Case Report:
Chondro-osseous Respiratory Epithelial Adenomatoid Hamartoma (COREAH) : A rare entity of nasal mass
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Abstract:
Chondro-osseous respiratory epithelial adenomatoid hamartoma (COREAH) is an extremely rare benign lesion of the nose and paranasal sinuses. Clinically the lesion presents as a polypoid mass and histologically can be easily confused with more threatening tumors such as inverted papilloma and sinonasal carcinoma. A correct and accurate diagnosis is really important as all of these lesions are treated by different surgical approach. Here we report a case of COREAH in a middle-age lady who presented with unilateral nasal mass with chronic nasal obstruction.

Keyword: Hamartoma; nasal obstruction; paranasal sinuses; paranasal sinus neoplasms

Introduction
In 1904, Albrect, who is the first person to describe the term “hamartoma”, defined it as an aberrant cell differentiation which may produce a mass of disorganized but mature specialized cells or tissue indigenous to the particular site. Generally, head and neck hamartomas specifically in the sinonasal tract are extremely rare entities. Most of the hamartomas commonly found in the lung, liver, spleen, kidney, and intestine, but uncommon examples have been noted in the upper aerodigestive tract. The majorities of sinonasal hamartomas are from the mesenchymal origin and may rarely include epithelial elements. Respiratory epithelial adenomatoid hamartoma (REAH) consists of an epithelial component and chondro-osseous respiratory epithelial adenomatoid hamartoma (COREAH) is an example of hamartoma comprising both epithelial and mesenchymal components. These lesions are benign and do not clearly represent a neoplastic or an inflammatory process. However, they may grow to a large size and trigger a clinical concern. It is very important to recognize and differentiate this lesion from any malignant lesion of the sinonasal tract as the treatment is different. Here, we report a case of COREAH which was first diagnosed in our center.

Case report
46 years old lady presented with progressively worsening right nasal blockage for 3 years duration which was associated with rhinorrhea and anosmia. She had no symptoms of epistaxis, excessive sneezing, itchiness of the face, eyes or throat and no symptoms of post nasal drips. There were also no constitutional symptoms such as loss of weight and loss of appetite. Rigid nasoendoscopy showed a polypoidal yellowish mass which was arising from the middle meatus and extending until the floor of the nose and posterior nasal space (Figure 1).

Figure 1: Rigid nasoendoscopy showed polypoidal yellowish mass arising from the middle meatus. (S) = Septum, (IT) = Inferior turbinate

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Computed tomography (CT) scan showed a soft tissue mass with calcification located in the right nasal cavity, measuring 5.0cm (AP) x 1.6cm (W) with mixed opacity suggesting fluid, mucin and hypodense lesion seen in the right maxillary sinus. The mass caused widening of the right ostio-meatal complex (OMC) with minimal sclerotic changes of right maxillary sinus wall. The mass also compressed the right inferior turbinate and abutting nasal septum and lateral nasal wall medially, extending into the right anterior ethmoid air cells superiorly and protruding into the right nasopharynx posteriorly. It crossed the midline to the left nasopharyngeal region without any significant bony erosion (Figure 2&3).

The patient underwent excision of nasal mass via functional endoscopic sinus surgery (FESS). Intraoperatively, there was a firm, whitish mass seen originating from the right lateral wall just posterior to the uncinate process. The mass occupied the right middle nasal cavity, obstructing the right OMC and extended posteriorly to the left choanae. The tumour was excised successfully and no immediate complication documented. Grossly, the tumour is a firm polypoid lesion measuring 65mm x 28 mm x 15 mm (Figure 4). Cut section of the tumour shows a yellowish-white centre with areas of hemorrhage. Microscopically, the mass is covered by respiratory epithelium with fibrous stroma and contains multiple aggregates of tubular glands lined mainly by benign respiratory epithelium. The stromas are cystically dilated and contain eosinophilic material with easily identifiable mature bony tissue within it (Figure 5). There are no cartilaginous components, epithelioid granuloma or evidence of malignancy seen.
Discussion

Wenig and Heffner were the first to describe REAH back in 1995. In their series of 31 cases taken from the Otolaryngic Tumor Registry at the Armed Forces Institute of Pathology in United States, they had included one case which contained mixed epithelial and mesenchymal elements with osseous metaplasia. In a later abstract, the term COREAH was used by the same authors referring to this chondro-osseous lesion. The first reported case of COREAH was published in 2005 by Flavin et al. The exact etiology of COREAH is still unknown. To date, two hypotheses have been proposed for its etiology. The first theory is that similar to other hamartomatous lesions; COREAH is believed to be congenital in nature and results from an inborn developmental error.

The second hypothesis proposed is that COREAH may represent the effect of inflammatory process on nasal polyps. This theory explained the association of hamartomatous lesions with chronic rhinosinusitis. In 2004, Delbrouck et al. reported a case of REAH associated with nasal polyps which also show similar clinical presentation, histopathologic changes and behaviour with sinonasal inflammatory polyps. Despite their unknown pathophysiology, both REAH and COREAH are completely benign lesions and their proliferation is self-limiting.

In the majority of cases, REAH and COREAH originate in the posterior nasal cavity. About 70% of them arise from the posterior nasal septum. Other sites of occurrence include the maxillary sinus, ethmoid sinus, frontal sinus, and nasopharynx. In our case, the mass originated from the right lateral wall just posterior to uncinate process which is similar to previously reported cases. R. Flavin et al. in 2005 reported a case of COREAH in an 11 year-old boy that also originated from the lateral nasal wall. In another reported case, a 38 year-old lady showed a tumour attached to the lateral nasopharyngeal wall.

All reported cases of COREAH have a common presenting symptoms which include nasal obstruction, nasal stuffiness, rhinorrhea, hyposmia, epistaxis and symptoms of chronic rhinosinusitis with variable duration of symptoms from few months up to 8 years. Due to their similarities in clinical presentations as well as the histopathological findings, inflammatory polyps, inverted papillomas, and low-grade sinonasal adenocarcinoma should be considered as differential diagnoses. It is really important to distinguish COREAH from all those lists of differentials as they might need a significantly different treatment approach.

Clinically, most of the COREAH can be differentiated from inflammatory polyps according to its location and involvement which is at the posterior nasal septum. However, it can be tricky because it also can originate from the lateral nasal wall mimicking the inflammatory polyps as seen in our case. In this situation, CT scan may be helpful as it shows mass with lots of calcification. Histopathologically, both lesions may show similar features of fibroblastic and vascular proliferation with stromal edema and mixed inflammatory cell infiltration. Florid adenomatoid proliferation and stromal hyalinization present in COREAH are important histological features which distinguish it from inflammatory polyps which typically have loose edematous stroma. COREAH can be misdiagnosed as inflammatory polyps in cases of lack of adenomatoid proliferation. Inverted papillomas arise from the inverted growth of stratified squamous epithelium which is not seen in COREAH.

Inverted papillomas commonly involved the lateral nasal wall, locally aggressive, have a tendency for recurrence and has the potential for malignant transformation which is never been described in COREAH. Clinical presentation of COREAH may also mimick low-grade sinonasal adenocarcinoma which can originate from the surface epithelium as well as seromucinous glands of the submucosa. Histologically, sinonasal adenocarcinoma typically shows cribriform architecture, the absence of ciliated epithelium lining the glands, desmoplastic stroma, and an elevated mitotic rate.

In conclusion, despite its rarity and varied clinical presentations, COREAH should be considered as one of the differential diagnosis in patients presented with obstructive symptoms with unilateral nasal mass. COREAH is a benign lesion which is treated with complete local excision. In reported literatures, after a successful surgery there was no reported evidence of recurrence in 5 years follow up. Clinicians and pathologists must be aware of this entity because misdiagnosis of COREAH for an inverting papilloma or sinonasal adenocarcinoma may result in unnecessary excessive surgical procedures for the patient.
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References


