ORIGINAL RESEARCH PAPER

General Surgery

HEMANGIOPERICYTOMA OF TONGUE: A RARE CASE REPORT

KEY WORDS:
Hemangiopericytoma, Long term follow-up, Vascular tumor

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ABSTRACT

Background: Hemangiopericytoma (HPC), described by Stout and Murray (1942), is a rare spindle cell tumor of mesenchymal origin with an incidence of about 0.06% per 100,000 population, affecting both genders equally. HPC typically develops in deep soft tissues, especially in the extremities, with only 16-25% of cases reported in the head-neck region, commonly affecting middle-aged individuals. **Case:** A 49-year-old male presented with a painless swelling in the tongue for two years. Intraoral examination revealed a large mass occupying the mouth. MRI suggested differential diagnoses of schwannoma or hemangioma. The mass was excised, and histopathologic examination confirmed a benign vascular neoplasm, diagnosing it as Hemangiopericytoma. **Conclusion:** Hemangiopericytoma is a rare vascular tumor with high recurrence and malignant potential, requiring awareness and long-term follow-up for effective management. Documenting such cases is crucial to increase awareness and improve understanding.

INTRODUCTION:

Hemangiopericytoma (HPC) is a rare mesenchymal cancer originating from Zimmerman's pericytes, the modified smooth muscle cells in capillary and postcapillary venule walls. (1) Histologically, HPC is characterized by randomly arranged spindle-shaped to ovoid cells, prominent staghorn vasculature, and the NAB2-STAT6 gene rearrangement. (2) HPC predominantly affects the soft tissues of the limbs and retroperitoneal space, but it is also commonly found among primary intracranial tumors. (1) Unlike meningiomas, which they resemble clinically, HPCs grow more rapidly and can cause symptoms through mass effect or local edema, leading to compression of the adjacent cerebral parenchyma and increased intracranial pressure. (3) In this rare case report we have presented a rare case of HPC of tongue.

Case Report

A 49 years old male presented with complaint of swelling in tongue in the last 2 years and having no pain or any discomfort. Intra-oral examination revealed large tongue almost completely occupying mouth of size 4 x 4 cm with dilated veins. **(Figure 1)** Palpation revealed mass to be non-tender, firm in consistency and no pulsations were felt with the lesion. MRI of tongue gave possible differential diagnosis of 1. a schwannoma or 2. a hemangioma involving anterior 2/3rd of tongue. **(Figure 2: A & B).** Intra–op findings- Longitudinal incision taken over left side. There was minimal bleeding present initially as multiple dilated veins in mucosa and submucosa. In deep submucosal layer mass is evident with well circumscribed and non-vascular structure. The mass was completely excised with blunt dissection. Polyglactin sutures used for layered closure. Oral gauze packing removed after 1 hour and replaced twice for minimal oozing. Ice packs given for 24 hours. Patient was relatively asymptomatic on IV antibiotics and analgesics for 2 days with post-operative minimal pain and slurred speech for 3 days. Liquids started on post op day 2 and soft diet on day 4. Patient was discharged on day 5 post surgery with advise of maintaining oral hygiene, soft and bland diet for another week and avoiding hot- spicy food. Histopathologic examination revealed benign vascular neoplasm composed of bland fusiform spindle cells with branching stromal vascular pattern with characteristic staghorn configuration. **(Figure 3)** Suggested final diagnosis as Hemangiopericytoma. Follow up was taken after 1 week, 2 weeks, 4 weeks, 8 weeks and 6 months and 1 year- patient was started on full diet after 2 weeks and he has no symptoms or signs of recurrence. If recurrence, it is planned to opt radiotherapy in future.

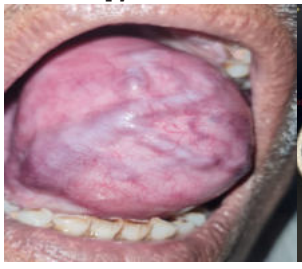

Fig. 1: Large tongue with dilated veins.

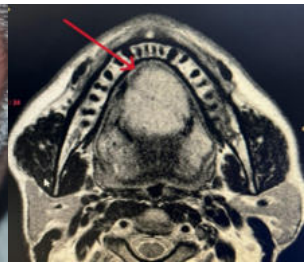

Fig. 2 A: MRI tongue Transverse plane.

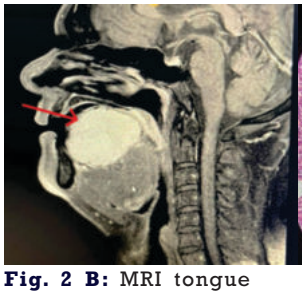

Fig. 2 B: MRI tongue Saggital plane

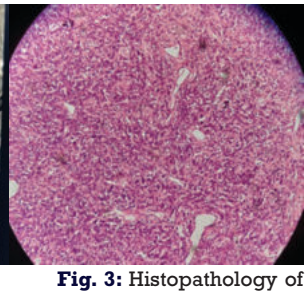

Fig. 3: Histopathology of tongue specimen

Fig. 2 A: Transverse plane: MRI tongue showing lesion involving anterior 2/3rd of tongue. **Fig 3:** Slide showing dilated vasculature and stroma hyalinized to collagenous to myxoid change.

DISCUSSION:

Hemangiopericytoma (HPC) is an uncommon neoplasm, with fewer than 300 reported cases, predominantly involving the trunk and lower limbs. [4] Involvement of the head and neck, particularly the tongue, [5] is exceedingly rare, making our case noteworthy. HPCs are more common in middle-aged individuals, [1-3] aligning with our case of a 49-year-old male presenting with HPC in the anterior two-thirds of the tongue.

Histologically, In our case, Examination revealed benign vascular neoplasm composed of fusiform spindle cells with a staghorn vascular pattern, consistent with other case reports describing similar spindle cell arrangements and vascular proliferation. [5]

HPCs can remain asymptomatic until advanced stages due to their indolent nature. [1-5] Our patient had an asymptomatic tumor for two years. MRI was utilized to assess tumor extension, corroborating with other reported cases. [1-5]

Effective management of HPC remains controversial due to its rarity and unpredictable behaviour. Surgical resection is the primary treatment, significantly improving overall survival and cancer-specific survival. However, over 30% of patients experience recurrence post-surgery, commonly in the retroperitoneum and pelvis. [6,7] Radiotherapy, although not providing significant survival benefits, is recommended to reduce local recurrence risk in tumors larger than 5 cm or with inadequate resection margins. [8,9]

The efficacy of chemotherapy in HPC patients is unclear, though it may be beneficial for metastatic cases. [7,10] Due to the high recurrence rate and unpredictable clinical behaviour, long-term follow-up is essential for HPC patients.

CONCLUSION

In conclusion, hemangiopericytoma (HPC) of the tongue presents as an unusual tumor that poses both diagnostic and management challenges. Surgical resection is the cornerstone of treatment, aiming for complete excision despite the tumor's propensity for recurrence. Adjuvant radiotherapy should be considered in cases with high-risk features to minimize recurrence. Given the unpredictable nature of HPC, long-term surveillance is imperative to detect and manage any recurrence early. Multidisciplinary collaboration and ongoing research are crucial to improving the prognosis and outcomes for patients with this rare condition.

Conflict Of Interest: None

Financial Support: None

Ethical Approval: NA

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