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Spindle cell hemangioma in the infratemporal fossa: A unique case report

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ABSTRACT

INTRODUCTION: Spindle cell hemangioma (SCH) is a rare vascular tumor which was first described in 1986. It affects mostly the distal extremities. The head and neck are rarely involved. This article reports the first case of SCH in the infratemporal fossa.
PRESENTATION OF CASE: A 41-year-old woman presented with an 8-month history of right cheek swelling. Facial CT scan and MRI showed an intensely and heterogeneously enhancing tumor of the infratemporal fossa suggesting an angiomatous neoplasm. The mass was excised surgically through an anterior maxillary approach. The histopathological and immunohistochemistry analysis revealed a SCH.
CONCLUSION: This case report presents a unique presentation of a Spindle cell hemangioma in an unexpected location of the head and neck region. it underlines the importance for clinicians and pathologists to consider the Spindle cell hemangioma as a possible etiological diagnosis of infratemporal fossa tumors.

1. Introduction

Spindle cell hemangioma (SCH) is a rare vascular tumor which was first described in 1986 as a low grade angiosarcoma [1].

By the advent of immunochemistry, this tumor was no longer considered as malign [2].

Because of its indolent clinical course, it is usually present for many years before diagnosis.

Habitually, the SCH affects almost exclusively the dermis and subcutaneous tissues of distal extremities. Till present, only 16 cases involving the head and neck areas have been reported. Most of them presenting as submucosal nodules although, intra-muscular and intra-orbital presentations have been reported [3]. As far as the authors know, cases located in the infratemporal fossa have never been described.

2. Presentation of case

A 41-year-old woman, with no medical history, presented with an 8-month history of a slowly growing painless right cheek swelling, with no other associated signs. Extraoral examination noted a facial asymmetry due to the presence of a bulging right cheek with no inflammatory signs. The swelling was solid, poorly circumscribed, no pulsatile, fixed to the deep planes. Facial nerve function was retained and no cervical lymphadenopathy was found.

In the intraoral examination, the mass was at the level of the right intermaxillary region with a regular overlying mucosa, without trismus.

No skin abnormalities, vascular and lymphatic malformations or associated tumors were found on the clinical examination.

Enhanced computed tomography (CT scan) of the face revealed a tissue density voluminous mass of the right Infratemporal Fossa, intensely and heterogeneously enhancing (Fig. 1). The mass expands the posterior wall of the maxillary sinus anteriorly without bone destruction. Superiorly, it extends to the inferior orbital fissure without orbital invasion and medially to the sphenopalatine foramen.

On magnetic resonance imaging (MRI) of the face, T1-weighted image showed heterogeneous isointense signal relative to adjacent muscle with considerable enhancement after gadolinium administration. On diffusion-weighted images, the tumor was of high intensity with low ADC value (Fig. 2).

Neither biopsy nor embolization were performed prior to surgery.

The surgical excision was performed by a senior ENT professor under general anesthesia through a large anterior maxillotomy approach with resection of the posterior wall of the maxillary sinus (Fig. 3). A monobloc resection of the tumor was achieved after bipolar cauterization and ligation of inflow feeder vessels (Fig. 4). The cavity was checked endoscopically for any residual tumoral tissues and hemostasis was ensured. The postoperative course was uneventful. On gross examination, the tumor had a reddish-white surface covered with a thin capsule, its consistency was firm. The weight and size were respectively 50 g and 8 × 6 ×

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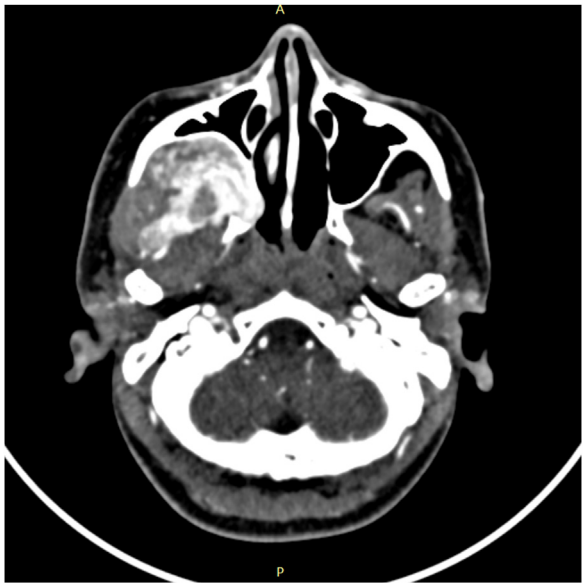

Fig. 1. Computed tomography (CT scan) showing a tissue density voluminous and intensely enhanced mass of the right Infratemporal Fossa.

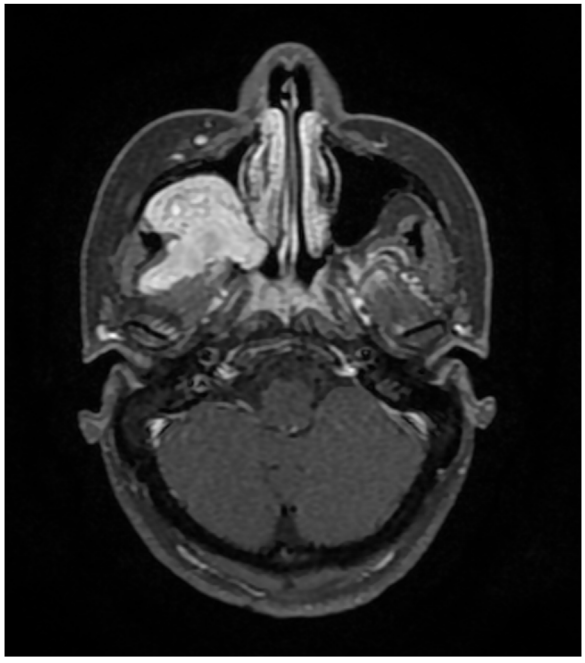

Fig. 2. MRI of the face: T1-weighted image showing heterogeneous and considerable enhancement of the tumor after gadolinium administration.

3 cm. Microscopic examination and immunohistochemical study, showed highly vascularized proliferation resembling Kaposi sarcoma associated to solid areas composed of spindle shaped cells and some endothelial cells which was positive for CD31 and CD34. Immunostaining for Desmine, Cytokeratin, S-100 protein and SMA were negative. Cellular atypia and abnormal mitotic figures were not observed (Figs. 5–7). These findings led to the diagnosis of SCH. The postoperative course was uneventful and the patient was discharged home 48 h later. After a multidisciplinary team meeting, and given the benign nature of the neoplasm, the total excision and the risks of side effects and malignant transformation, the decision was to not add adjuvant radiation or chemotherapy. At 12 months of follow up, no signs of recurrence have been noted.

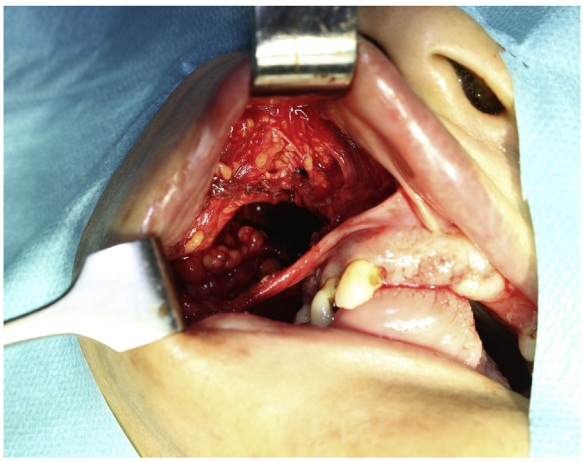

Fig. 3. Peroperative view of the transmaxillary approach.

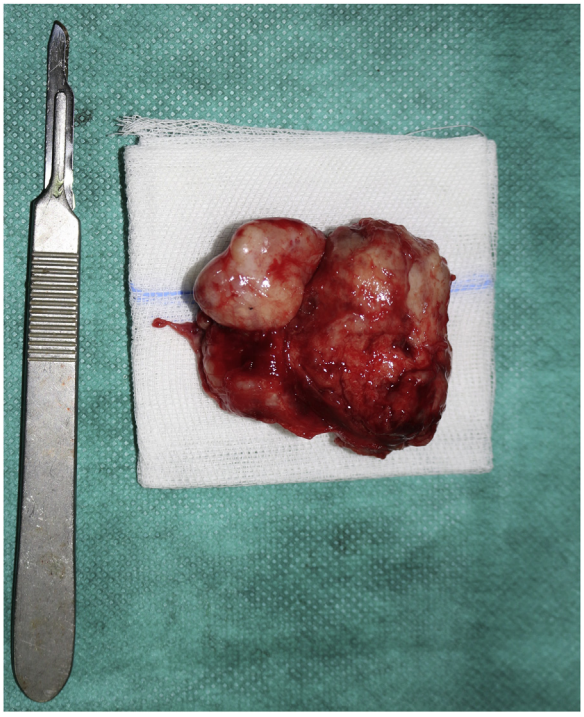

Fig. 4. Complete en-bloc excision of the tumor.

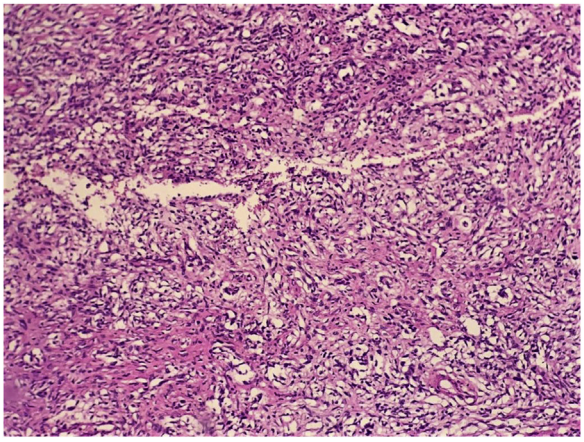

Fig. 5. Microscopic findings of the spindle cell hemangioma with no evidence of abnormal mitotic activity or nuclear atypia in spindled cells (hematoxylin and eosin stain, original magnification ×100).

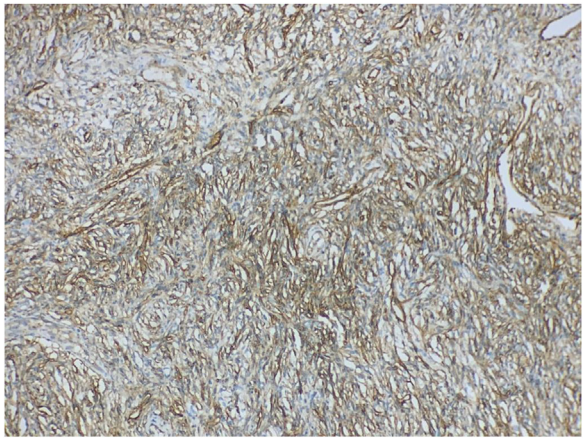

Fig. 6. Photomicrograph showing positivity for CD31 immunohistochemical marker.

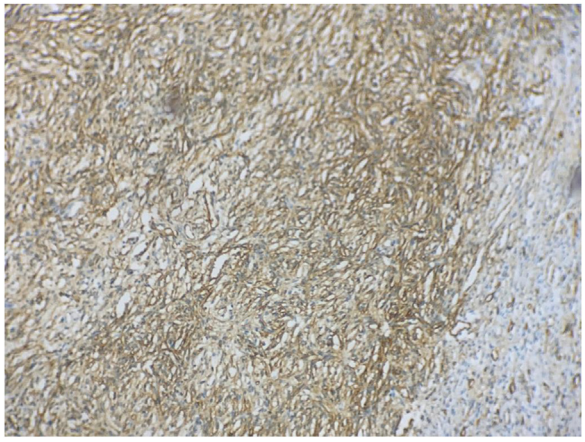

Fig. 7. Photomicrograph showing positivity for CD34 immunohistochemical marker.
No patient or author details are included in the figures.

This case has been reported in line with the SCARE 2018 criteria [4].

3. Discussion

In 1986, Weiss and Enzinger described the SCH as a unique vascular tumor with combined features of cavernous hemangioma and Kaposi's sarcoma. The tumor was considered to be intermediate -or low-grade malignancy, with biological behavior between a hemangioma and angiosarcoma [1].

But in 1991, Fletcher et al. suggested, on the basis of a clinicopathological and immunohistochemical study, that SCH were more likely to be a non-neoplastic lesion and that its development correlates with histological evidence of a malformed vascular at the affected site [2].

In a large case series of 78 patients, Perkins and Weiss, noted no evidence of metastatic potential and described the SCH as a benign vascular neoplasm based on the resemblance of SCH with angiomatosis. They designated the terms SCH for solitary lesions and "spindle cell hemangiomatosis" for multifocal lesions [5].

The latters have been associated with Maffucci syndrome, Ollier disease, Millroy disease, Klippel-Trenaunay syndrome, von Willenbrand disease and acute myelomonocytic leukemia [6].

Clinically, this neoplasm is characterized by an indolent but progressive growth, local recurrence and multifocality [7]. It occurs at all ages with equal sex prevalence [5].

Commonly, the tumor is found on the cutaneous and subcutaneous tissues of the distal extremities. So far, only 16 cases have been reported in the head and neck region [3]. To the best of authors knowledge, the present case is the first to be described in the infratemporal fossa. In addition, with 8 × 6 × 3 cm in size, it represents, with the case located in the orbit reported by Gbolahan et al., the largest SCH tumors ever described in the head and neck [8].

Histologically, SCH shows a proliferation of spindle cells composed of endothelial cells, pericytes and fibroblasts between dilated vascular spaces. Lesional cells show immunoreactivity for endothelial markers such as CD34, CD31, vimentin and factor VIII-related antigen [9].

SCH shows no or only a low level of mitotic activity. The distribution and percentage of the main histologic components may be highly variable [10].

Kaposi sarcoma lacks the cavernous spaces of spindle cell hemangioma, shows intracytoplasmic hyaline globules in spindle cells, lacks epithelioid vacuolated cells, and is invariably positive for human herpesvirus 8 (HHV-8) latent nuclear antigen, unlike spindle cell hemangioma [11].

Until recent years, it was thought that no genetic mutations or molecular characteristics were associated to SCH [12]. But Recent studies have shown that most sporadic SCH not associated with Maffucci syndrome also harbor mutations in isocitrate dehydrogenase. This mutation seems to be unique to SCH, because a wide range of other vascular tumors that have been studied lack mutations in isocitrate dehydrogenase [13].

Local excision is the treatment of choice for most lesions, while postoperative radiotherapy, low-dose interferon -2b, and intralesional and intra-arterial administration of recombinant interleukin-2 have been successful in treating and/or preventing recurrence of inaccessible or multiple SCH [14].

SCH is a benign lesion and the most widely acceptable treatment option presently is conservative excision without adjuvant chemotherapy or radiotherapy [15].

Following surgical excision, local recurrence rate of up to 58% has been reported. Recurrences occur more commonly in patients with multiple lesions at presentation, occurring near surgical sites rather than within them [5]. It is likely related to local intravascular propagation of the tumor [13].

Overall, prognosis is excellent and no incident of death from SCH have been reported till date [6].

4. Conclusion

This case report presents a unique presentation of a Spindle cell hemangioma in an unexpected location of the head and neck region. Currently, there is no case reported of a SCH in the infratemporal fossa. This article underlines the importance for clinicians, radiologists and pathologists to consider the SCH as a possible etiological diagnosis of infratemporal fossa tumors.

Declaration of Competing Interest

None.

Funding

None.

Ethical approval

The study is exempt from ethical approval in our institution as it is a "Case report" and not a research study.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

Not applicable.

Guarantor

Y. Oukessou.

Provenance and peer review

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CRediT authorship contribution statement

Y. Oukessou: Investigation, Resources, Writing - original draft, Writing - review & editing, Visualization. **M. Lyoubi:** Investigation, Resources, Writing - review & editing. **Y. Hammouda:** Writing - review & editing. **S. Rouadi:** Validation, Supervision. **R.L. Abada:** Validation, Supervision. **M. Roubal:** Validation, Supervision. **M. Mahtar:** Validation, Supervision.

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