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Diagnosis and Treatment of Bilateral Respiratory Epithelial Adenomatoid Hamartomas With and Without Sinonasal Polyposis

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Case 1

A 68-year-old man presented in the consulting room of the Otorhinolaryngology MD of the "A.O. Papardo" in Messina. His complaints were nasal obstruction and anosmia without headache and nasal bleedings. The patient had a history of unsuccessful administration of corticosteroids and antihistamines.

Endoscopy (performed with rigid endoscope 30° angle Karl Storz) showed a protruding solid mass located between the middle turbinate and the nasal septum on both nasal cavities. In the left nasal fossa, the mass occupied all the olfactory cleft from the body of middle turbinate extending anteriorly under the nasal bones (Figure 1). Indeed, the right mass arised approximately next to the head of middle turbinate and protruded toward the roof of nasal fossa, occupying just one-third superior part of the olfactory cleft.

Both formations showed quite solid consistence (tougher than nasal polyps), low bleeding during palpation (in contrast to inverted papillomas and malignant neoplasms), and the chromatic features of common nasal polyps.

On coronal and sagittal planes of computed tomography (CT) scan, hamartomas appeared as hypodense expansive lesions that enlarged olfactory clefts on both sides and extended to the turbinate regions (Figure 1).

The CT scan showed on both sides an enlargement of the olfactory cleft, with a maximum distance of 10 mm between the 2 walls. This, in line with the hypothesis of Hawley and colleagues, is a suggestive sign of respiratory epithelial adenomatoid hamartoma (REAH).[1]

The patient underwent magnetic resonance imaging (MRI) for further investigation: An abnormal tissue proliferation bilaterally reaching olfactory clefts, more evident on the right side, was documented by T_1-weighted sequences.

Case 2

A 65-year-old woman presented to the consulting room of Otorhinolaryngology MD of the "A.O. Papardo" complaining nasal obstruction and anosmia. The anamnesis was negative for headaches, epistaxis, or rhinorrhea.

Flexible endoscopy was performed, revealing the presence of neoformations with a polypoid appearance occupying both the left and right middle meatus. Furthermore, it also showed 2 huge bilateral masses in the context of the olfactory clefts, resembling nasal polyps both for chromatic features and consistence.

The CT scan confirmed a solid tissue similar to a massive polyposis that widened the olfactory clefts. Subsequently, MRI also was carried out, which confirmed the sinonasal mass at the ethmoidal level (Figure 2).

Both patients underwent surgical treatment by centripetal endoscopic sinus surgery (CESS) with Draf III procedure (Lothrop): The portion of the nasal septum presenting neoformations has been removed,[2] eradicating all the pathology "en bloc." Furthermore, in the second patient, frontal–ethmoidal–maxillary sphenoidectomy has been performed for the treatment of concomitant sinonasal polyposis.

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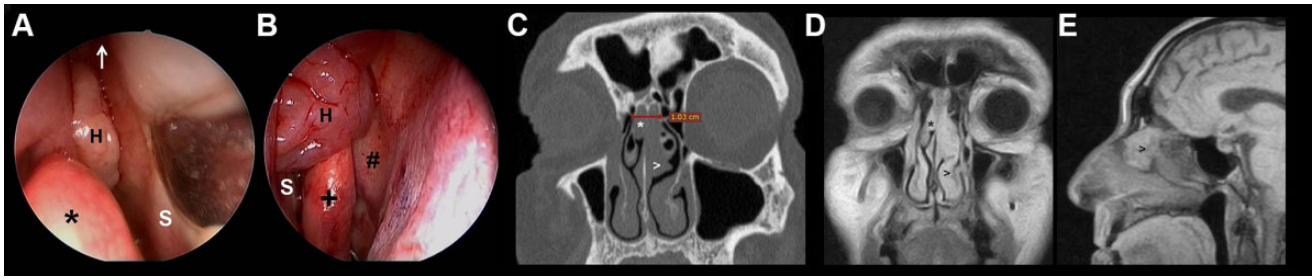

Figure 1. A and B, Endoscopic images of case 1: On right side (A) and left side (B), it's possible to see H (hamartoma), * (middle turbinate), > (nasal septum), ↑ (olfactory cleft), and # (uncinate process). C, Computed tomography scan of case 1 on coronal plane: Hamartoma appears as a hypodense expansive lesion that enlarges olfactory cleft on both sides and extends to the turbinate regions. It's possible to see > (hamartoma on left side) and * (hamartoma on right side). D and E, Case 1: Magnetic resonance imaging and T2-weighed sequence: On coronal (D) and sagittal (E) plane, hamartoma appears as a nonhomogeneous hyperintense tissue that extends from the olfactory cleft to the turbinate regions.

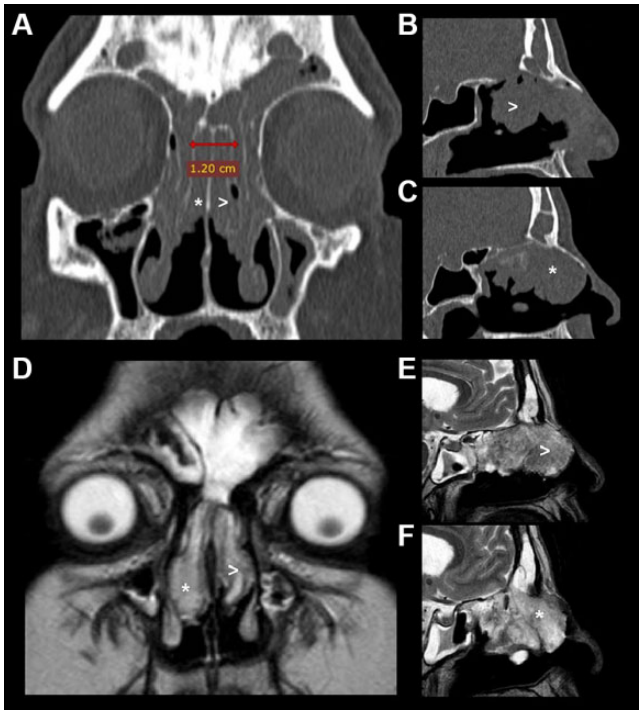

Figure 2. A-C, Case 2: Computed tomography scan on coronal (A) and sagittal (B, C) planes shows a hypodense tissue that enlarges olfactory cleft, thinning the surrounding bone and extending into the nasal cavities. D-F, Case 2: Magnetic resonance imaging and T_2-weighed sequences: coronal (D) and sagittal (E, F) planes show the presence of a nonhomogeneous hyperintense mass, arising from the olfactory cleft and extending to the turbinate regions.

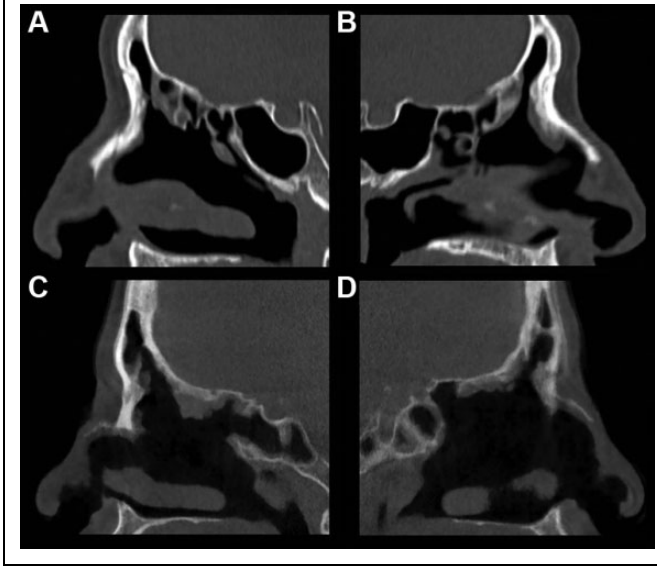

Figure 3. A and B, Case 1: Postoperative computed tomography scan on sagittal planes shows the excision of the neoformations. C and D, Case 2: Computed tomography scan on sagittal planes after treatment with corticosteroids shows disappearance of the polypoid lesions.

Histological analysis confirmed in both cases the diagnosis of glandular rhinosinusal hamartomas. After surgery, the second patient had a relapse of nasal polyps, which has been successfully treated. In both patients, after the interventions, there was no evidence of relapse of REAH (Figure 3).

A REAH is a benign proliferation of various differentiated epithelial cells with no atypia or metaplastic changes with an edematous stroma infiltrated by chronic inflammatory cells.[3,4]

The diagnosis of this pathology is very rare: Only 394 cases have been published as case reports or case series between 1995 and 2013.

The REAH can occur in nasal fossa, nasopharynx,[5] ethmoidal, maxillary, and frontal sinuses.[6,7] A bilateral REAH occupying both the right and left olfactory clefts, such the abovementioned, is not a common occurrence.[8-10]

As one could expect, the definitive diagnosis of REAH depends mainly on its histological features.[11]

Being able to feasibly distinguish between different benign pathologies is important to plan a proper therapeutical approach. We performed a subperiosteal dissection with adjacent bone drilling all done by CESS, in line with Bignami and colleagues, who stated that an aggressive resection with subperiosteal dissection and drilling into the adjacent bone gave the same outcomes of standard

endoscopic sinus surgery.[4,12] Sinonasal polyps can be often observed together with REAH.[1]

An interesting aspect that could show a difference between polyposis and REAH is that the latter can frequently occupy the olfactory cleft and cause a widening due to its growth.[13,14]

Together with clinical features, imaging findings of a discoid-shaped lesion reported on both CT scan and MRI T_1-weighted sequence with contrast medium, in absence of any bony damage, could suggest a possible REAH.[1,15]

Both MRI and CT scan are advanced tecniques with several applications in clinical imaging, and could be important to find features that could help to detect REAH[16-22] and evaluate its dimensions, shape, and location. Therefore, preoperative biopsy should be recommended if the presence of REAH is suspected,[4,16,17] for example, in case of widening of the olfactory clefts[1] or when a discoid-shaped lesion in the olfactory cleft is present without any bony changes.

Declaration of Conflicting Interests

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