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Case Report

External Auditory Canal Angiofibroma: A Case Report And Literature Review.

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Abstract

Angiofibromas are rare benign fibrovascular tumors typically found in the nasopharynx. Extra-nasopharyngeal angiofibromas are even more uncommon, with limited reports of their occurrence in the external auditory canal (EAC). We present a case of an angiofibroma located in the left EAC in a 33-year-old woman who presented with the gradual onset of multiple nodules on the pinna and EAC, along with left-sided otalgia for 6 months. Clinical examination revealed angiomatous papulo-nodules, the largest measuring 1 cm. Histopathological analysis confirmed angiofibroma. Preoperative audiometry showed moderate conductive hearing loss in the left ear. A CT scan revealed polypoid mucosal thickening in both the bony and cartilaginous EAC without bone erosion. Given the clinical and radiological findings, the patient first underwent an incomplete excision of the lesions on the pinna. Subsequently, she received external radiation therapy using intensity-modulated radiation therapy (IMRT), receiving a total dose of 50 Gray in 25 fractions. The patient experienced grade 1 radiodermatitis, which resolved without complications. Follow-up CT at 6 months showed regression of the polyps and post-therapy changes in the pinna. The patient remains stable with no further progression.

Keywords : Angiofibroma, External Auditory Canal, Radiation therapy, Local control.

INTRODUCTION

Angiofibromas account for less than 1% of all head and neck tumors, primarily occurring in the nasopharynx and predominantly affecting adolescent males, hence termed juvenile nasopharyngeal angiofibromas (NPA) [1-3]. In contrast, extranasopharyngeal angiofibromas (ENPA) are much rarer, with fewer than 80 cases reported in the literature [1, 2]. Among these atypical locations, angiofibromas of the external auditory canal (EAC) are extremely rare, with only a few documented cases [1.2.4]. This extreme rarity presents significant diagnostic and therapeutic challenges.

In this article, we present the case of a female patient with an angiofibroma of the external auditory canal, highlighting treatment strategies, particularly the use of radiation therapy, in the management of this rare tumor.

CASE PRESENTATION

Our patient is a 33-year-old woman with no prior medical history, admitted for the management of an angiofibroma of the left ear. Approximately two years ago, she noticed the gradual appearance of multiple nodules on the pinna and external auditory canal (EAC) of the left ear, associated with left-sided otalgia, which led her to seek consultation. She was initially seen at the dermatology department of University Hospital Hassan II in Fez, where a biopsy confirmed the diagnosis of angiofibroma. She underwent an excision of the nodules, but the excision was incomplete and she was subsequently referred to our center for adjuvant radiation therapy.

On clinical examination in dermatology, angiomatous papulo-nodules and skin-colored nodules were observed, the largest measuring 0.5 cm, located on the concha and EAC of the left ear, with a soft consistency. Histopathological examination of the biopsy revealed features of an angiofibroma, with no eosinophilic infiltrate, ruling out a diagnosis of angiolymphoid hyperplasia with eosinophilia (ALHE). Histopathological examination of the excised lesions from the pinna showed an angiofibroma in two fragments and angiolymphoid

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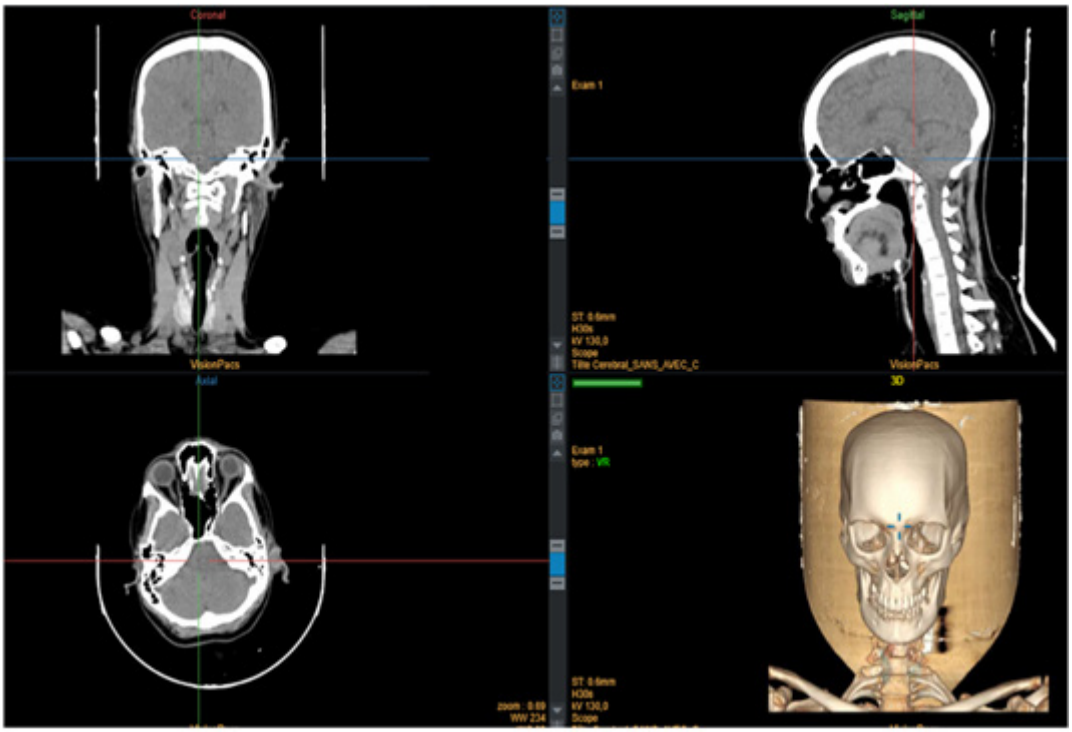
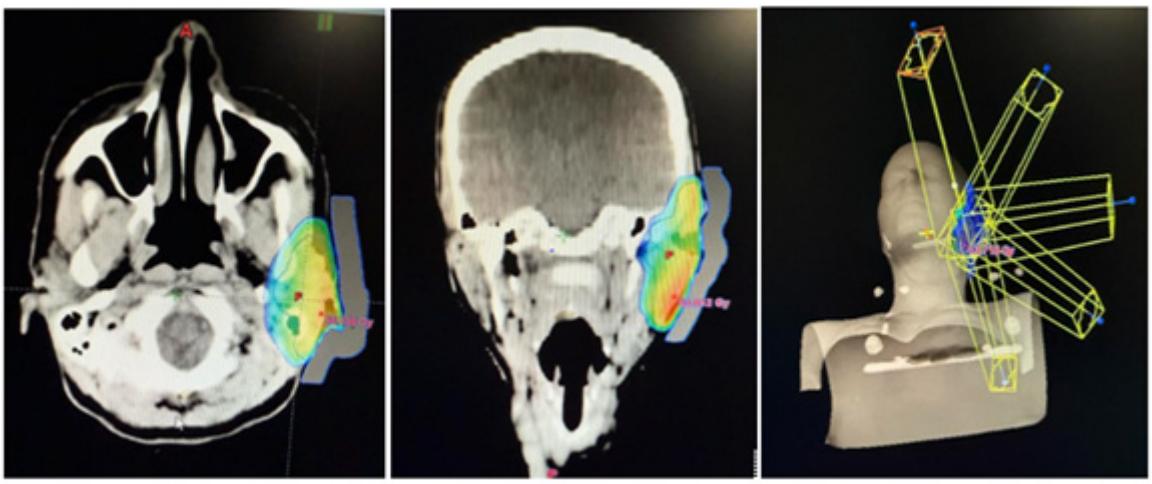
hyperplasia with eosinophilia in one fragment. A computed tomography (CT) scan of the temporal bones revealed polypoid mucosal thickening in the anterior part of the bony EAC, with no bone erosion, measuring 6x5 mm, remaining distant from the tympanic membrane. On the left side, polypoid mucosal thickening of the cartilaginous EAC was noted, without deep extension, measuring 9 mm at its thickest point, with no bone erosion (**Figure.1**). Audiometry showed moderate conductive hearing loss in the left ear, while the right ear was normal.

Figure 1. CT scan showing Polypoid Mucosal Thickening in the Left Cartilaginous External Auditory Canal Without Bone Erosion.



On clinical examination, soft, angiomatous nodules were found on the pinna and external auditory canal (EAC) of the left ear, with the largest measuring 1 cm. The cervical and supraclavicular lymph nodes were non-enlarged. The patient was scheduled for external radiation therapy using the intensity-modulated radiation therapy (IMRT) technique, with a total dose of 50 Gray administered in 25 fractions of 2 Gray each, delivered once daily, five days a week (**Figure.2**)

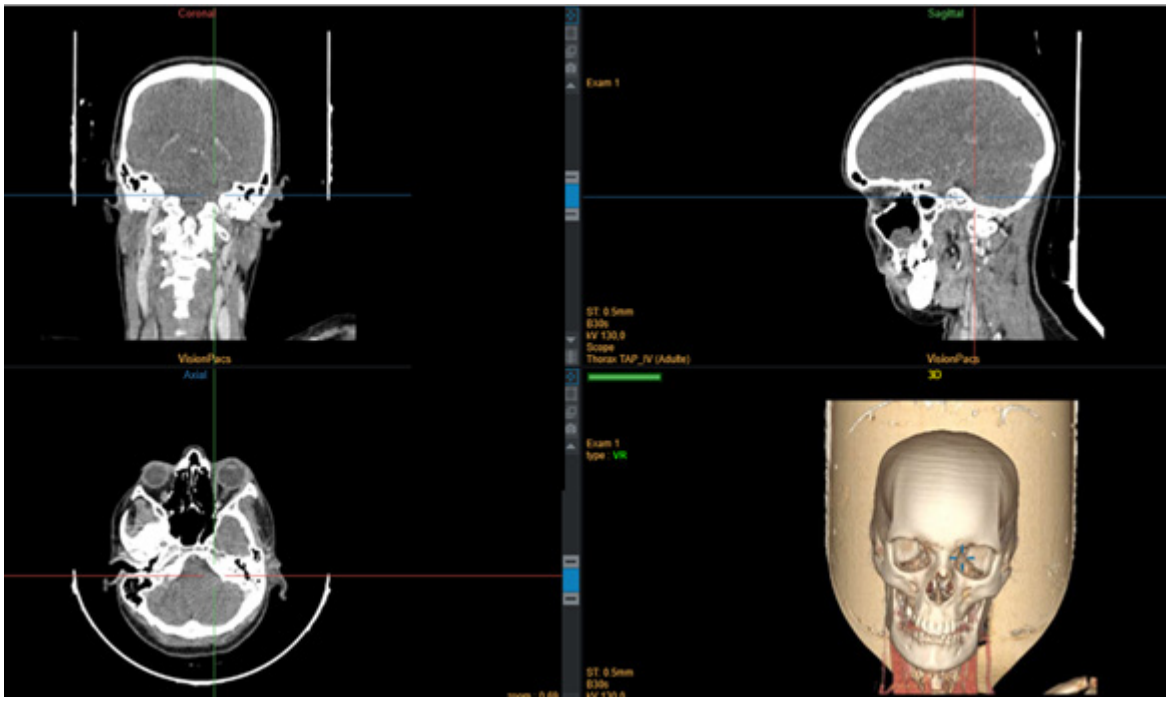
Figure 2. Radiation Dose Distribution to Target Volumes.



The acute toxicity of radiation therapy was marked by the appearance of grade 1 radiodermatitis, which resolved well.
A follow-up CT scan at six months showed stability of a 6 mm hypervascular polyp in the right EAC, with regression of three

polyps in the left EAC. Post-therapeutic changes were noted in the pinna (**Figure.3**). The treatment was completed, and the patient remains under regular follow-up with no further progression.

Figure 3. Six-Month Follow-Up CT Scan Showing Regression of Three Polyps in the Left EAC, and Post-Therapeutic Changes in the Pinna.



DISCUSSION

Angiofibromas are benign fibrovascular tumors characterized by irregular, thin-walled blood vessels embedded in a fibrous or cartilaginous stroma containing numerous spindle-shaped or stellate cells [2]. These tumors are generally categorized into two distinct types: juvenile nasopharyngeal angiofibromas (PNA) and extra-nasopharyngeal angiofibromas (ENPA). The primary distinction between these two types lies in their anatomical locations. While ENPAs have been occasionally reported in areas such as the maxillary sinus, ethmoid sinus, nasal septum, and nasal cavity, cases involving the external auditory canal (EAC) are extremely rare [1–3].

Although PNA and ENPA share histological features, they exhibit distinct clinical characteristics, supporting their classification as separate entities [1–3].

PNA is typically diagnosed without difficulty, thanks to its characteristic clinical presentation, which includes a predominance in adolescent males, along with specific imaging features [2]. Conversely, ENPA can be challenging to diagnose due to the often subtle and variable clinical signs, which are heavily influenced by the tumor's location [1–3]. This was evident in the case of our patient, where the clinical presentation of nodules in the pinna and external auditory canal, along with left-sided otalgia, initially led to a delayed diagnosis. Timely recognition of ENPA is critical to ensure appropriate management and avoid unnecessary delays in treatment.

Imaging modalities, such as computed tomography (CT) and magnetic resonance imaging (MRI), are indispensable for evaluating the tumor's location, size, extent, and its relationship with adjacent structures. In some cases, carotid or selective angiography may be used to visualize the vascular supply of the tumor and assist in preoperative embolization, which helps minimize the risk of bleeding during surgery [2]. For ENPA, preoperative biopsy is generally not recommended due to the elevated risk of hemorrhage; this is a concern that is more pronounced in PNA. Additionally, ENPA typically follows a less aggressive clinical course, leading to a favorable prognosis and a lower recurrence rate after surgical resection [1–3].

The differential diagnosis for angiofibromas includes conditions such as angiofibrolipomas, solitary fibrous tumors (SFTs), vascular tumors, infected polyps, myxomas, fibromyxoid sarcomas, and liposarcomas [1–3]. Angiofibrolipomas, which consist of a mix of vascular, adipose, and collagenous tissue, are histologically distinct from angiofibromas and are usually less aggressive due to their lower cellularity. These tumors are also not encapsulated, making them easier to differentiate from angiofibromas [5]. Other lesions, such as angiolipoleiomyomas, which are commonly found in the earlobe, should also be considered in the differential

diagnosis. The distinction between these tumors relies on specific histological and immunohistochemical markers, such as Masson's trichrome staining for collagen or the presence or absence of smooth muscle actin [6].

SFTs, which are of mesenchymal origin, may also present similarly to angiofibromas. These tumors are composed of spindle-shaped cells with collagen-rich stroma and branching vessels, and may include multinucleated giant cells, a feature historically associated with giant cell angiofibromas (GCA). Though rare, SFTs in the external auditory canal have been documented in a small number of cases [7–11].

Radiation therapy is occasionally used in the management of angiofibromas, especially for unresectable tumors or those with intracranial extension, as well as for cases of incomplete tumor removal. Despite the rare occurrence of malignant transformation following radiation therapy, the local control rate of 80%-85% in terms of symptom regression has been reported, with outcomes comparable to surgical interventions [12-15]. A total irradiation dose of 30-35 Gy is generally recommended, although doses of 40-45 Gy may be used for larger or more extensive lesions [13.16.17].

While radiation therapy was rarely used for ENPA, its use has been documented in a few cases between 1961 and 1979. For example, Alajmo and Storchii in 1961 applied radium treatment (10 mg for 72 hours) after partial resection of an ethmoid lesion, while another patient received similar treatment following a lateral rhinotomy [18]. Other reports, such as those by Gudea and Kitano, describe the successful use of 40 Gy and 20 Gy of radiation, respectively, for managing large lesions in the maxillary region [13]. However, radiation therapy is not always effective and may require repeated surgical interventions for recurrences, as seen in the case of Supiyaphun, who administered radiation to shrink a large ENPA in the oral cavity but found the therapy ineffective [19].

In addition to radiation therapy, other treatment modalities for angiofibromas include cryotherapy, embolization, hormone therapy, chemotherapy, arterial ligation, and sclerosing agents. These options can be used either alone or in combination, depending on the tumor's location, size, and aggressiveness. For cases with intracranial extension, a multidisciplinary approach may be necessary. Chemotherapy, specifically with doxorubicin or dacarbazine, has been suggested for aggressive or recurrent angiofibromas, though it is not indicated for ENPA, as evidenced by a case where pre-surgical chemotherapy for an ENPA was ineffective [20-23].

CONCLUSION

Angiofibromas, though benign, can be challenging to diagnose and treat due to their varied clinical presentations and locations. While surgical resection is the primary treatment with good outcomes, radiation therapy can serve as an alternative for inoperable or extensive tumors. Although rarely used for extra-nasopharyngeal angiofibromas (ENPA), radiation therapy has shown efficacy in some cases. Other treatments like cryotherapy and chemotherapy may also be considered for aggressive or recurrent cases. A multidisciplinary approach is often beneficial, and further research is needed to optimize treatment strategies for these rare tumors.

Data availability

The patient's data are available upon reasonable request to the corresponding author.

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All the authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

Conflict of interest statement

None declared.

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REFERENCES

1. Windfuhr JP, Remmert S. Extranasopharyngeal angiofibroma: etiology, incidence and management. Acta Otolaryngol 2004;124: 880–9.

2. Szymanska A, Szymanski M, Morshed K, Czekajska-Chehab E, ´ Szczer- bo-Trojanowska M. Extranasopharyngeal angiofibroma: clinical and radiological presentation. Eur Arch Otorhinolaryngol 2013;270:655–60.

3. Correia FG, Simões JC, Mendes-Neto JA, Seixas-Alves MT, Gregório LC, Kosugi EM. Extranasopharyngeal angiofibroma of the nasal septum uncommon presentation of a rare disease. Braz J Otorhinolaryngol 2013;79:646.

4. Bouhafs K, Lachkar A, Bouamama T, Miry A, Benfadil D, Ghailan MR. Angiofibroma of the external auditory canal: a rare case with literature review. J Surg Case Rep. 2022 Apr 3;2022(4):rjac117. doi: 10.1093/jscr/rjac117. PMID: 35382137; PMCID: PMC8977119.

5. Liggett AD, Frazier KS, Styer EL. Angiolipomatous tumors in dogs and a cat. Vet Pathol 2002;39:286–9

6. Kershisnik W, McCarthy DJ, O'Donnell E. Angiofibrolipoma: a histologic variant of lipoma. J Am Podiatr Med Assoc 1986;76: 67–70.

7. Guillou L, Gebhard S, Coindre JM. Orbital and extraorbital giant cell angiofibroma: A giant cell-rich variant of solitary fibrous tumor? Clinicopathologic and immunohistochemical analysis of a series in favor of a unifying concept. Am J Surg Pathol 2000;24: 971–9.

8. Dei Tos AP, Seregard S, Calonje E, Chan JK, Fletcher CD. Giant cell angiofibroma. A distinctive orbital tumor in adults. Am J Surg Pathol 1995;19:1286–93.

9. Thomas R, Banerjee SS, Eyden BP, Shanks JH, Bisset DL, Hunt R, et al. A study of four cases of extra-orbital giant cell angiofibroma with documentation of some unusual features. Histopathology 2001;39:390–6.

10. Rezk S, Yousef M, Zamansky M, Khan A. Solitary fibrous tumor of the auditory canal. Arch Pathol Lab Med 2004;128:e169–71.

11. Izumaru S, Yoshida Y, Nakashima T. A solitary fibrous tumor in the external auditory meatus. Auris Nasus Larynx 2004;31:65–7.

12. Makek MS, Andrews JC, Fisch U. Malignant transformation of a nasopharyngeal angiofibroma. Laryngoscope. 1989;99:1088-1092.

13. Gudea F, Vega M, Canals E, Montserrat JM, Valdano J. Role of radiation therapy for 'juvenile' angiofibroma. J Laryngol Otol. 1990;104:725-726.

14. Sinha PP, Aziz HI. Juvenile nasopharyngeal angiofibroma. A report of seven cases. Radiology. 1978;127:501-503.

15. De Blanc HJ, Chretien P. Angiofibroma clinically indistinguishable from carotid body tumor. Am J Surg. 1970;119:743-745.

16. Reddy KA, Mendenhall WM, Amdur RJ, Stringer SP, Cassisi NJ. Long-term results of radiation therapy for juvenile nasopharyngeal angiofibroma. Am J Otolaryngol. 2001;22:172-175.

17. Fitzpatrick PJ, Briant TD, Berman JM. The nasopharyngeal angiofibroma. Arch Otolaryngol. 1980;106:234

18. Alajmo E, Fini-Storchi O. Il fibroangioma primitivo delle fosse nasali e dei seni paranasali. Boll Mal Orecch. 1961;79:217-240.

19. Supiyaphun P, Sastarasadhit V, Chochaipanichnon L, Tonsakulrungruang K. Intraoral angiofibroma: a case report. J Med Assoc Thai. 1986;69:382-385

20. Goepfert H, Cangir A, Lee YY. Chemotherapy for aggressive juvenile nasopharyngeal angiofibroma. Arch Otolaryngol. 1985;111:285- 289.

21. Schick B, Kahle G, Hassler R, Draf W. Chemotherapy of juvenile angiofibroma–an alternative? HNO. 1996;44:148-152.

22. Gupta M, Motwani G, Gupta P. Extranasopharyngeal angiofibroma arising from the infratemporal region. Indian J Otolaryngol Head Neck Surg. 2006;58:312-315.

23. Goepfert H, Cangir A, Ayala AG, Eftekhari F. Chemotherapy of locally aggressive head and neck tumors in the pediatric age group. Desmoid fibromatosis and nasopharyngeal angiofibroma. Am J Surg. 1982;144:437-444.