CASE REPORT

Bilateral Olfactory Respiratory Epithelium Adenomatoid Hamartoma (REAH): A Forgotten Disease Entity

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ABSTRACT

Objective: **This clinical case report highlights the importance to recognize olfactory respiratory epithelium adenotomatoid hamartoma (REAH). Olfactory REAH is a benign disease that is located in the olfactory cleft. Olfactory cleft is an area which is often overlooked as it is situated in the narrow passage at the superior part of nasal cavities. Although olfactory REAH is rare, it is important to recognize this disease as it can cause a significant impairment towards a person's quality of life.**

Design: **A single case report.**

Introduction: **Respiratory epithelium adenomatoid hamartoma (REAH) is a rare, benign sinonasal glandular neoplasm. It is predominantly found in the nasal cavity followed by nasopharynx and anterior paranasal sinuses. The clinical presentation of REAH are non specific and it primarily mimics the symptom of chronic rhinosinusitis. Hyposmia is the symptom which is usually uniquely affecting patients with REAH that is located in the olfactory cleft.**

Case Report: **A 70-year-old housewife presented to us with increase hypersensitiveness to odour (hyperosmia) since 2 years ago. This symptom eventually led her to develop an avoidance behaviour towards certain offending odour until she could no longer tolerate her daily routine in preparing food for her household. She has moderate rhinitis symptom but there was no epistaxis, persistent headache or blurry of vision. Nasoendoscopic examination revealed a small polypoidal lesion at the bilateral olfactory clefts. CT scan showed soft tissue density within both of the olfactory clefts, which has caused widening of the clefts. Conservative management was opted with due consideration of the benign clinical disease entity as well as the patient's tolerance toward her symptoms following the commencement of intranasal corticosteroid.**

Conclusion: **Olfactory respiratory epithelium adenomatoid hamartoma (REAH) can significantly impair a person's quality of life if it is not treated. However, the disease can potentially be controlled with medication as shown in this case. This can eventually avoid unnecessary surgery.**

KEY WORDS

olfactory cleft, hyperosmia, rhinitis, hamartoma

INTRODUCTION

Respiratory epithelium adenomatoid hamartoma (REAH) is a rare, benign sinonasal glandular neoplasm. It predominantly affects men. REAH arises mainly from the nasal cavity. REAH can also occur in the nasopharynx and anterior paranasal sinus. The clinical presentation of REAH are non specific, which can mimic the symptoms of chronic rhinosinusitis and sinonasal malignancy. Hyposmia is the additional symptom which usually affects patient with olfactory REAH. Clinical examination may reveal polypoidal mass in the affected area. It is important to recognize REAH from chronic rhinosinusitis or sinonasal malignancy as REAH itself is a self limiting disease and can be safely managed conservatively. Early recognition of REAH may avoid unnecessary surgery as well as its potential surgical complications. We present a case of an elderly lady who develop hyperosmia which has significantly limit her performance in doing daily chores.

CASE REPORT

A 70-year-old housewife complaint of increase hypersensitiveness to odour (hyperosmia) for past 2 years. It was not preceded by head trauma or infection. This has led her to have avoidance towards selective offending odours, especially to onion and spices. Due to the hyperosmia, she has restricted her time spent in the kitchen. She preferred not to cook as it was no longer enjoyable. She would stay away from preparing meal, especially if the meal preparation involved slicing the onions. Fortunately, she was able to tolerate her hypersensitiveness toward the sweet smell of flower and parfum.

She has been under our follow up for moderate persistent allergic rhinitis since many years ago in which it has been well controlled with intranasal corticosteroids and antihistamine. She did not have epistaxis, persistent headache or blurry of vision.

Clinical examination showed no telecanthus. There was no hyponasal speech. Nasoendoscopic examination revealed a small, smooth polypoidal lesion at the olfactory clefts bilaterally. (Figure 1 and Figure 2).There was no ulcer, prominent vessel or nasal discharge. Nasal septum and turbinates were normal. Postnasal space examination was unremarkable. There was no palpable neck node.

Computed tomography of paranasal sinus showed soft tissue density within both the olfactory clefts that have caused minimal widening of the width of the clefts. The mass did not have any calcification or double density sign within. The adjacent bone was not eroded. (Figure 3)

Clinical diagnosis of respiratory epithelial adenomatoid hamartoma

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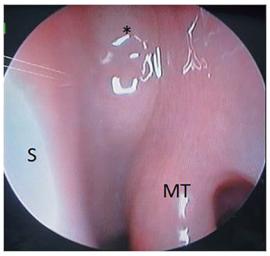

Figure 1: Endoscopic view of left nasal cavity at the olfactory cleft region. *polypoidal mass at the olfactory cleft. S= nasal septum MT = middle turbinate

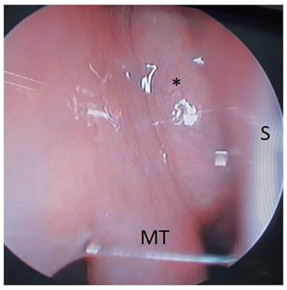

Figure 2: Endoscopic view of right nasal cavity at the olfactory cleft region. *polypoidal mass at the olfactory cleft. S= nasal septum MT = middle turbinate

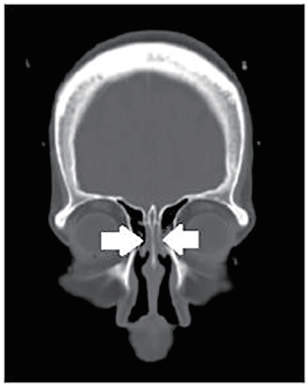

Figure 3: Coronal CT scan showed widening of the olfactory cleft bilaterally

(REAH) was made based on serial clinical examination which did not show any disease progression. The size of polypoidal mass at the olfactory region remained to be the same. The radiographical investigation was not suggestive of sinonasal malignancy or typical rhinosinusitis. We have also advised patient to be strictly adhered to her daily intranasal corticosteroids which has improved her hyperosmia to a tolerable level. In view of REAH is known to be a self-limiting disease, conservative management was opted.

DISCUSSION

Hamartoma was first described in 1904[1]. The term hamartoma is coined as primary, non-neoplastic malformations or inborn errors of tissue development and it is characterized by an abnormal mixture of tissues indigenous to a part of the body with an excess of one or more of the cellular components with the disease known to be self-limiting[2,3].

The term respiratory epithelial adenomatoid hamartoma (REAH) was subsequently introduced by Wenig and Heffner in 1995 as a subtype of hamartoma[3]. REAH mainly affects nasal cavity and it is most commonly involves the posterior nasal septum with unilateral predominance[3]. REAH predominantly affects male (80%), with age mostly affected between 30 to 90 years old[4].

The clinical presentation of REAH is non specific as it may mimics chronic rhinosinusitis and sinonasal tumour. They can present with nasal blockage, nasal congestion and epistaxis.

There is no specific characteristic for REAH on imaging. However, Lima et al studied CT scans comprising of 15 REAHs along with 36 patients with nasal polyposis and 49 patients without sinus diseases. It has shown REAH widen the widths of the olfactory clefts[5]. This finding corresponds to our clinical case which was shown on the coronal cuts of the CT scan.

The definitive diagnosis can be made by histopathology. In REAH, there is a proliferation and accumulation of glands and ducts lined by pseudostratified ciliated epithelial cells. There is no atypia seen. REAH also do not favour any specific immunohistochemical study. However, staining such as MIB-1 is done to differentiate it from neoplasms in which MIB-1 immunoreactivity is higher in neoplasm.[2]

Prolonged inflammation has been postulated as risk of developing REAH. To date, there is no clear aetiology of the pathogenesis of REAH. In a literature review done by Leet JT *et al*, 57.1% of the concurrent pathology in REAH group has the association with sinonasal polyposis, followed by 34.4% has chronic inflammation without sinonasal polyp and 8.6% has allergic fungal rhinosinusitis[6]. Patel DN *et al* described a single case report of REAH with previous history of radiation for nasal non melanotic skin cancer[7].

REAH has been treated with complete local excision. Fortunately, REAH has never been reported to cause malignant transformation, recurrence or progression of disease[4].

In view of its benign and self-limiting feature, REAH should be identified early by clinical history, endoscopic examination and radiological investigation to distinct it from other more aggressive diseases such as sinonasal malignancy, olfactory neuroblastoma, glioma, dermoid, inverted papilloma and lymphoma. This is crucial to avoid unnecessary surgery as REAH is a benign, non-progressive and self-limiting disease. The potential surgical risks of olfactory REAH include cerebrospinal fluid rhinorrhoea and anosmia due to its location. However, if during the follow up, the polypoidal mass appears suspicious of malignancy, it should not deter us from taking a histological biopsy and taking further action.

CONCLUSION

Olfactory respiratory epithelium adenomatoid hamartoma (REAH) can significantly cause impairment in quality of life if it is remain to be left untreated. The disease can be managed conservatively, controlled with medication as shown in this case, hence avoiding the unnecessary surgery and its potential surgical risk.

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