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Case Report

An Unusual Case of Intrapulmonary Schwannoma Associated with Sarcoidosis in Mediastinal Lymph Nodes

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Abstract

Background

The FDG uptake in schwannoma is variable. Studies report SUV ranging from 1.9 to 7.2. This can complicate the distinction between schwannoma, other neoplasms and non neoplastic lesions prior to histologic diagnosis. Here, we present an unusual case of intrapulmonary schwannoma associated with sarcoidosis in mediastinal lymph nodes.

Case report

A 75-year old asymptomatic man presented with a right lower lung nodule along with multiple enlarged lymph nodes in the mediastinum. The 18-fluorodeoxy-glucose (FDG) positron-emission tomography (PET) showed the pulmonary nodule to be slightly hypermetabolic, with a standardised uptake value of 3.3, as well as hypermetabolism in the enlarged lymph nodes. Endobronchial ultrasound-guided transbronchial needle aspiration of lymph nodes showed well- defined non-caseating granulomas without demonstrable microorganisms, which was consistent with sarcoidosis. The CT-guided transthoracic biopsy of the nodule led to a pathological diagnosis of schwannoma. Given the elevated SUV in the pulmonary nodule, a surgical resection was performed, which confirmed the diagnosis.

Conclusion

Intrapulmonary schwannomas are uncommon neoplasms. Preoperative evaluation can be difficult and an elevated SUV does not preclude the diagnosis. Surgical resection will often be needed to rule out more aggressive neoplasms; moreover, the complete excision appears to be an adequate treatment. Whenever possible limited resections should be favoured.

Keywords: Schwannoma; FDG-PET; Sarcoidosis

Introduction

Intrapulmonary schwannomas account for only 0.2% of pulmonary neoplasms [1]. The FDG uptake in schwannoma is variable and, according to the current literature, can reach a standardised uptake value (SUV) as high as 7.2. This can complicate the distinction between schwannomas, other neoplasms, and non-neoplastic lesions prior to pathological evaluation [2]. Here we present an unusual case of intrapulmonary schwannoma with moderately elevated SUV, associated with hypermetabolism in mediastinal lymph nodes secondary to sarcoidosis.

Case report

A 75-year old man was referred to our center after the incidental finding of a 25-millimeter nodule in the right lower pulmonary lobe on a computed tomography (CT) scan performed in the context of nephrolithiasis (Figure 1). There were also multiple enlarged infra-carenal and bilateral hilar lymph nodes. The 18-fluorodeoxy-glucose (FDG) positron-emission tomography (PET) showed the pulmonary nodule to be slightly hypermetabolic, with a SUV of 3.3, as well as hypermetabolism in the enlarged lymph nodes. The patient was completely asymptomatic. There was a history of smoking that was stopped twenty years ago as well as emphysema.

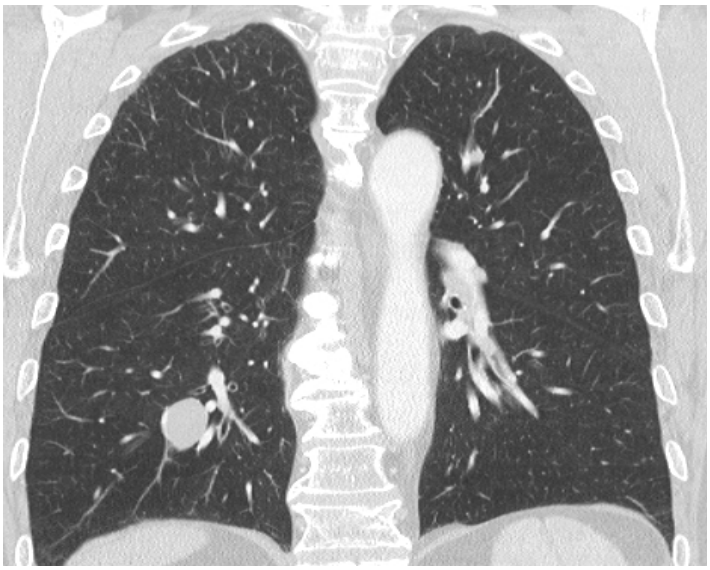

Figure 1. Chest computed tomography scan showing a well-circumscribed 25-millimeter nodule in the right lower pulmonary lobe.

Endobronchial ultrasound-guided transbronchial needle aspiration of lymph nodes stations 11R, 11R and 7L, as defined by the Internal Association for the Study of Lung Cancer (IASLC), showed non-caseating well-formed granulomas consistent with sarcoidosis. The CT-guided transthoracic biopsy of the pulmonary nodule led to a pathological diagnosis of schwannoma. Given the elevated SUV in the pulmonary nodule, the institutional tumour board suggested proceeding with a limited surgical resection. However, at surgery, the limited excision of the nodule was not feasible because of the position of the nodule inside the lobe. Therefore a right lower lobectomy was performed. Lymph node stations 11R, 7R and 11R were also sampled. The patient had a favourable evolution in the post-operative period.

Macroscopic evaluation of the lobectomy specimen showed a well-defined 2.4 cm tumor located at the bifurcation between two bronchi. Microscopically, the tumor consisted of a well-circumscribed proliferation of spindle cells with slightly enlarged nuclei and abundant cytoplasm (Figure 2). The tumor showed typical areas of Antoni A pattern with nuclear palisading and Verocay bodies. The neoplastic cells expressed S100, vimentin and pan-keratin (focally), while they were negative for thyroid transcription factor 1 (TTF-1), smooth muscle actin, desmin, CD34, and melan A. The proliferative index assessed by MIB-1 was less than 5%. These findings were consistent with schwannoma. The margins were free of tumor and there was no pleural invasion. The lymph nodes lacked metastatic involvement but revealed non-caseating, well-formed granulomas without demonstrable microorganisms, and were interpreted as consistent with sarcoidosis.

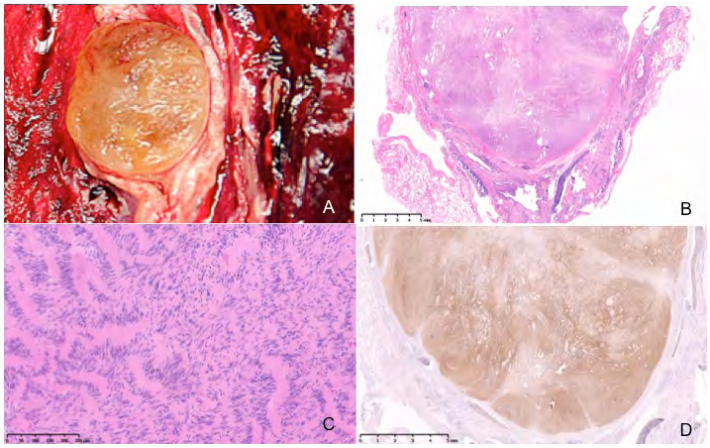

Figure 2. Macroscopic and histologic features. Well-defined tumour located at a bronchial bifurcation (A). Well-circumscribed proliferation of spindle cells (low magnification, hematoxylin-eosin stain) (B). Antoni A pattern areas with Verocay bodies (high magnification, hematoxylin-eosin stain) (C). Positive S100 immunostaining of the spindle cells (high magnification) (D).

Discussion

In the present report, we present a case of schwannoma with an unusual intrapulmonary location, increased SUV, in combination with mediastinal lymph node involvement by sarcoidosis. A study found only four primary intrapulmonary neurogenic tumours out of 1664 patients with pulmonary neoplasms (0.2%) - three of these were schwannomas [1]. A review of 62 cases of intrapulmonary schwannomas showed that 55% orig-

inated near the terminal segmental bronchus. Most peripheral lesions were asymptomatic [3].

The FDG uptake in schwannomas is variable. A series of nine cases showed SUV ranging between 1.9 and 7.2. This can help distinguish schwannomas from other neoplasms but the diagnosis is challenging prior to pathological evaluation [2]. The preoperative evaluation in this case was further complicated by the hypermetabolism observed in mediastinal lymph nodes, likely related to idiopathic sarcoidosis or sarcoid-like reaction to the tumour. The prevalence of sarcoidosis ranges form less than one to 40 cases per 100,000 people depending on estimates. About 95% of patients with sarcoidosis show bilateral hilar lymph node enlargement, with or without mediastinal lymph node enlargement [4]. A study found a median maximal SUV of 8.2 (range 1.2-16.5) in sarcoidosic mediastinal lymph nodes, which can therefore mimic malignant lymphadenopathy [5]. To our knowledge, this is the first report of intrapulmonary schwannoma combined with mediastinal lymph node sarcoidosis which both showed increased SUV at PET-scan evaluation.

Schwannomas are generally considered neoplasms with low malignant potential. Many authors favor a limited resection whenever it is technically feasible, [3, 6, 7]. However Domen et al. suggested that secure margins should be achieved given reports of a local recurrence arising from an inadequately removed tumour and malignant transformation occurring in a previously quiescent schwannoma [8]. In cases for which follow-up was available, the prognosis was excellent when a complete resection had been achieved [9, 10].

Conclusion

Intrapulmonary schwannomas are uncommon neoplasms. Preoperative diagnosis can be difficult and an elevated SUV does not preclude the diagnosis. Surgical resections are often needed to rule out more aggressive neoplasms and to confirm the diagnosis made on small biopsy specimens. Futhermore, complete resection appears to be the treatment of choice as prognosis is excellent when complete resection with negative surgical margins is achieved.

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