Extranasopharyngeal angiofibroma of the nasal septum - uncommon presentation of a rare disease

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INTRODUCTION

The extranasopharyngeal angiofibroma (ENPA) is a tumor which is histologically similar to juvenile nasopharyngeal angiofibroma (JNA), differing from the latter in clinical and epidemiological characteristics.1-3 Prevalence, gender, age, affected site, pathogenesis, clinical-CT and recurrence are completely different, which leads some authors to classify the ENPA as a disease which is different from the JNA.1

There are less than a hundred cases of ENPA described in the literature, and the maxillary sinus is the most frequently affected site, followed by the ethmoid, being rare in the nasal septum.4 The objective of this study is to report a case of ENPA with a rare presentation in the nasal septum.

CASE PRESENTATION

WSR, 10 years and 11 months of age, complained of constant bilateral nasal obstruction for six months, worse on the right side, with hyposmia and snoring, without epistaxis. Nasal endoscopy showed a pinkish lesion, smooth, non-friable, non-ulcerated, apparently inserted into the nasal septum, obstructing the right nasal cavity (RNC) in its posterior third all the way to the choanae, and it could be viewed through the other nostril. Middle meatuses and sphenoid recesses were free. Computed tomography (CT) showed a lesion with soft tissue density in the RNC (Figure 1A-B). We performed an endoscopic approach, identifying the lesion inserted in the nasal septum, doing a subperiosteal dissection and excision with a margin at its insertion. Postoperative follow-up of 1 year and 9 months without recurrence (Figure 1C). Histopathology reported it to be an angiofibroma (Figure 1D).

DISCUSSION

The JNA is the most common benign neoplasm of the nasopharynx, despite representing less than 0.05% of tumors of the head and neck.2-4,5 It affects almost exclusively males, between 12 and 14 years of age.2-4,5 But the ENPA is even more unusual, more common in women between 17 and 22 years, and its most common site is the maxillary sinus, followed by the ethmoid, being very rare in the nasal septum and inferior turbinates.1 The origin of the JNA is at the top of the sphenopalatine foramen, with controversial etiology. ENPA’s etiology is associated with a migration error of the fascia basalis, justifying its presence in varied locations. Our patient had age, gender and location different from most ENPAs, confirming the rarity of this case.

The initial growth of the JNA follows a well-defined pattern in the nasal cavity, nasopharynx and pterygopalatine fossa, leading to the triad: nasal obstruction, recurrent epistaxis and nasopharyngeal tumor.1,2 The JNA has characteristic radiological signs: Holman-Miller (anteriorization of the posterior wall of the maxillary sinus) and enlargement of the sphenopalatine foramen - pterygopalatine fossa.2-4 The ENPA can evolve with a variety of symptoms and radiological signs, depending on its site.2-4 Our patient reported nasal obstruction due to a rare location in the nasal septum.

Histologically, the ENPA is similar to the JNA, with connective tissue stroma and a matrix of dilated vessels without a muscular layer.2-4 As for differential diagnosis, we have the hemangioma and the hemangiopericytoma. While the JNA can be suspected based on known clinical and CT characteristics2-4,5, histopathological examination is essential to confirm the ENPA diagnosis.

REFERENCES