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Case Report

Bilateral Frontal Sinus Ectopic Angiofibroma: Focus on Management of a Rare Lesion with Unusual Presentation

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Abstract

Extranasopharyngeal Angiofibromas (ENA) are rare benign tumors originating in the majority of cases from the nasal septum, while involvement of the frontal sinus is extremely rare, with few cases reported in literature. ENA present the same histological characteristics and immunophenotype of Nasopharyngeal Angiofibromas (NA), which are more commonly found; however, they do not present the typical clinical presentation of their nasopharyngeal counterpart, therefore biopsy is mandatory to obtain the diagnosis. A thorough preoperative assessment by means of nasal endoscopy, computed tomography scan and enhanced magnetic resonance imaging provides essential information for treatment planning. The authors present the case of a 34-year-old man with a massive fronto-ethmoidal angiofibroma. A combined endoscopic transnasal and osteoplastic flap approach was employed to resect the tumor. After 2 years of clinical and radiological follow-up, no recurrence of disease was observed. Despite being rare entities, ENA must be considered in presence of frontal sinus neoformations. A combined endoscopic endonasal and osteoplastic flap approach provides a safe and effective treatment for these lesions when a massive involvement of the frontal sinus is present, overcoming the limits of single-approach procedures.

Keywords: Sinonasal angiofibromas; Frontal sinus; Endoscopic; Osteoplastic flap; Surgery.

Introduction

Extranasopharyngeal Angiofibromas (ENA) involving paranasal sinuses represent an unusual finding and the localization at the level of frontal sinus is an extremely rare event with few cases reported in literature [1,2]. The clinical presentation is aspecific, lacking the typical features of their more common nasopharyngeal counterpart with which they share the histological characteristics.

We present our experience in the diagnosis and management of a massive frontoethmoidal angiofibroma, treated with a combined endoscopic transnasal and Osteoplastic Flap (OPF) approach.

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Case presentation

A 34-year-old man was referred to our department for a nine-month history of bilateral nasal respiratory obstruction and right epistaxis.

The endoscopic endonasal evaluation revealed a pinkish, firm, capsulated mass of apparent vascular nature, completely obstructing the right nasal fossa. A CT scan, as first line radiological investigation, revealed an expansive mass occupying the right nasal fossa, with opacification of the ipsilateral antrum and both frontal sinuses, thinning of bony septa of the anterior ethmoidal complex as well as remodeling of interfrontal septum and frontal beak. An MRI with gadolinium showed an inhomogeneous neoformation both on T2-weighted and enhanced T1-weighted sequences, involving the right ethmoid and bilateral frontal sinuses (Figure 1). A biopsy of the mass was performed under local anesthesia. Histopathological examination revealed fibrocollagenous stromal proliferation with an admixture of variably sized and angulated vascular spaces.

Immunohistochemistry showed that endothelial cells were reactive with endothelial cell markers (e.g., CD31, CD34, others). Smooth muscle actin-positive cells were found around the circumference of the vascular spaces. Spindle-shaped and stellate stromal cells were positive for nuclear androgen receptor, nuclear β-catenin and vimentin, whilst negative for S100 protein, estrogen and progesterone receptor (Figure 2). The histology and immunohistochemistry were compatible with juvenile angiofibroma. The patient was then scheduled for a combined

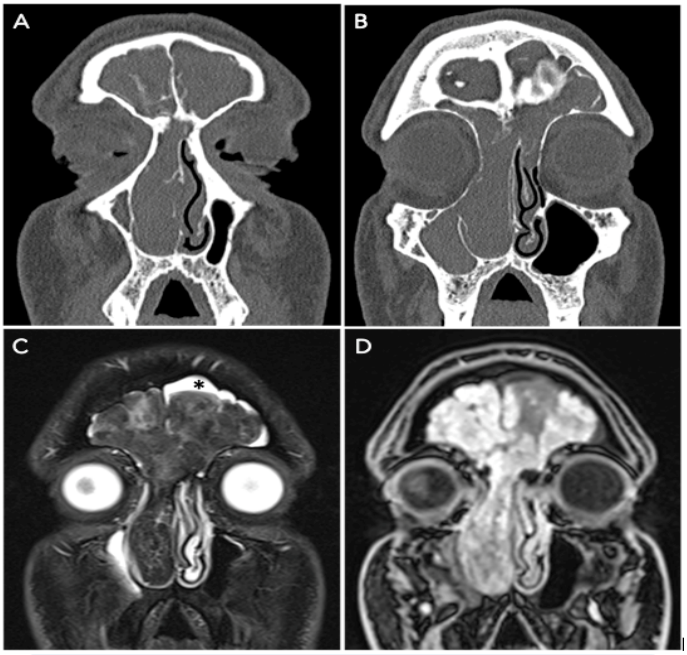

Figure 1: Preoperative imaging investigations showing a neoformation involving the right ethmoid sinus, bilateral frontal sinuses and completely occupying the right nasal fossa. CT scan shows thinning of bony septa of right anterior ethmoidal complex as well as remodeling of interfrontal septum and frontal beak **(A-B)**. At MRI, the lesion appears hypointense on coronal T2-weighted **(C)**, retained secretions lie in the superior portion of left frontal sinus (asterisk). Post contrast spoiled 3D gradient echo T1-weighted fat saturated sequences show diffuse inhomogeneous enhancement **(D)**, the mass is characterized by a "salt and pepper" appearance due to the presence of intralesional flow-voids **(C)**.

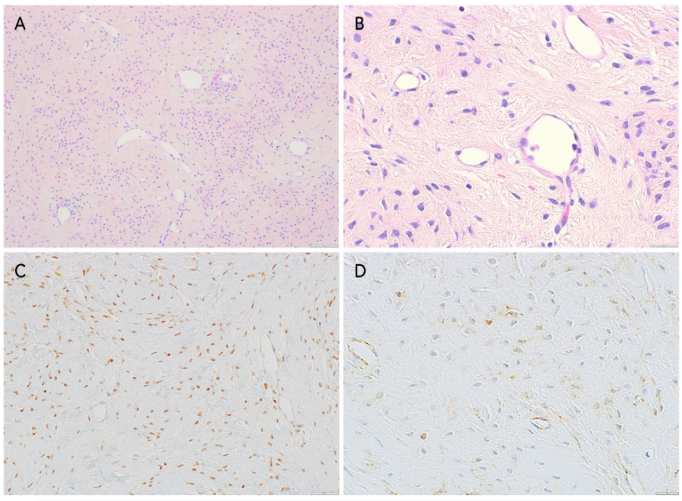

Figure 2: (A) - Microscopic examination showing an admixture of fibrocollagenous stroma, and thin-walled blood vessels (H&E 100x). **(B)** - Spindle-shaped and stellate cells with plump nuclei are visible. Angulated blood vessels lined by a single layer of endothelial cells are detectable inside the stroma (H&E 400x). **(C)** – Cells with nuclear immunoreactivity for androgen receptor. **(D)** - Cells with nuclear immunoreactivity for β-catenin.

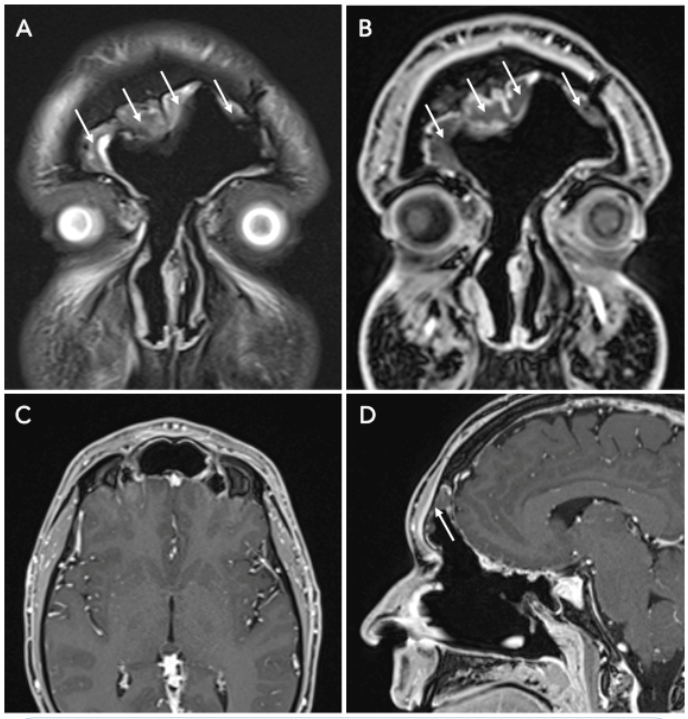

Figure 3: Postoperative magnetic resonance imaging performed 2 years after resection showing good surgical results without evidence of recurrence of angiofibroma on coronal T2-weighted sequence **(A)** and coronal **(B)**, axial **(C)**, sagittal **(D)** post contrast spoiled 3D gradient echo T1-weighted fat saturated sequences. A signal with inhomogeneous intensity on T2-weighted sequence and peripheral contrast enhancement, compatible with scar tissue, lies on upper/lateral portions of frontal sinuses (white arrows).

endoscopic endonasal and OPF approach. The portion of the lesion obstructing the right nasal cavity and involving the anterior ethmoid was removed trans-nasally. The lesion appeared to originate from the frontal beak, which was accurately drilled out after a right frontal sinusotomy type IIB sec. Draf was performed. An ethmoidectomy, middle antrostomy and sphenoidotomy were performed without evidence of involvement by the lesion. The OPF approach allowed complete removal of the lesion occupying both frontal sinuses, and the bony walls' surface was drilled out, in order to eliminate possible microscopic remnants. An MRI performed 48 hours after surgery excluded any persistence or postoperative sequelae. Postoperative course was uneventful, and the patient was discharged 72 hours after surgery. The follow-up was performed with nasal endoscopy at 1, 3, 6, 12 months for the first year after surgery and then two times per year, combined with MRI performed every 6 months; 2 years after surgery no recurrence was observed (Figure 3).

Discussion

ENA is a rare entity, with less than 200 cases reported in medical literature. Although the histologic features are the same of much more common Juvenile Angiofibroma (JA), the clinical appearance differs significantly; therefore, they are considered as two different entities by many authors. In contrast with JA, which has a characteristic localization, due to its precise site of origin at the level of pterygoid, ENA could involve any site of the upper aerodigestive tract, but nasal septum appeared to be involved in the majority of cases. Windfur et al. in their large literature revision of 174 cases of ENA did not report any case of frontal sinus involvement, which appears to be an unusual site of presentation for this pathology with few cases reported in current literature [3]. The age of presentation is typically around the second and third decade of life. In contrast with JA, the occurrence in females is not unprecedented (M:F 2:1). The main symptom is nasal obstruction, either in combination with epistaxis (25.8%) or other symptoms (12.6%). Diagnosis of JA is confirmed by imaging due to the characteristic radiological features; this is not possible when dealing with ENA. Considering the aspecific presentation of ENA, the clinical diagnosis of this pathologic entity is not straight forward, and differential diagnosis includes the majority of sinonasal vascular benign tumors and low-grade malignant tumors. In presence of lesions compatible with JA, biopsy is strongly contraindicated to avoid bleeding which could be difficult to deal with; fortunately, the reduced vascular component of ENA compared to JA, makes biopsy a feasible procedure with a low risk of massive bleeding. Histologic findings and immunohistochemistry play an essential role in differentiating angiofibromas from other vascular malformations or tumors (e.g. lobular capillary hemangioma, sinonasal polyps, peripheral nerve sheath tumors, solitary fibrous tumor and desmoid tumor) [4,5]. Complete surgical resection is considered the best treatment option. The reduced vascularization of ENA compared to JA accounts for limited intraoperative bleeding, making preoperative embolization of the mass unnecessary in the majority of cases. Nowadays, the majority of sinonasal benign and malignant tumors may be treated with an exclusive endoscopic approach, with recurrence and complication rates similar to that reported for external approaches. In the present case, the choice of a combined endoscopic endonasal and external approach via an OPF, was based on the massive involvement of well pneumatized frontal sinuses by the lesion. Kim et al. reported a case of frontal sinus ENA treated with transnasal approach and frontal sinus trephination which did not allow a complete excision of the lesion [6]; to gain a complete exposition of the sinus and easily reach radicality, we chose to perform an OPF approach.

Conclusion

The localization of angiofibromas at the level of frontal sinus is extremely rare, however they must be taken into account in differential diagnosis of frontal sinus neoformations. A biopsy is necessary for the diagnosis and it's a safe procedure, with massive bleeding being a rare occurrence.

A combined endoscopic endonasal and osteoplastic flap approach provides a safe and effective treatment of these lesion when a massive involvement of the frontal sinus is present, overcoming the limits of the single-approach procedures.

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