margins is generally the only effective treatment (1). It is indispensable to recognize, clinically and radiologically, these tumors for the timely treatment of patients, as well as to identify the ideal site for taking biopsies.

Clinical case

Female patient of 34 years old, with three months of burning pain in the right upper limb, no improvement despite the use of non-steroidal anti-inflammatory drugs.

Later, with edema in right hemiface, ipsilateral ptosis, anisocoria, anhydrosis and worsening of the pain in the extremity. Chest x-ray showed a right apical mass. Computerized tomography (CT) of the neck, thorax and abdomen was performed, with finding of mass in the posterior mediastinum, right paratracheal, with invasion of the right radicular emergency foramen T1-T2 and the medullary canal, which compresses the superior vena cava and is associated with blast lesions in the vertebral bodies from C3 to T1 (figure 1). A magnetic resonance imaging (MRI) with spinal contrast medium (figure 2) was performed, demonstrating a heterogeneous mass, with enhancement after administration of the contrast medium, right upper paravertebral location, invading the lateral channel of T1-T2, with intradural invasion and signs of myelopathy. Due to the findings, percutaneous biopsy of the lesion was performed as a result of benign fusocellular lesion pathology, with neural aspect without atypia, pleomorphism or tumor (figure 3).

Due to the rapid growth of the lesion, clinical worsening and blast lesions, the suspicion of malignant tumour persists. Surgery was performed to decompress the medullary canal with a new sample for pathology of the paravertebral region. The result indicated low-grade benign peripheral nerve sheath tumor.

An 18F-FDG PET/CT study was performed to stage the tumor, characterize its metabolic behavior, and determine the ideal biopsy site. The 18F-FDG PET/TC demonstrated mass with peripheral hypermetabolism and central amethobolic areas, with SUVmax of 8.5 in the anterosuperior region of the mediastinum. This metabolic behavior suggests malignant origin with different degrees of differentiation, so a new sample was taken in the region with the highest 18F-FDG enhancement (Figure 4). The new mediastinoscopic biopsy demonstrated malignancy within the context of a heterogeneous/borderline tumor originating from a neurofibroma (Figure 5).

Discussion

Los tumores de la vaina nerviosa periférica se originan en los nervioPeripheral nerve sheath tumors originate in the spinal nerves and cranial pairs (except II). These tumors are derived from Schwan's cells and may be benign or malignant. Benign tumors include schwanomas, neurofibromas, and plexiform neurofibromas. Plexiform neurofibromas have an abundant extracellular matrix that is susceptible to malignant degeneration (1).

Malignant tumors of the peripheral nerve sheath are aggressive, with early metastases and low survival rates. They affect 2-5 % of patients with neurofibromatosis type 1. They can have sporadic origin or in a plexiform neurofibroma. Early surgical treatment with wide resection margins is generally the only effective treatment (2). Because of its poor prognosis, it is important to differentiate between malignant and benign lesions. Most have heterogeneous composition, with a combination of benign and malignant tissue, which complicates their evaluation as they can give false negatives in the biopsy (3).

Imaging of malignant nerve sheath tumors includes CT, MRI, and 18F-FDG PET/CT. MRI is the best modality to determine the anatomy of the brachial and lumbosacral plexus and although there are no findings that indicate with certainty the presence of malignancy, the following characteristics suggest it: ill-defined margins, perilesional edema, cystic degeneration/necrosis and heterogeneity in sequences with pre- and post-contrast medium administration T1 information (4, 5).

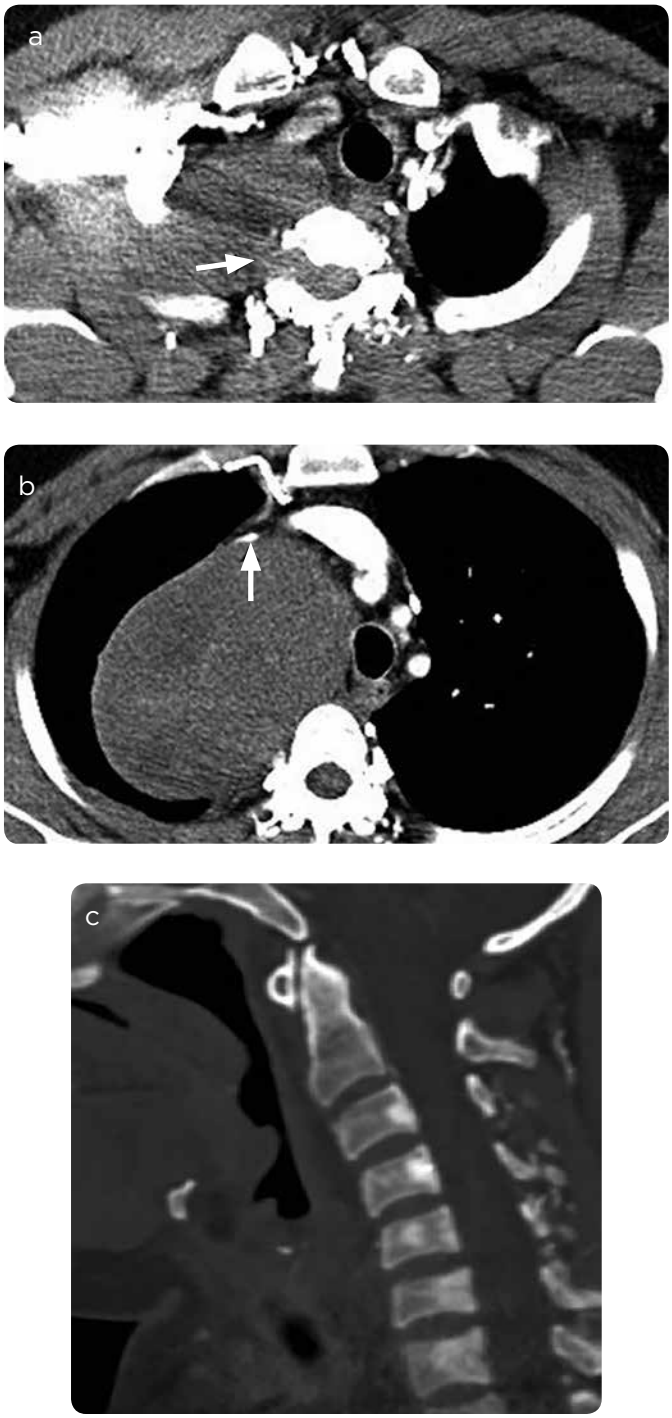

Figure 1. Chest CT with contrast medium. a and b) Mediastinum window: A well-defined mass of the posterior mediastinum invading the right root emergency foramen T1-T2 (arrow in a) and the central canal. It contacts the right lateral wall of the trachea by moving it to the left and the visceral pleura at the ipsilateral apex. It contacts and displaces the superior vena cava previously, causing a decrease in its calibre (arrow in b). c) Sagittal CT reconstruction of the neck in the bone window: Blast lesions in the vertebral bodies from C3 to T1.

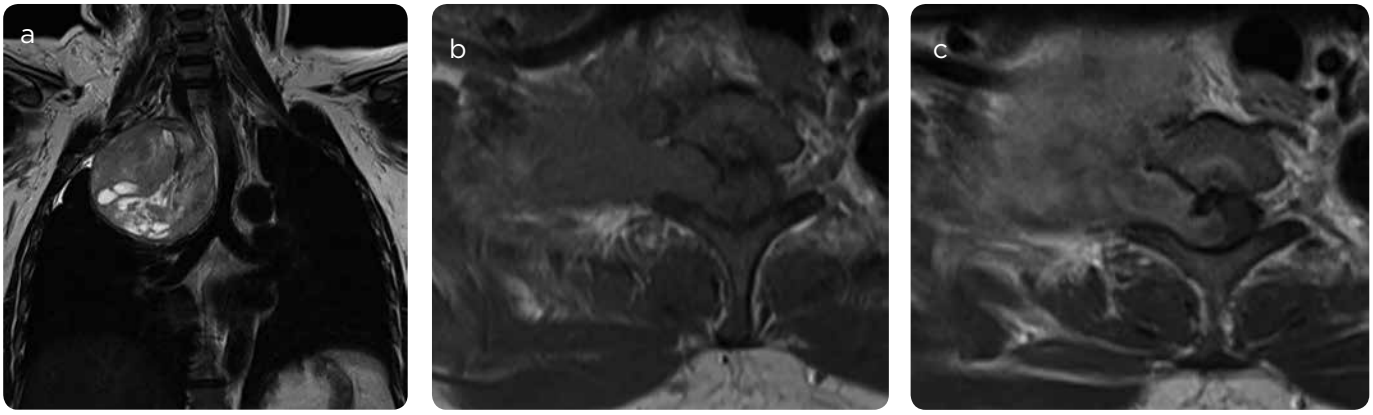

Figure 2. MRI with spinal contrast medium. a) Coronal, with T2 information, b) simple axial with T1 information, c) with contrast medium: right paravertebral mass, heterogeneous, with enhancement, invading the lateral channel of T1-T2. Intradural invasion with compression and displacement of the medullary cord and increase in its signal suggesting myelopathy.

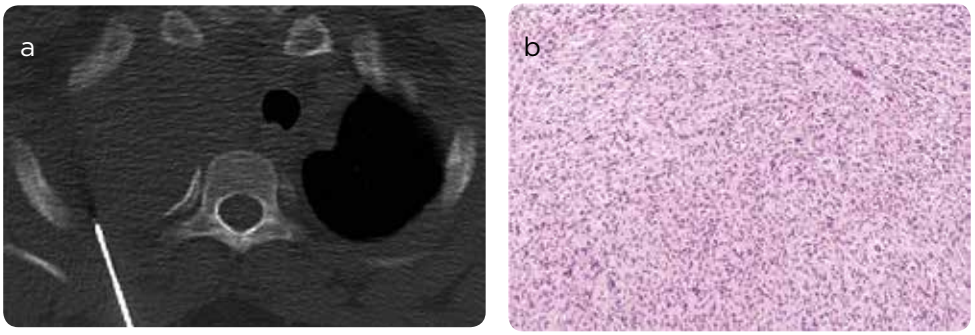

Figure 3. a). Percutaneous biopsy guided by tomography. b) Peripheral nerve sheath tumor. Benign fusocellular lesion of neural aspect without atypia, pleomorphism or tumor.

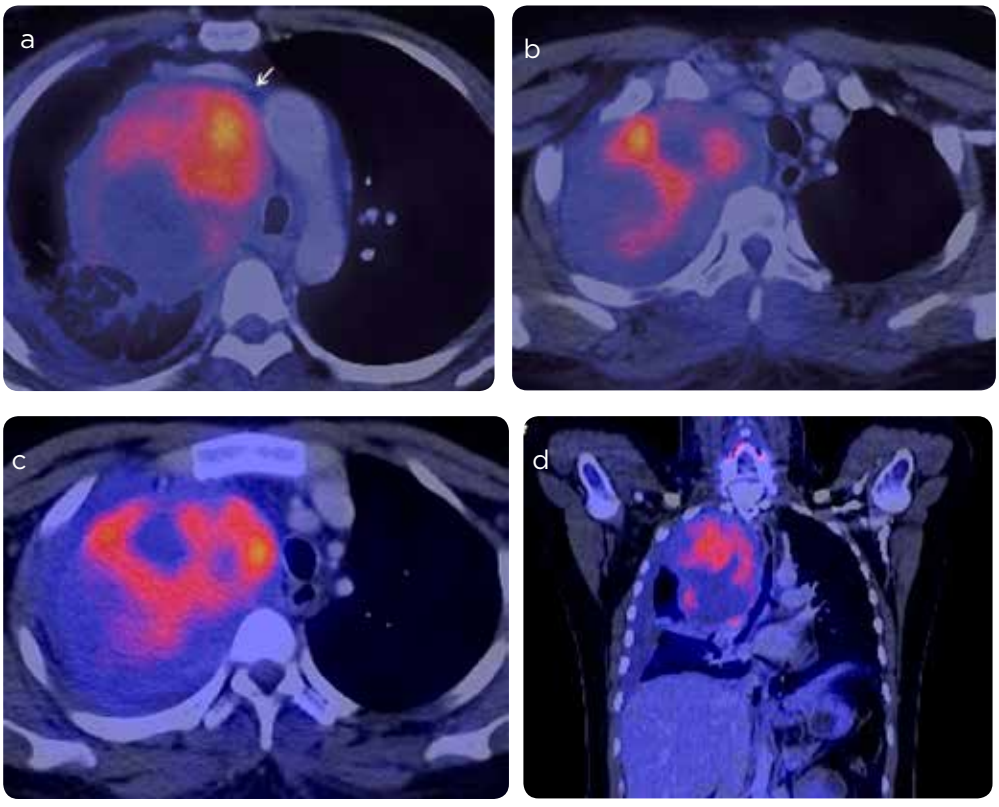

Figure 4. 18F-FDG PET/CT images: mass with peripheral hypermetabolism and central metabolic areas, with greater activity in the anteromedial and superior (a) regions, with SUVmax of 8.5.

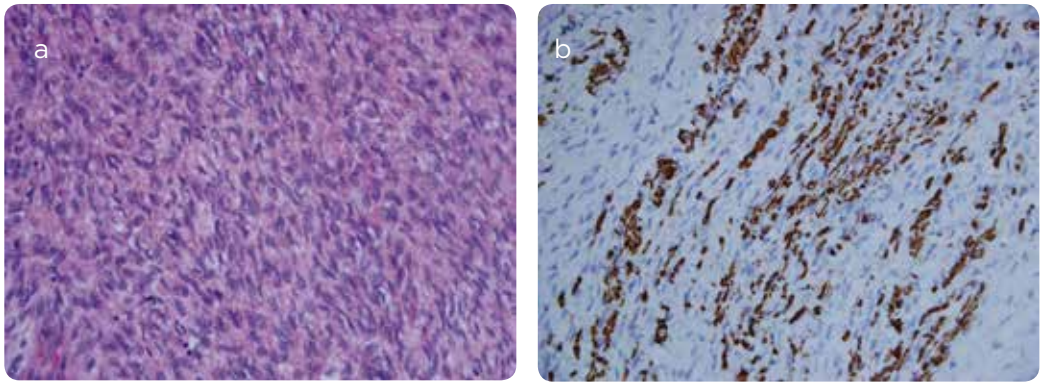

Figure 5. a) Hematoxylin and eosin. Fusocellular tumor with marked increase in cellularity and slight nuclear atypia. b) Immunohistochemical marker Ki 67 with 40% positivity.

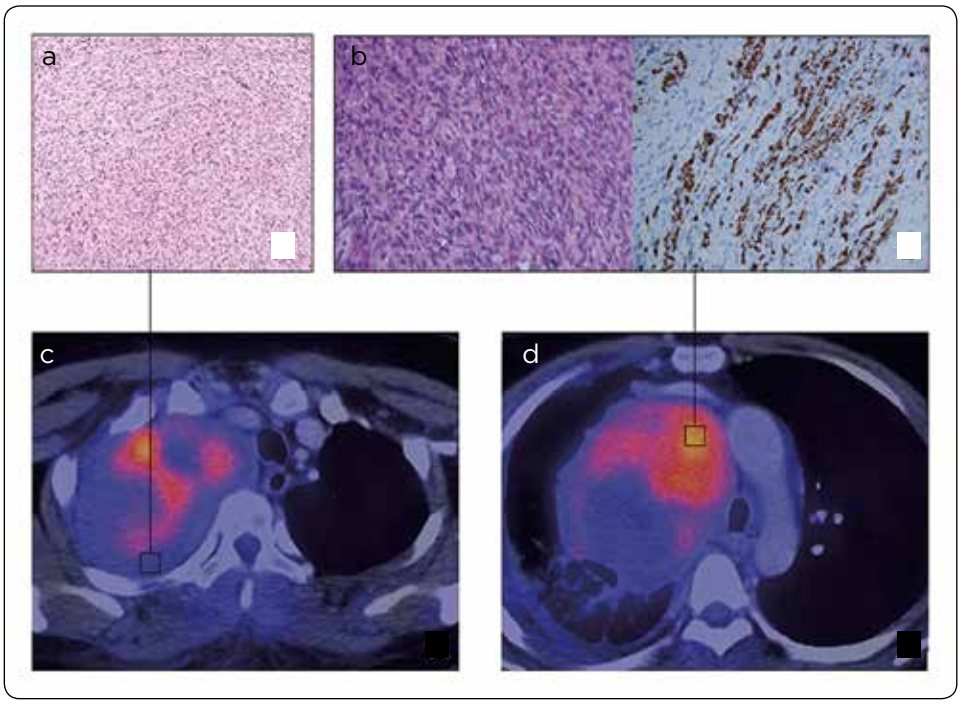

Figure 6. Correlation between biopsy results and 18F-FDG PET/TC: a) relationship of low metabolic activity and benignity result; b) high metabolic activity (SUVmax 8.5) with malignancy result.

The main objective of 18F-FDG PET/TC is to determine cellular metabolic activity, which is particularly high in oncological, infectious and inflammatory processes. Tumour metabolism is increased by cell de-differentiation. Tumour cells have an overexpression of glucose transporters (GLUT), secondary to the increase in glucose metabolism. The lower the degree of tumour differentiation, the higher the glucose expenditure and therefore the more aggressive the tumour (3, 6).

Peripheral nerve sheath tumors usually contain several cell types. The heterogeneous enhancement of both 18F-FDG and contrast medium is suggestive of malignancy (7). The SUVmax is a semiquantitative parameter that has a high sensitivity for the characterization of these tumors. An SUVmax greater than 3.5 is suggestive of malignancy in this type of tumors, with a sensitivity of 97% and specificity of 87%. For this reason, in some studies, it is the cut-off point for deciding whether the tumor requires histopathological study or resection (8, 9). Other parameters that can be evaluated with 18F-FDG PET/CT are tumor metabolic volume and total tumor glycolysis rate, which could be prognostic indicators (10).

18F-FDG PET/CT has the following advantages over other modalities: 1) high sensitivity for the detection of tumour lesions, 2) the capacity to determine cell behaviour and therefore is ideal for guiding the taking of biopsies, which should be in areas with higher metabolism, and 3) its usefulness in monitoring tumours, as it evaluates the response to treatment and the appearance of metastasis (1, 3).

The case of this study is interesting because, in the context of a peripheral nerve sheath tumor, 18F-FDG PET/CT was highly suggestive of malignant lesion and gave indispensable information for a third biopsy, which provided the definitive histological diagnosis (Figure 6).

Conclusion

18F-FDG PET/CT is a useful imaging modality in the diagnosis and follow-up of nerve sheath tumors, as it has high precision and sensitivity in the classification and staging of these tumors. Additionally, it serves as a guide for taking biopsies and provides prognostic information in the follow-up.

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Correspondence

Mónica Vidal
Departamento de Medicina Nuclear
Hospital Pablo Tobón Uribe
Medellín, Colombia
monicavidalg@hotmail.com

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