

Abstract

A 38 year old female with germline succinate dehydrogenase subunit D (SDHD) mutation presents with multifocal paragangliomas.

Negative distant PET dotatate imaging and negative urine and plasma metanepherines allowed for resection of the left sided carotid body and jugulotympanic lesions.

Preoperative embolization of both lesions was performed prior to the resection. The resection was completed without vessel injury or large cerebrospinal fluid leak. After a brief inpatient hospital stay, the patient was discharged to pursue radiation therapy for the residual jugulotympanic lesion and contralateral paragangliomas.

Case

A 38 year old female with germline succinate dehydrogenase subunit D (SDHD) mutation presents with multifocal paragangliomas.

On exam she has significant cranial neuropathies including dense left VII (HB 6/6), paretic X on flexible laryngoscopy and weak XII with tongue atrophy. Additionally, she has a left profound sensorineural hearing loss.

Workup with PET dotatate was negative for abdominal or thoracic paragangliomas (Figure 1A). CT with contrast demonstrated bilateral Shamblin II carotid lesions, left larger than right, small right vagal and right paratracheal lesion (Figure 1B).¹ MRI demonstrated large left sided jugulotympanic lesion eroding the jugular foramen abutting the posterior fossa and invading the ear canal, Fisch type C/Glasscock Type 3 (Figure 1C).^{2,3}

Plasma and urine metanepherine levels were within normal limits. Preoperative balloon occlusion test of the left internal carotid demonstrated poor contralateral collateral flow. Preoperative embolization of the left carotid body and jugulotympanic lesion was performed targeting the proximal and distal ascending pharyngeal artery as well as feeding vessels from the vertebral and occipital arteries. She suffered post-embolization seizures controlled medically.

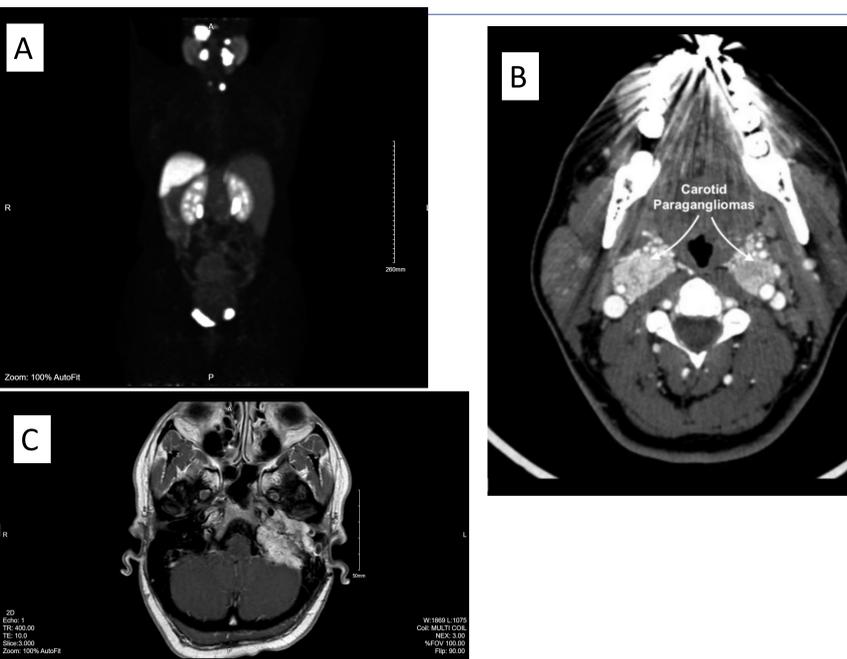


Figure 1: A. PET dotatate demonstrating no evidence of abdominal or intrathoracic paragangliomas. B. CT with contrast demonstrating bilateral Shamblin I carotid body lesions. C. MRI IAC demonstrating large jugulotympanic lesion abutting CPA

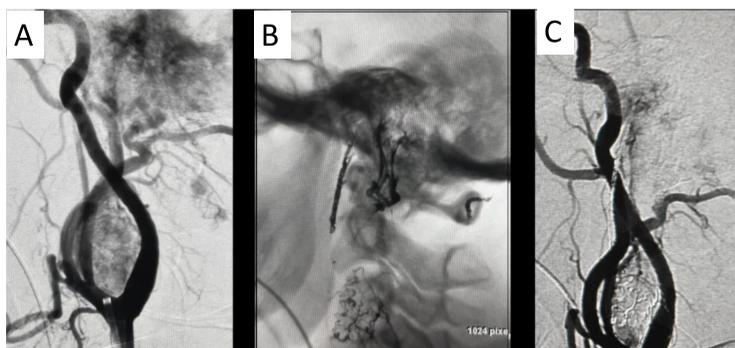


Figure 2: A. Pre embolization angiography demonstrating extensive vasculature of left carotid body lesion and jugulotympanic lesion, lyre sign of the carotid lesion is appreciated. B. Deployment of coiling and onyx to achieve embolization. C. Post embolization angiography demonstrating significant reduction in blush.

Intervention

Surgical resection of the left sided lesions was undertaken with plan for post operative radiation of residual tumor and contralateral lesions.

Neck dissection was performed obtaining vessel control of the distal and proximal carotid as well as jugular vein. The carotid body lesion was resected without vessel injury and the internal jugular vein was ligated.

A modified Fisch A approach was utilized to access the glomus jugulare.⁴ Given gross involvement of the facial nerve and baseline dense facial paresis, the facial nerve was sacrificed. The dissection was carried to the petrous apex leaving residual tumor over the petrous carotid to avoid injury. The jugular bulb was resected, preserving the medial aspect to avoid permanent cranial neuropathy. The tumor was abutting the posterior fossa dura inferiorly to the level of the V3 vertebral artery and was noted to involve the dura extensively. The dura was decompressed but not violated. The dissection was carried minimally into the cerebropontine angle, at which a low flow cerebrospinal fluid leak was encountered. The leak was repaired using a fascia graft, Surgicel, and fibrin glue. Further dissection to the hypoglossal canal was completed. The eustachian tube was obliterated with a fascia graft and Surgicel.

A platinum eyelid weight was placed and post operatively the patient was noted to have a left vocal fold paralysis despite stimulation of X at the skull base intraoperatively after tumor resection. She was later rehabilitated with vocal fold injection. She was discharged on an oral diet and did not develop post operative leak.

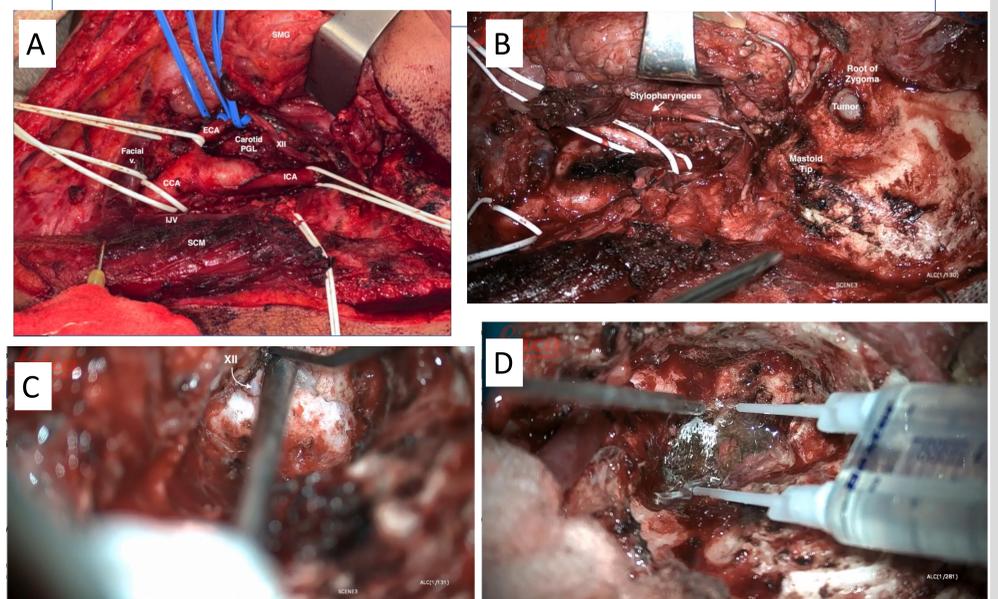


Figure 3: Intraoperative footage of resection. A. Neck dissection demonstrating carotid body lesion with anatomic structures labeled. B. Surgical view prior to mastoidectomy with EAC resected. C. Skull base resection down to hypoglossal canal. D. Surgical and fibrin glue skull base reconstruction.

Discussion

The decision to surgically remove the larger carotid body lesion may be considered a deviation from the typical treatment for bilateral carotid body lesions.^{5,6} The already present cranial neuropathies and need for vessel dissection for jugular vein sacrifice provided the justification for addressing the ipsilateral carotid body lesion surgically. This allowed for the implementation of a comprehensive treatment plan of her multifocal disease.

Addressing both lesions does present the risk of significant intraoperative blood loss and prolonged operative times. As such, preoperative embolization was leveraged to reduce blood loss which was recorded at ~1L.⁷

Subtotal but extensive resection of the jugulotympanic lesion allowed the surgical team to avoid durotomy and significant cerebrospinal fluid leak with confidence that post operative radiation would achieve a high rate of control.

Conclusions

Management of multifocal paragangliomas in the setting of germline SDHD is challenging. Workup with PET dotatate and urine/plasma metanepherines is essential prior to developing treatment plans for skull base lesions.

Careful consideration of cranial neuropathies should be involved in treatment planning. The combination of surgical intervention with adjuvant radiation can optimize outcomes.

Preoperative embolization may be helpful to reduce intraoperative blood loss to provide comprehensive surgical intervention.

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