Introduction

Hamartoma can be defined as a non-neoplastic malformation or inborn error of tissue development constituted by a mixture of tissue. Hamartomas can occur in any area of the body, with a predilection for the lung, kidney, and intestine. Respiratory epithelial adenomatoid hamartomas (REAHs) are rare and most often of the pure epithelial type first described by Wenig and Heffner in 1995.1 REAHs usually arise from the posterior nasal septum, paranasal sinuses, or olfactory cleft. We report a case of nasopharynx REAH mimicking a malignant tumour with incidental high 18-FDG uptake in a patient with colon cancer. Less than five similar cases have been reported to date, and this is the first case of REAH to show high uptake on PET/CT scans. Although hamartoma arising from the nasopharynx region is very rare, it should be considered in the differential diagnosis because it is a benign lesion and complete surgical resection is curative.

Case

A 47-year-old male smoker with a history of colon cancer was referred to our department for disease staging due to high 18F-FDG uptake in the nasopharynx on PET/CT (SUVmax: 11.2). The patient was asymptomatic and had no history of any other sinonasal disease. PET-CT revealed a mass in the nasopharynx that partially obliterated the airway. Endoscopic evaluation demonstrated an irregular polypoid structure on the posterior nasopharyngeal wall. Endoscopic examination revealed no evidence of nasal polyp or papillomas. A primary nasopharyngeal cancer was suspected due to the examination and PET-CT uptake (Figure 1). Therefore, biopsy specimens were taken endoscopically from the nasopharynx. The histopathological diagnosis was consistent with REAH. Histopathological examination revealed that subepithelial glands proliferated in continuity with the overlying epithelium. The stroma was well vascularized, edematous, and contained scattered chronic inflammatory cells with a prevalent eosinophilic component. The glands were lined by ciliated respiratory epithelium of various sizes, and separated by stromal tissue, characteristic of the diagnosis of REAH (Figure 2). Pathologists confirmed the lack of evidence of diseases such as inflammatory polyp, inverted papilloma, squamous cell or adenocarcinoma of the nasopharynx. Excision was performed under general anesthesia through the nasal endoscope using a bipolar suction forceps and rotation suction knife (shaver). Because of the irregularity of the tumour, en bloc resection of the specimen could not be performed. Follow-up at 12 months has not shown evidence of disease recurrence.

Discussion

Hamartomas are caused by errors in tissue development and can grow from any tissue in the body, including surface epithelium, seromucinous...
glands, fibrous stroma, or vessels. Hamartomas can occur at any body site, but they are most commonly encountered in the lungs, liver, spleen, kidneys, and intestines.

Wenig and Heffner first described respiratory epithelial hamartomas in 1995, including 31 cases with symptoms at presentation: facial pressure, facial pain, nasal congestion, headaches, and hyposmia. The most common site of occurrence was the nasal septum, and paranasal involvement was quite rare. Vira et al. described 54 cases, which is the largest series described to date, and suggested REAH predominance within the sinus contents rather than the nasal cavity (85% of 54 patients). Nasopharyngeal occurrence is extremely rare, and less than five cases have been reported in the literature to date.1,3

The characteristic glandular components consist of respiratory epithelium originating from the surface epithelium with an absence of seromucous glands (Figure 2); the polypoid growth results from the respiratory epithelia-lined adenomatoid proliferation.1 REAH must be differentiated from other diseases, especially malignant tumours, and includes inflammatory polyp, inverted papilloma, low grade adenocarcinoma in the sinonasal tract, and nasopharyngeal carcinoma (NPC). Histopathological examination of the tissue can differentiate between these different disease entities. Malignancies demonstrate some degree of cellular atypia, pleomorphism, and increased mitotic index, which are not present in REAH and lymphoid hyperplasia.

Two clinical types of REAH in the nasal cavity have been described previously: solitary pseudotumoural lesions confined to the olfactory cleft and lesions that frequently occur in the ethmoid region are associated with nasal polyposis. Most REAH lesions in the olfactory clefts have been suggested to regress after the excision of nasal polyposis or to

Figure 1
Axial PET/CT images showing a polypoid mass in the nasopharynx (SUVmax 11.2) that mimics nasopharyngeal carcinoma (NPC).
The amount of FDG uptake on PET/CT is not a decisive indicator of malignancy because moderate to extremely intense (SUVmax: 5 to 40) uptake is seen with benign sinonasal papillomas. The degree of uptake cannot reliably differentiate between squamous cell carcinoma and benign papilloma, even when intense uptake is present. REAHs may have the same uptake characteristics as sinonasal papillomas. However, more REAH patients with high PET scan uptake are needed to confirm this hypothesis.

Endoscopic biopsy and pathological examination should be performed on nasopharyngeal masses in adults in order to make the differential diagnosis. In this case, REAH was successfully treated by a minimally invasive endonasal endoscopic approach and complete removal of the lesion from the nasopharynx without any morbidity because it was small and non-invasive. Because hamartomas do not have malignant potential, conservative surgery may be curative.

As a result, metabolic imaging with FDG-PET plays an important role in the management of oncological disease. However, false positive or negative FDG uptake should be excluded by physical and histopathological examination in a systematic manner. PET/CT is useful not only for cancer diagnosis and staging, but also for the detection and follow-up of other diseases.
References


Abdurrahman Bugra Cengiz
Yurtaslan Onkoloji Training and Research Hospital,
13.Street, No: 56 Demetevler
Yenimahalle
Ankara, Turkey
Tel.: 00904742251660
E-mail: drcengiz@gmail.com