Agenesis of the Vomer Bone

David Herrero Calvo, Mario Cuetos Azcona, Luis A. Vallejo Valdezate, and Luis M. Gil-Carcedo García
Servicio de Otorrinolaringología y Patología Cérvico-Facial, Hospital Universitario del Río Hortega, Valladolid, Spain

CASE STUDY

Thirty-four-year-old female attending the clinic due to chronic mild difficulty in breathing through the nose, together with a sensation of inflammation of the soft palate and occasional left earache. She has had a hypernasal voice since childhood and reports the emergence of the stomach contents through the nose with the effort of vomiting. She does not indicate any clinical signs of infection or any prior craniofacial trauma, as well as no consumption or substance abuse.

The endoscopic examination of the nose revealed an S-shaped deformity of the cartilaginous septum, as well as a complete defect of the bony wall from the medium turbine to the choanae; the entire mucosa structure is entirely lacking and the underlying bony skeleton (Figure 1). The rest of the nasal mucosa has normal colouring and no signs of acute or chronic rhinosinusal inflammation are evident.

The examination of the oropharynx and the oral cavity did not reveal any pathological finding. Otoscopy and acumetry results are normal, with type A (Jerger) tympanographic curves in both ears. Active anterior rhinomanometry provided flow values at 150 Pa of 589 mL/s in the right nasal fossa and 803 mL/s in the left nasal fossa.

The computerized tomography (CT) imaging study revealed the complete absence of the vomer bone, without alterations in the shape, bone density or pneumatization of the remaining bones of the face (Figure 2). The patient was seen again 9 months after diagnosis and no alteration in the nasal endoscopy findings was seen, and with hardly any variation in clinical situation, except for the disappearance of the left otalgia.

The nasal septum comprises the membranous wall, the quadrangular cartilage, the perpendicular lamina of the ethmoid, the intermaxillary and palatal crests, and the vomer.[1] This begins to grow in the form of 2 parallel laminas of cartilage

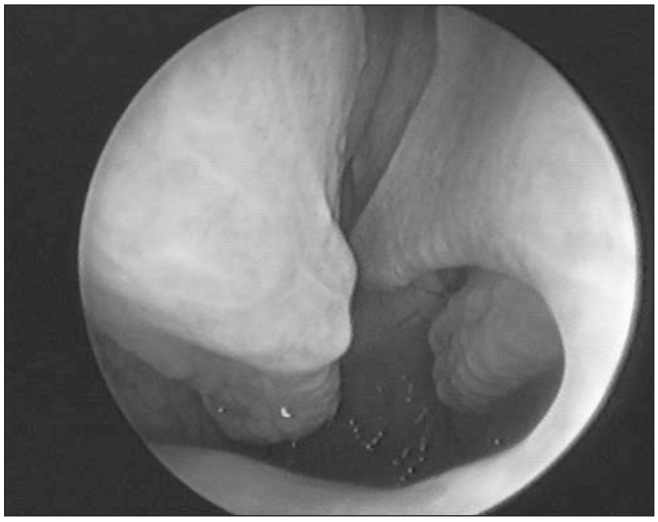

Figure 1. Image of the right nasal endoscopy in which it is possible to observe the defect in the bony septum. Note the tail of the contralateral inferior turbine.

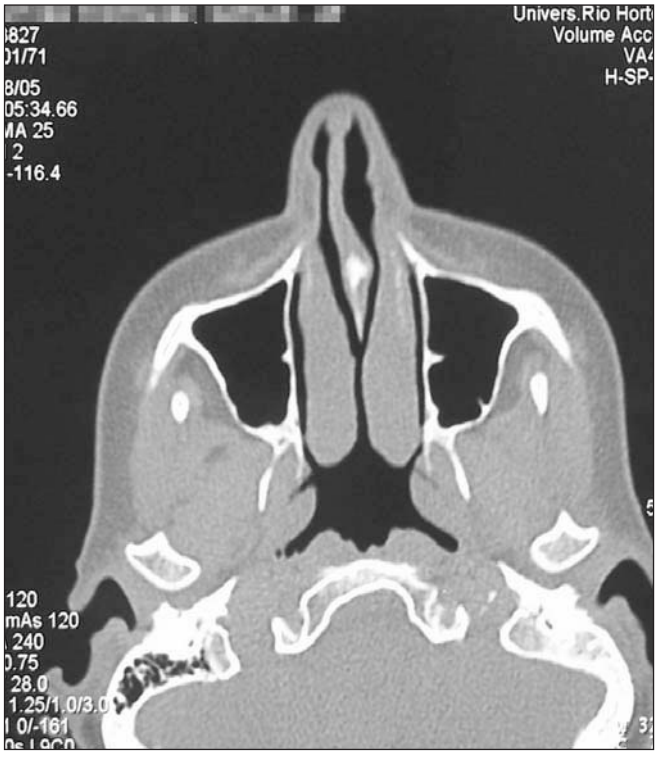

Figure 2. Axial computerized tomography revealing the absence of almost all the bony nasal wall together with the deformity of the cartilaginous portion. Note the hypertrophy of the posterior area of both inferior turbines.

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Correspondence: Dr. D. Herrero Calvo.
Alamillos, 9-11, 3.º B. 47005 Valladolid. España.
E-mail: dhcalvo@hotmail.com

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from the eighth week of gestation, at which point 2 ossification centres, one on either side, emerge. Around the third month, both these laminas begin to join together from the caudal region at the same time as they turn to bone and the cartilage is reabsorbed, as described by Sandikcioglu et al.[2] Later on, it begins to take on a Y-shaped morphology until it becomes completely fused, at around week 17 of gestation, and joins the rostral edge of the sphenoid. At the onset of puberty, the laminas are almost completely fused, although signs remain of their bilaminate origin in the wings of the upper edge and in the striation of the anterior border. On the other hand, it is essential to have a correct formation and union of the palatal bones and the palatal apophyses of both maxillary bones for the complete development of the vomer.[3]

Mohri et al[4] put forward 2 theories to explain the pathogenesis of this structural defect. First of all, they point to an "immature ossification centre" that might give rise to an alteration in the bone's development and the possible re-absorption of the original lamina of cartilage. On the other hand, they propose a "descending growth defect" in the primitive nasal septum, so that its caudal edge is prevented from progressing towards the floor of the fossa, and the normal ossification process cannot take place.

So far, only 9 cases of isolated agenesis of the vomer[5] have been described in the literature. Mohri et al[4] present a series of 6 cases, the most numerous group published, in the first reference in the scientific literature of a congenital defect of the vomer bone as a pathological entity.

In the cases published, the diagnosis was most often achieved during the third decade of life through the use of endoscopic techniques. A constant finding is the compensatory hypertrophy of the posterior third of both inferior turbines; in some cases this is accompanied by otitis media with effusion or other otological condition. We do not accept that the possible mild incompetence of the velar and palatal region suspected from the clinical presentation has any link with this structural defect, as other members of the same family present hypernasality of the voice despite lacking any anatomical alterations.

The rhinomanometry test provided flow/pressure curves that are consistent with the mild right septal deviation, with an increased flow for her gender at 150 Pa in the left nasal fossa. This fact, not cited in other cases described, may be due to the reduction in resistance because of the absence of choanae. Nonetheless, the scant symptoms presented by the patient, despite having such a striking defect in the septum, have led us to point out that large-scale resections of the posterior bony wall (due to congenital or acquired structural alterations) do not significantly alter the nasal ventilatory function and have much less functional repercussion than smaller defects in the cartilaginous septum.

In order to reach the diagnosis of agenesis of the vomer bone, it is necessary to rule out all the causes that might affect the nasal septum, such as traumatic injury, a prior history of surgery, abuse of inhaled substances, infections, inflammatory processes or neoplasias. Furthermore, it is necessary the assess the posterior floor of the fossa to discard the existence of a submucous cleft palate, as the absence of the vomer bone is a constant in patients presenting this defect.[6]

We share the opinion expressed by Yilmaz et al[5] that these isolated defects in the vomer bone are more numerous than reflected in the bibliography and that the ever more frequent use of endoscopic techniques and imaging tests will lead to an increase in the cases diagnosed.

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