SKULL BASE LEIOMYOSARCOMA MIMICKING NEUROFIBROMA

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1. Parapharyngeal Space Tumors

- 0.5-1.5% of head and neck neoplasms[7]
- Most common types[7]:
  - Benign (71%), ex. pleomorphic adenoma
  - Malignant (29%), ex. squamous cell carcinoma of parotid
- Presentation:
  - Dysphagia
  - Difficulty breathing, speaking
  - Swelling
  - Discomfort in neck
- Evaluation:
  - MRI or CT
  - Fine needle aspiration
  - Immunohistochemical staining following resection
- Management:
  - Resection via transparotid, transoral, and/or transcervical approach,
  - Postoperative radiation therapy

Leiomyosarcoma

- Immunohistochemistry:
  - S-100 negative
  - Smooth muscle actin (SMA) positive

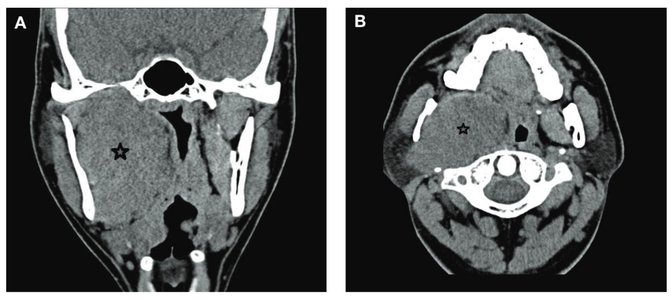

Fig 1: Image showing a leiomyosarcoma of the right parapharyngeal space (indicated with a star). A) coronal CT. B) axial CT. Image source: Locatello et. al.[5]

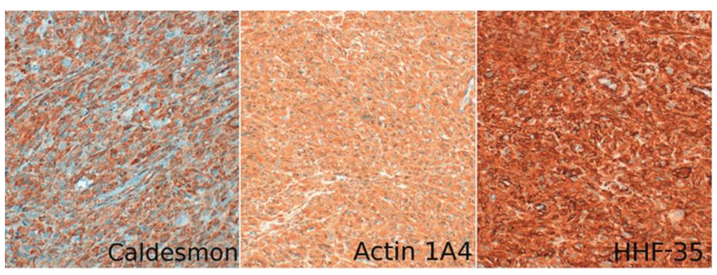

Fig 2: Image showing immunohistochemical staining for a leiomyosarcoma of the right parapharyngeal space. From left to right: caldesmon, actin 1A4, HHF-35.
Image source: Locatello et. al.[5]

Neurofibroma

- Presentation and evaluation are typically alike that of leiomyosarcomas, but management can just be observation
- Immunohistochemistry:
  - S-100 positive
  - Smooth muscle actin (SMA) negative

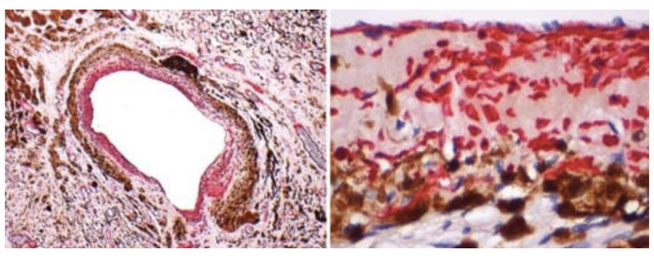

Fig 3: Image showing immunohistochemical staining for S-100 and actin for a benign neurofibroma of the spine.
Image source: Yang et. al.[6]

3. Case Presentation

A 55-year-old female presented with progressive hoarseness, ipsilateral otalgia, and right true focal fold paralysis. Neck CT demonstrated a 3 cm right carotid space mass suspicious for neurofibroma. CT-guided biopsy demonstrated spindle cells compatible with a peripheral nerve sheath tumor (Fig. 4). Preoperative MRI showed a T2 hyperintense mass in the right carotid space consistent with neurofibroma (Fig 5). Preoperative examination demonstrated the neurologic deficits listed below.

Physical Exam:
- Hoarseness
- Ipsilateral otalgia
- Right true focal fold paralysis
- Palatal asymmetry
- Nasopharyngeal regurgitation
- Ptosis (suggestive of sympathetic chain involvement)
- Tongue weakness (suggestive of hypoglossal nerve dysfunction)

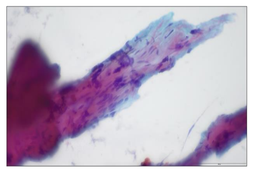

Fig 4: Pre-operative fine need aspiration showing fragments of bland spindled cells.

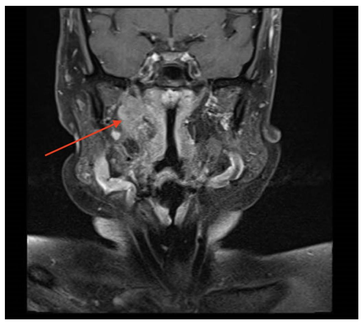

Fig 5: Pre-operative T2-weighted coronal MRI: enhancing mass of right carotid space with anteromedial displacement of hypoglossal nerve and posterolateral displacement of internal carotid artery.

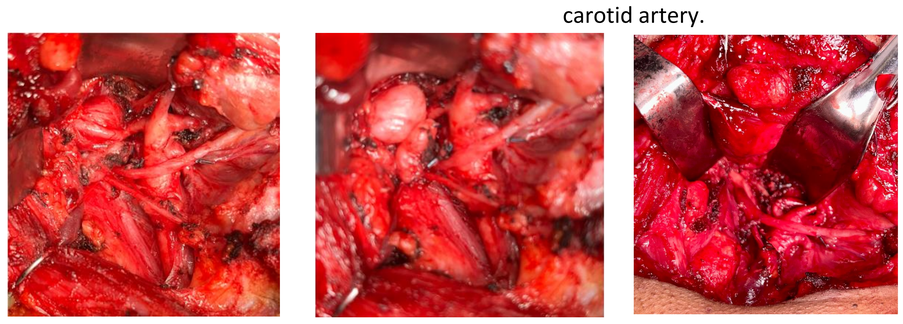

Fig 6: Intraoperative images of the parapharyngeal tumor. Left: occipital artery crossing superficial to tumor. Center: tumor overlying hypoglossal nerve at the skull base after ligation of occipital artery. Right: resection bed after tumor extirpation.

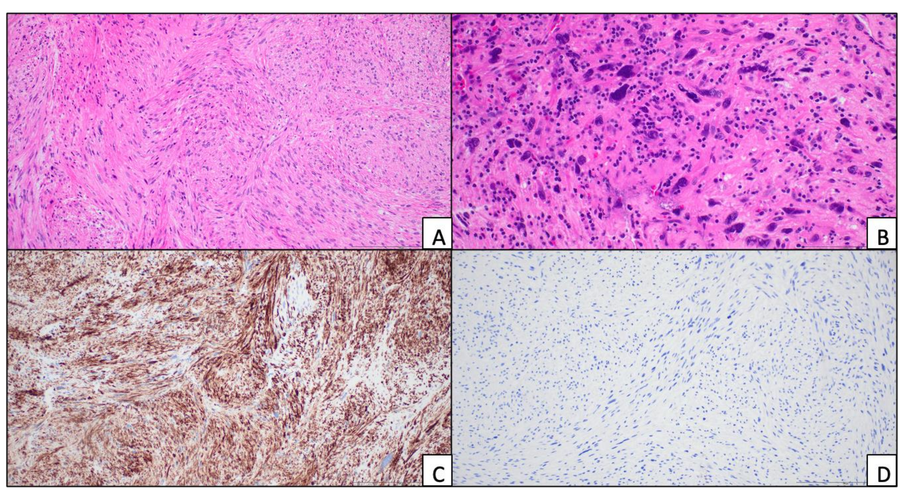

Fig 7: Resection specimen pathology. A) H&E section showing spindled cells with mild cytologic atypia similar to biopsy. B) Other areas show significant pleomorphism with large nuclei and hyperchromasia. C) Immunohistochemical stain for desmin was strongly, diffusely positive, indicating smooth muscle differentiation. D) Stain for S-100 was negative, providing no support for nerve sheath differentiation and neurofibroma.

The patient then underwent trans-mastoid occlusion of the sigmoid sinus, transcervical approach to the right infratemporal fossa, and right laryngeal innervation using right ansa cervicalis for tumor resection (Fig. 6).

Final pathology was a grade 2 leiomyosarcoma. Immunohistochemistry of the resected tumor was notable for S-100, a marker of Schwann cells, negativity and smooth muscle actin positivity, consistent with leiomyosarcoma (Fig. 7). Additionally, the tumor had increased Ki-67 proliferation and 0-5 mitoses per 10 hpf, consistent with a malignant process.

4. Post-operative Course

Patient discharged on a full liquid diet. Staging PET CT was consistent with hepatic metastases, confirmed with biopsy. Recommendation was for adjuvant chemoradiotherapy. Simulation CT for radiation therapy demonstrated rapid recurrence at the skull base, confirmed on MRI (Fig. 8).

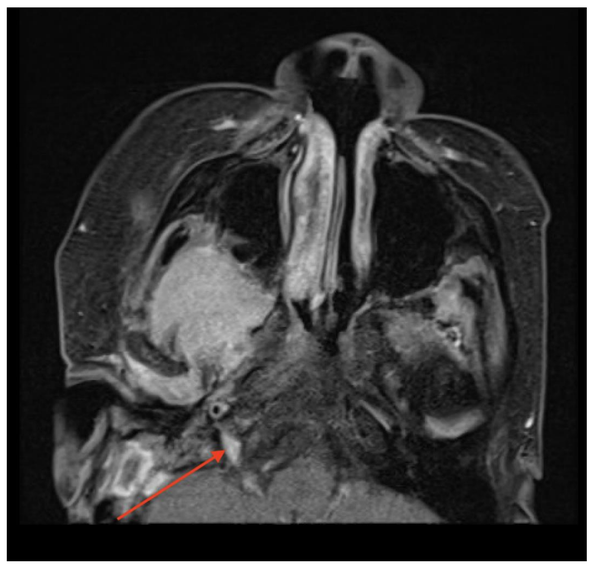

Fig 8: Two-month post-operative MRI with red arrow indicating recurrence of large 6.5 cm trans-spatial mass

5. Discussion

- Parapharyngeal space tumors are often benign
- Neurofibromas and leiomyosarcomas are rare tumors of the head and neck with similar imaging characteristics of T2 hyperintensity on MRI that can present with neurological deficits
- Leiomyosarcomas: S-100 negative, smooth muscle protein positive
- Neurofibromas: S-100 positive, SMA negative

5. Conclusion

- This is the 6th reported case of a leiomyosarcoma of the parapharyngeal space
- This case was complicated by preoperative image-guided biopsy suggestive of benign nerve sheath tumor.
- In patients without a history of a genetic neurocutaneous disorder, additional spindle cell tumors should be considered via immunohistochemistry
- Metastatic leiomyosarcoma has a poor prognosis and early diagnosis is key for improved survival

6. References

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