CASE STUDY

Bilateral Respiratory Epithelial Adenomatoid Hamartoma With Atypical Behaviour

Hamartoma respiratorio epitelial adenomatoso bilateral de comportamiento atípico

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A 69 year-old man, with bilateral inflamed sinonasal polyps that had been treated medically for years, referred for surgical treatment suffering from moderate progressive bilateral nasal obstruction, sinus pressure with no rhinorrhea or other symptoms and typical bilateral inflamed polyps emerging from the middle meatus, not exceeding the level of the lower concha (Endoscopic Appearance Score of 8, Lindholdt scale grade 2 and Lund-Mackay grade 2 occupation shown on nasal endoscopy).

CT requested and we maintained medical treatment with fluticasone furoate 27.5 μg/day/nostril.

Twenty days later, awaiting the results, the patient reported a sudden worsening of symptoms: serious nasal obstruction, rhinolalia clausa and mild spontaneous nose-bleeds with massive growth of the ‘polyps’, each seen from both nostrils as voluminous reddish masses, prone to bleeding, deforming the nose. The CT revealed polyoid lesions filling the nostrils with mild diffuse chronic frontal ethmoidal maxillary inflammation, with no bone erosion, infiltration or spread to adjacent structures. Nuclear magnetic resonance revealed bilateral hyperintense polyps with hypointense axis on T2 sequences with origin in the upper area of the head and body of middle conchae (Fig. 1).

A complete endoscopic en bloc resection of the tumour was performed including a partial middle turbinectomy to minimise bleeding and achieve eradication, after control of the sphenopalatine pedicle flaps and conservative cleaning of the inflammatory sinus disease. We obtained 2 voluminous smooth elastic polypoid masses, yellow-reddish in colour (Fig. 1).

Histopathological analysis revealed bilateral respiratory epithelial adenomatoid hamartoma REAH) with inflamed mucosa (Fig. 2).

No recurrence was observed after 18 months’ follow-up.

Discussion

Hamartomas are ubiquitous benign tumours due to proliferation of local native tissue. Sinonasal REAH are a rare polyoid subtype with proliferation of the epithelial glands in continuity with normal epithelium and normal submucous glands, of slow expansive growth, non-aggressive, filling the nostrils and producing secondary symptoms.

They appear in adult males in the sixth decade of life, and their aetiology and precipitants remain unknown, although it has been suggested that they are induced by chronic inflammatory processes due to frequent concomitance, as in our
patient, although it cannot really be ascertained whether this inflammation is the cause or the consequence of REAH or whether it is simply concomitant. An embryological cause has also been proposed: the development of disorganised immature tissues and mature specialised cells, of which the trigger is unknown.\textsuperscript{2,3}

Their most common and specific location is the posterior area of the nasal cavity, of septal origin. Other described locations are: intrameatal, olfactory cleft, nasopharynx, intrasinusal and lower conchae.\textsuperscript{1,3\textendash}5

Patients suffer nasal obstruction and symptoms of non-specific chronic sinus disease, slowly progressing over months to years. REAH appear as voluminous, elastic, meaty polyps, but are more indurated than inflamed polyps, filling the nasal cavity, and, very exceptionally, they are bilateral.\textsuperscript{4,5} An independent bilateral presentation, both emerging from the upper part of the middle conchae, of rapid growth (20 days), congested reddish in colour, prone to bleeding, as in our case, has not been described before, as far as we know. In some publications we read

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\includegraphics[width=\textwidth]{figure1}
\caption{Clinical aspect. (A) Prior to surgery after administering vasoconstriction of the masses emerging through both nostrils. (B) Intraoperative period. (C) CT: polypoid masses growing from the middle conchae filling nasal cavities, mild signs of inflamed sinus and small polyps not compromising sinus drainage. Absence of bone erosion. (D) NMR: hyperintense lesions in T2 with hypointense axis.}
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\begin{figure}[h]
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\includegraphics[width=\textwidth]{figure2}
\caption{Histology. (A) Polymorphic epithelial glandular accumulations separated by fibrous stromal tissue and mixed reactive inflammatory infiltrate in continuity with epithelium (haematoxylin\textendash eosin 40\times). (B) Essentially normal glands in submucosa with atrophic changes, covered with a single layer of cuboidal epithelium (haematoxylin\textendash eosin 100\times).}
\end{figure}
that "hamartomas are not capable of constant growth and, therefore, their proliferation is self-limited". This is the case as long as there are anatomical barriers to this growth or in the absence of growth-promoting factors. REAH, however, grow uncontrollably in areas where there is no resistance, as in the nostril.

We cannot explain the behaviour described in our case: we did not find ischaemic, haemorrhagic or infectious processes inside the hamartoma. We can only conjecture exacerbation of inflammatory disease with glandular growth as the explanation. A pre-operative histological examination would have been appropriate; we did not obtain this, with reason, for fear of haemorrhage, and relied on the imaging studies.

CT and MR imaging are non-specific with no described typical pattern: they differ according to the proportions of the components of the REAH. Enlargement >10 mm of the olfactory clefts associated with sinusitis has been suggested as a characteristic; this enlargement was not observed in our patient. However, its expansive, non-infiltrating behaviour was demonstrable, suggestive of a benign process, and its origin and extension, and consequently we were able to plan treatment.

Histopathology showed polymorphic accumulations of glandular proliferations in continuity with the surface of the normal ciliated epithelium covering the lesion, fibrosis of the stroma with chronic inflammatory infiltrate separating the gland groups and oedema. Reactive atrophic changes were also observed and the absence of proliferation of submucosal glands.

A differential diagnosis should be made, especially with inflamed polyps, antrochoanal polyps, inverted papilloma, with very marked thickness of squamous epithelium and adenocarcinoma, with stroma between the glands, cellular atypia, pleomorphism, mitotic activity and aggressive infiltrating behaviour destroying underlying bone.

Conservative endoscopic surgical resection in a monoclob fashion is the treatment of choice. An external paralateralonasal approach can be used if there is no previous histological diagnosis. Surgery should include the seat of the lesion with healthy margin to the extent necessary to prevent both recurrence and iatrogenesis on the drainage of the paranasal sinuses or olfactory system. Concomitant inflammatory disease is treated in accordance with usual practice. If this requires surgical treatment, it should be performed as if there had been no previous REAH. It is also reasonable to consider surgery in 2 stages, with appropriate medical treatment to minimise sinus surgery or even avoid it. Recurrence is very rare.

In conclusion, although a unilateral refractory non-aggressive mass, concomitant with inflammatory disease, might raise suspicion of REAH, this suspected diagnosis is almost impossible if there is bilaterality or unusual behaviour. It is not always possible to obtain biopsy results before surgery, particularly when uncontrollable bleeding is to be expected due to its atypical behaviour. Treatment planning lies in knowledge of this disease and studying the images (origin, extension and non-malignant pattern) to achieve complete removal, minimise iatrogenesis and prevent unnecessary over-treatment given the non-specificity of CT and MR imaging.

Conflict of Interests

The authors have no conflicts of interest to declare.

References