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What is your diagnosis?

Atypical nasal mass

E. Drapier , M. Makeieff , X. Dubernard *

Service d'oto-rhino-laryngologie, CHU de Reims, rue du General-Koening, 51100 Reims, France

1. Case report

A 60-year-old man presented with chronic left unilateral epistaxis for at least 3 months. He had never been exposed to wood dust and did not present either diabetes or hypertension. He had recently been managed for thyroid papillary cancer by thyroidectomy and cervical lymph node dissection followed by ^{131}Iodine radioiodine therapy.

He did not report any pain, nasal obstruction, anosmia or rhinorrhoea. Clinical examination revealed a clearly demarcated, hypervascular, oval-shaped lesion, bleeding on contact, with no signs of purulent secretion, attached by a pedicle to the middle turbinate (Fig. 1). No ophthalmological or cranial nerve, especially trigeminal nerve, disorders were observed. Imaging assessment comprising contrast-enhanced CT scan and magnetic resonance imaging (MRI) was performed (Fig. 2).

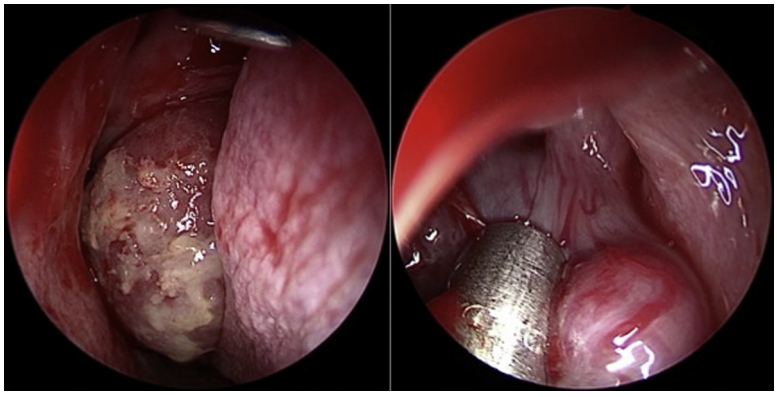

Fig. 1. Endoscopic view of the left nasal cavity at the level of the middle meatus.

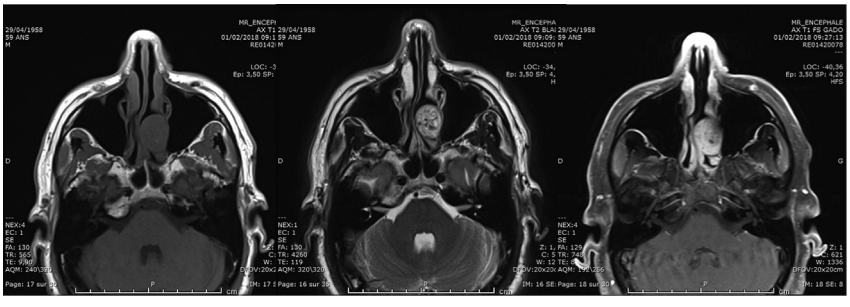

Fig. 2. MRI, axial T1-, T2- and gadolinium-enhanced T1-weighted sequences.

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* Corresponding author.
E-mail address: xdubernard@chu-reims.fr (X. Dubernard).

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2. Response

Angioleiomyoma of the left nasal cavity.

Initially described by Maesaka et al. in this site, angioleiomyoma is a rare tumour of the nasal cavity [1], representing less than 1% of all angioleiomyomas of the human body [2,3]. Angioleiomyomas arise from smooth muscle fibres and their sites of predilection are the female genital tract (95%), skin (3%) and gastrointestinal tract (1.5%) [2]. According to various hypotheses, this tumour arises in the nasal cavity from smooth muscle cells of the walls of blood vessels, arrector pili muscles of hair follicles around sweat glands or reorganized and undifferentiated embryonic tissue [2,3]. Angioleiomyomas have a nonspecific clinical presentation and essentially present in the form of repeated epistaxis and nasal obstruction. The peak incidence is observed between the fourth and sixth decades [2]. This tumour often presents as a clearly demarcated and highly vascular mass, contraindicating biopsy in the outpatient setting. It can arise anywhere in the nasal cavity, but predominantly involves the inferior turbinate, septum and nasal vestibule. In this patient, the lesion was located underneath the middle turbinate. Clinical examination did not reveal any signs of ophthalmological or cerebral involvement, as this lesion only has a potential for local growth without invasion of adjacent structures. Contrast-enhanced CT scan and magnetic resonance imaging are essential to characterize the mass and exclude other differential diagnoses.

Contrast-enhanced CT scan typically reveals a clearly demarcated, vascular lesion, isodense to muscle, not involving the sphenopalatine artery, which excludes the diagnosis of nasopharyngeal angiofibroma. The tumour is not associated with any osteolysis and, depending on its size, the nasal septum may be deviated. Gadolinium-enhanced MRI classically demonstrated a lesion with a T1 signal isointense to muscle, a hyperintense T2 signal and homogeneous and intense gadolinium enhancement [4].

The differential diagnoses that must be considered are primarily malignant tumours of the nasal cavity and paranasal sinuses, angiomyolipoma, botriomycoma, haemangioma, leiomyosarcoma, and fibromyoma.

Malignant tumours of the nasal cavity correspond to various histologies and can sometimes be related to risk factors (exposure to wood dust). These tumours are sometimes diagnosed at an advanced stage. Clinical examination may reveal neurological (sensory loss and/or facial neuralgia due to trigeminal nerve involvement) and ophthalmological lesions (diplopia, exophthalmos). In contrast with angioleiomyoma, imaging reveals poorly demarcated lesions with osteolytic invasion of adjacent structures (orbital cavity), frequently associated with intratumoural calcifications. Gadolinium enhancement is very often heterogeneous.

Angiomyolipoma, a benign tumour of adipose connective tissue, cannot be considered in this patient due to the absence of a hyperintense T1 signal on MRI.

Botriomycoma is a common, highly vascular, solid tumour, often occurring in patients after the fifth decade. Only histological examination can confirm the diagnosis, as the clinical features are very similar to those of angioleiomyoma.

Histological examination plays an essential role in this context, as it is the only way to reliably exclude all potential differential diagnoses. Primary biopsy in the operating room or primary surgical resection when the lesion is clearly demarcated is therefore mandatory. When the diagnosis of angioleiomyoma is confirmed, as in the present case, histological examination reveals a hyperplastic perivascular smooth muscle and vascular organoid proliferation. This concentric perivascular formation is composed of large spindle-shaped cells with an eosinophilic or clear cytoplasm and a low nucleocytoplasmic ratio. Immunohistochemical analysis of this lesion shows the presence of a mature smooth muscle phenotype with expression of positive markers such as h-caldesmon and negative markers such as HMB45 [5]. The presence of less than 3% of mature adipose tissue excluded the diagnosis of angiomyolipoma. The absence of nuclear atypia, cigar-shaped tumour nuclei arranged in intertwined fascicles, and the absence of numerous mitotic figures also excluded the diagnosis of leiomyosarcoma in this patient [3].

Angioleiomyoma has an excellent prognosis following complete resection. Treatment is exclusively surgical and is associated with a very low recurrence rate. We recommend clinical follow-up at 6 months and 1 year, combined with radiological follow-up to detect possible recurrence.

Tumours of the nasal cavities present a wide range of benign and malignant histology and must always be characterized histologically and radiologically. Angioleiomyoma is an unusual benign tumour, which must be considered in the differential diagnosis, even in these rare sites.

Disclosure of interest

The authors declare that they have no competing interest.

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