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Recurrent Sporadic Malignant Triton Tumor in the Carotid Sheath in a Patient Without Neurofibromatosis

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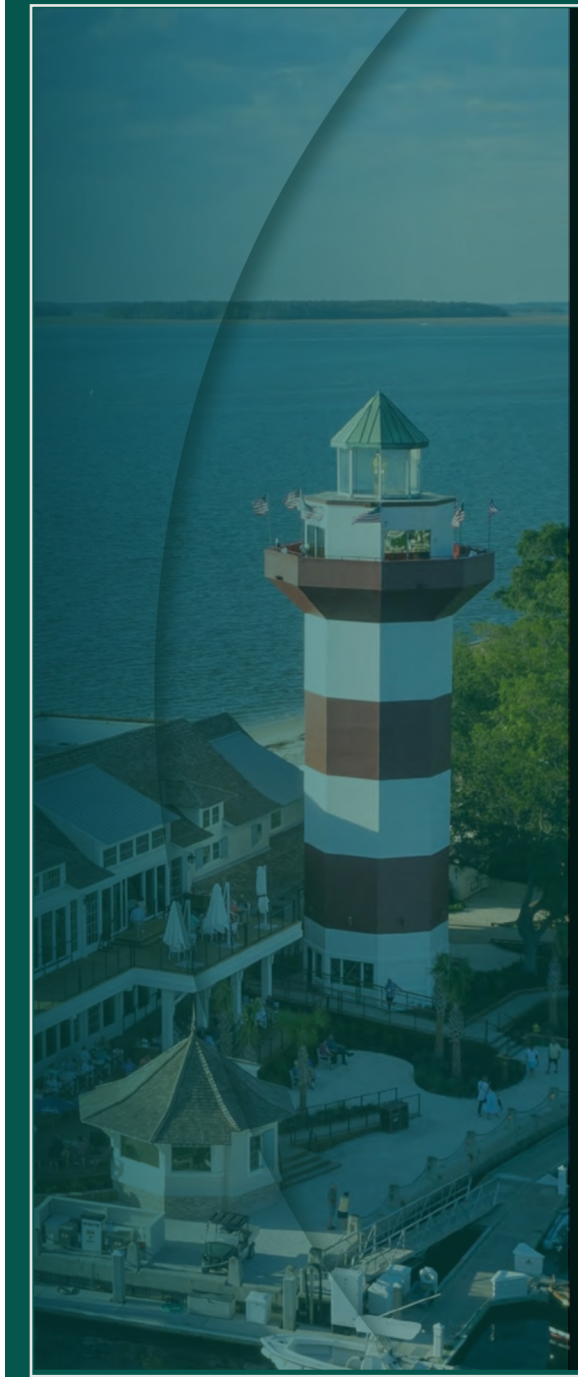


The authors have nothing to disclose

Case presentation

Middle aged male

ER

- Fatigue
- Swelling
- Non-radiating pain in the right neck

+

- gagging sensation
- persistent recurrent hiccups that would last up to 5 minutes.

Case presentation

Examination findings

1. Neck: Tender to palpation
2. Neurological assessment: Unremarkable
3. Carotid bruit: Not detected

Laboratory findings

Negative for leukocytosis or electrolyte imbalance

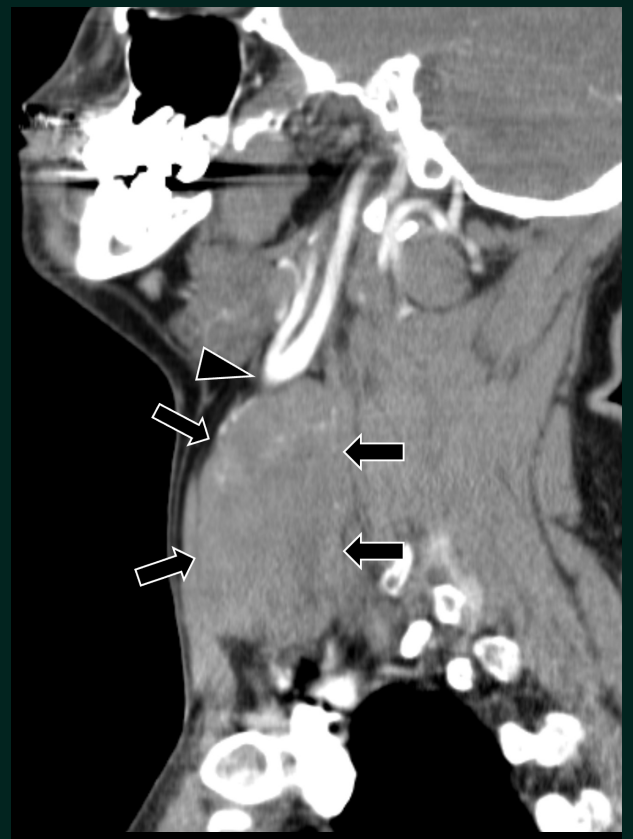
Pertinent prior history

1. History of biopsy-confirmed malignant peripheral nerve sheath tumor (MPNST) exhibiting rhabdomyoblastic and cartilaginous differentiation in the right neck, involving the carotid body and brachial plexus, 8 years prior to presentation.

2. History of partial resection of the MPNST (with positive margins), interposition saphenous vein graft to the right carotid artery, radiation therapy, and chemotherapy using sodium 2-mercaptoethane sulfonate (*Mesna*), Ifosfamide, and Adriamycin at an outside hospital.

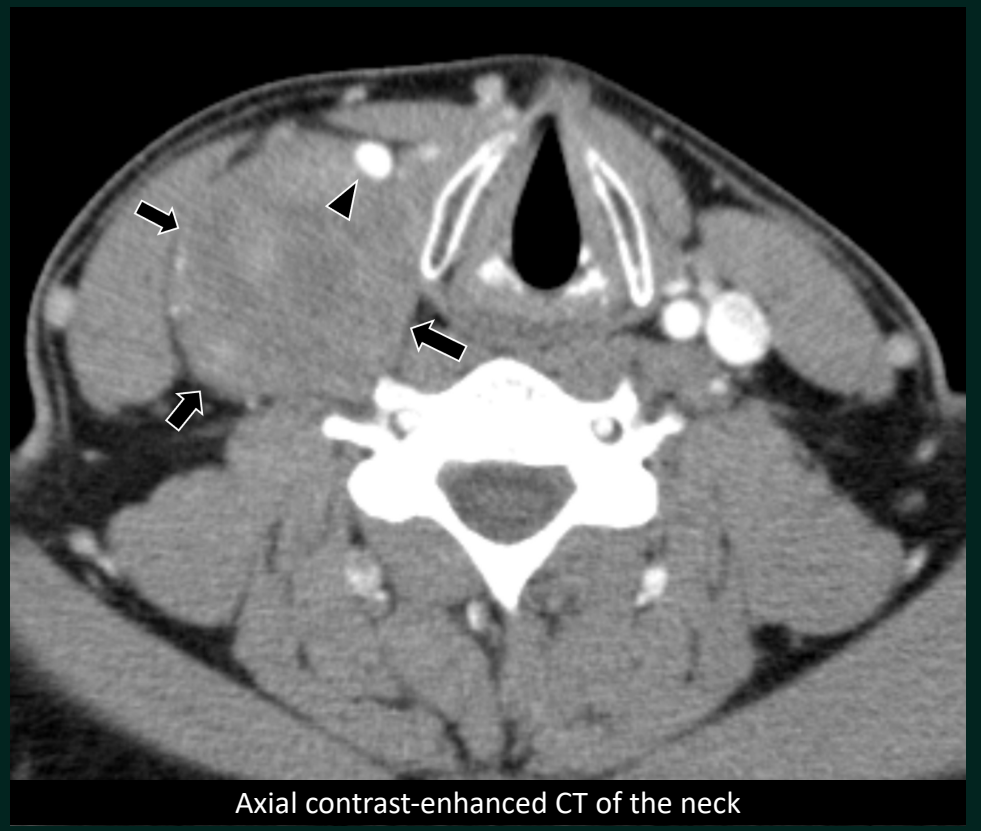
Case presentation

Imaging findings



Sagittal contrast-enhanced CT of the neck

Homogenous, rim-enhancing solid mass in the right carotid sheath (arrows), immediately caudal to the carotid bifurcation (arrowhead)

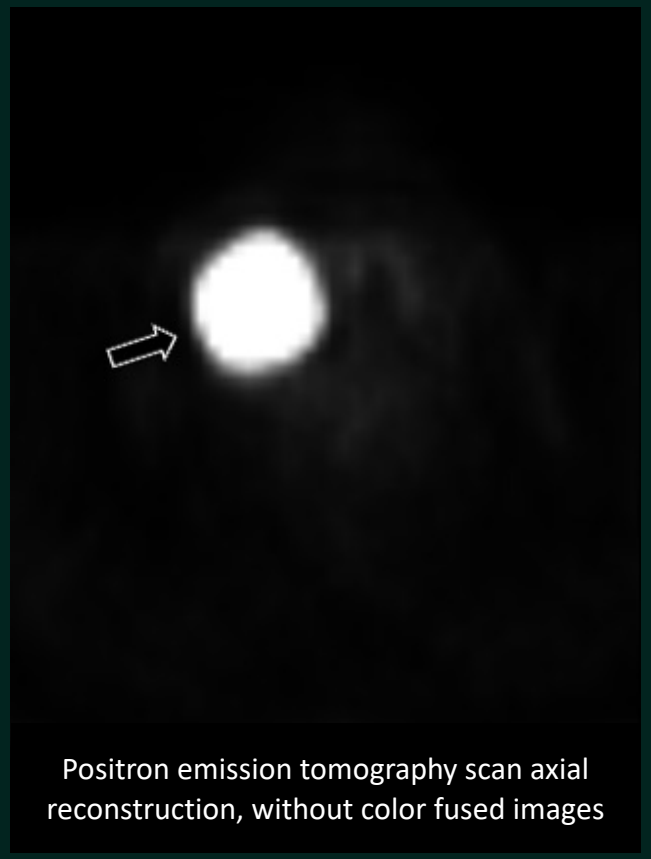


Axial contrast-enhanced CT of the neck

Homogenous, rim-enhancing solid mass in the right carotid sheath (arrows). The right common carotid artery was observed coursing within the margins of the mass, with mild focal stenosis (arrowhead). No evidence of wall violation.

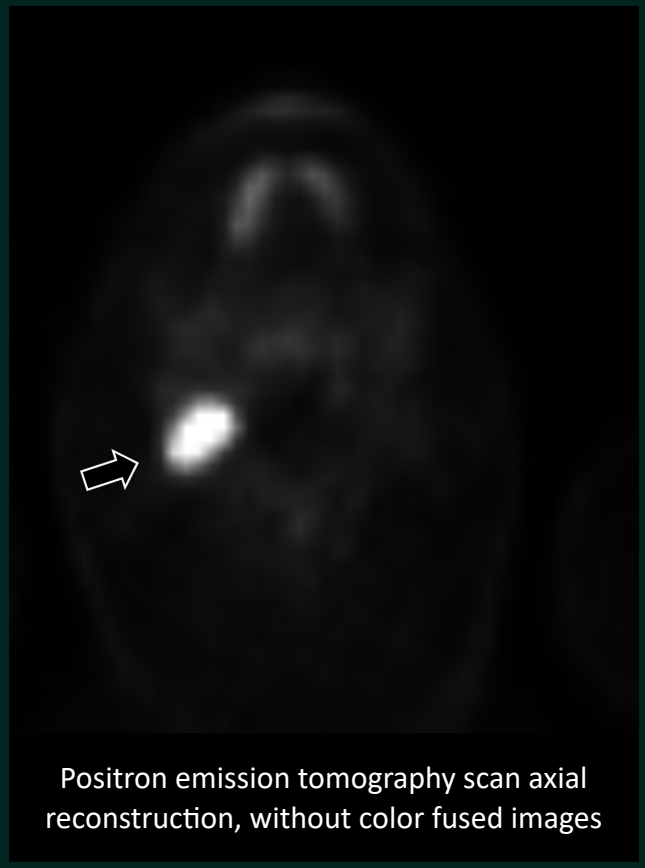
Case presentation

Imaging findings



Positron emission tomography scan axial reconstruction, without color fused images

Intense fluorodeoxyglucose (FDG) avidity in the right neck mass (arrow), with no metastatic disease evidence



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Case presentation

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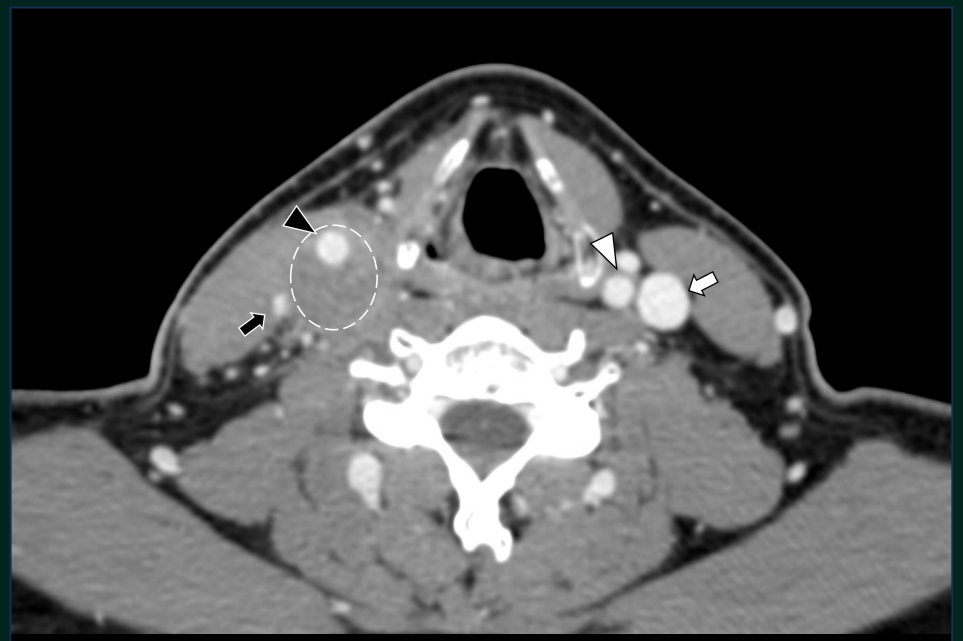
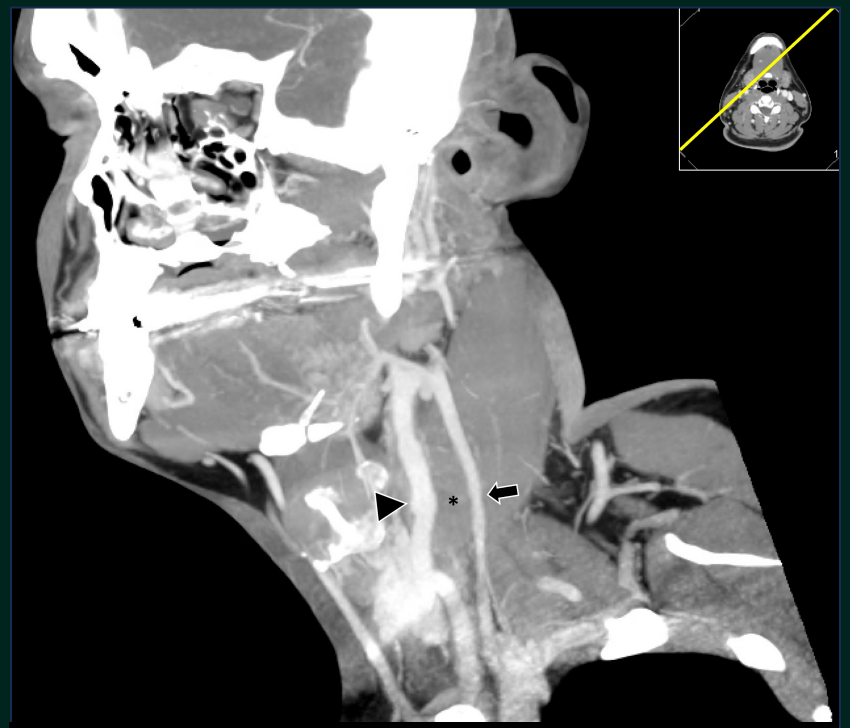


Contrast-enhanced axial CT of the neck

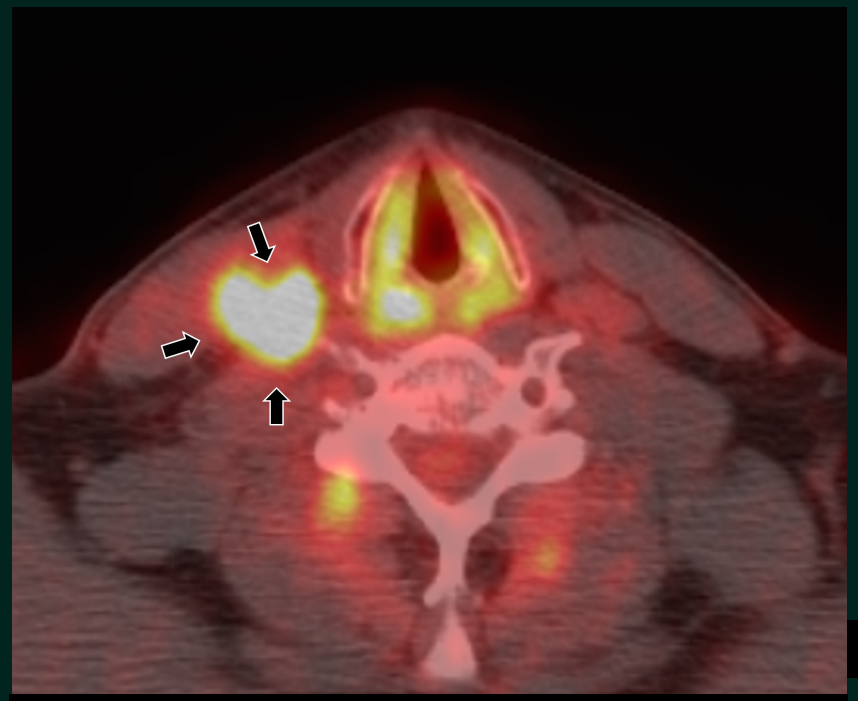
Splaying and anterior displacement of the right common carotid artery (black arrowhead) and posterior displacement of the right internal jugular vein (black arrow) by the mass (dotted circle). Note normal left-sided common carotid artery (white arrowhead) and internal jugular vein (white arrow) for comparison.



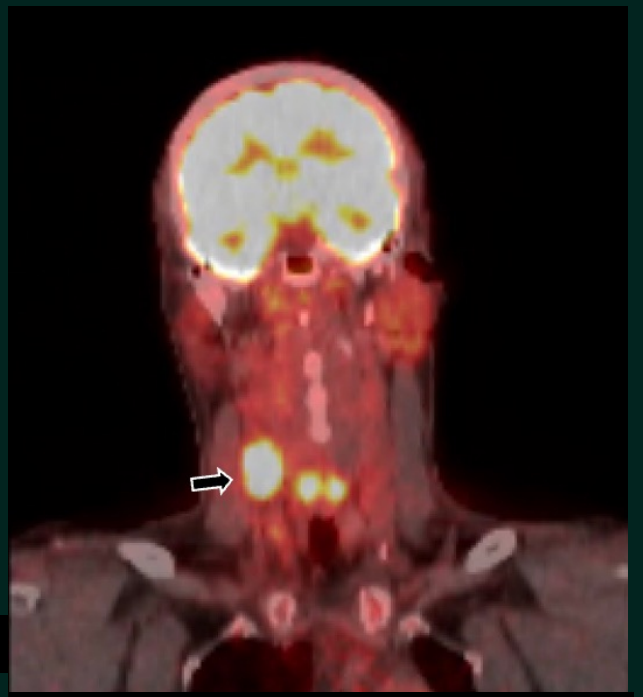
Contrast- enhanced CT of the neck maximal intensity projection (MIP) reconstruction in the plane of the right common carotid artery and the right internal jugular vein (inset)

Splaying of the common carotid artery (arrowhead) and the internal jugular vein (arrow) by the mass (*).

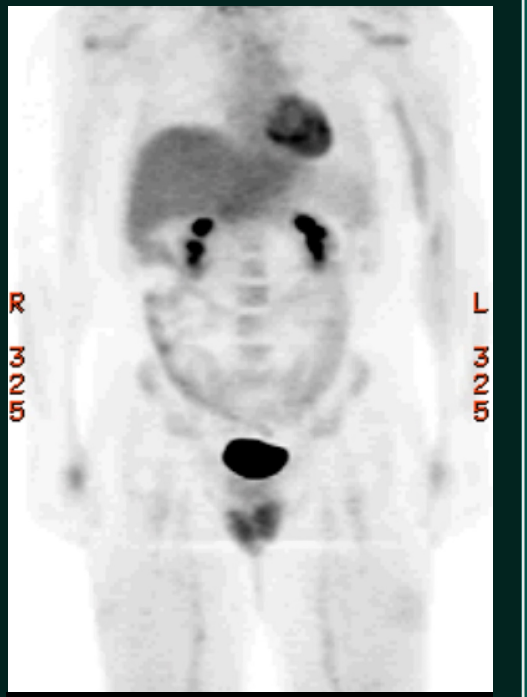
Imaging findings



Axial fused positron emission tomography (PET) scan



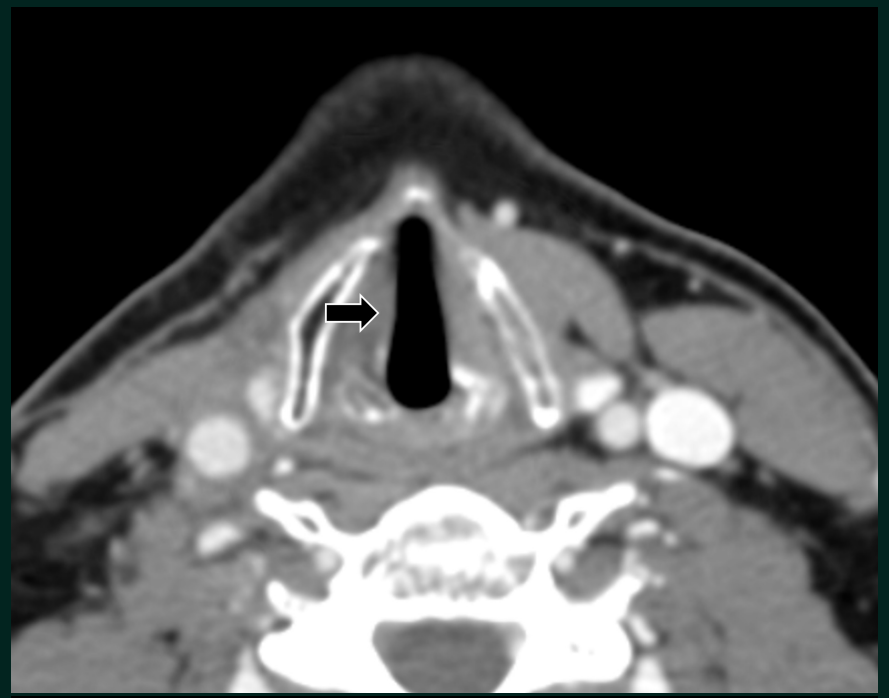
Coronal fused positron emission tomography (PET) scan



Coronal maximal intensity projection

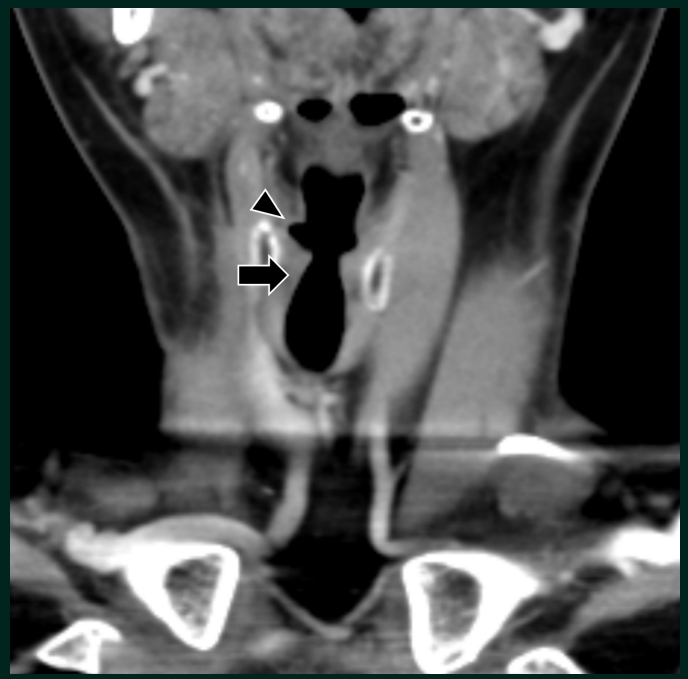
Intense fluorodeoxyglucose (FDG) avidity in the right neck mass (arrows) without demonstrated evidence of metastatic disease.

Imaging findings



Contrast-enhanced CT of the neck axial reconstruction

Slight medialization of the right vocal cord (arrow), suggesting right vocal cord palsy.

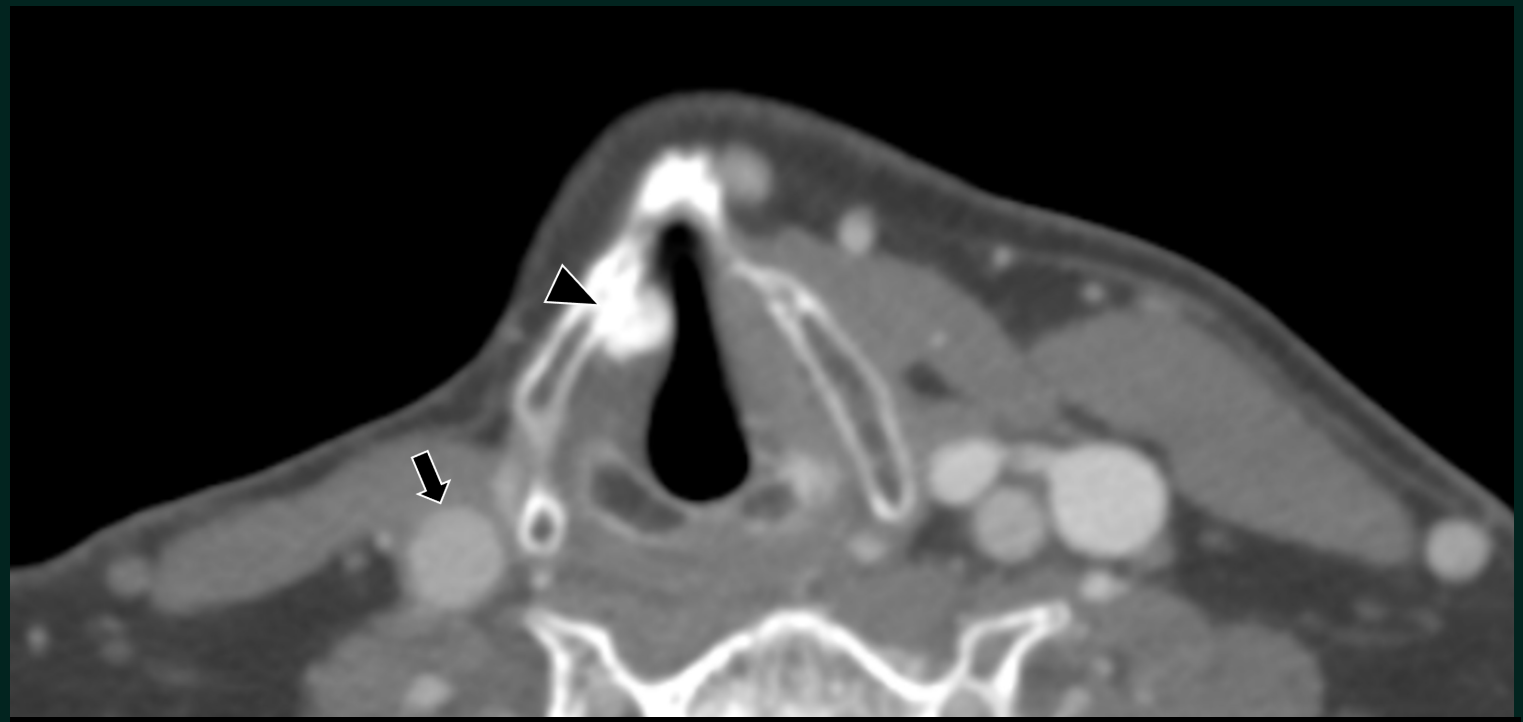


Contrast-enhanced CT of the neck coronal reconstruction

Slight medialization of the right vocal cord (arrow) and dilated right laryngeal ventricle, suggesting right vocal cord palsy.

Case presentation

Imaging findings



Contrast-enhanced CT of the neck axial reconstruction

No signs of recurrence after 8 years (arrow). Additionally, the arrowhead points to evidence of right vocal cord medialization treatment.

Case presentation

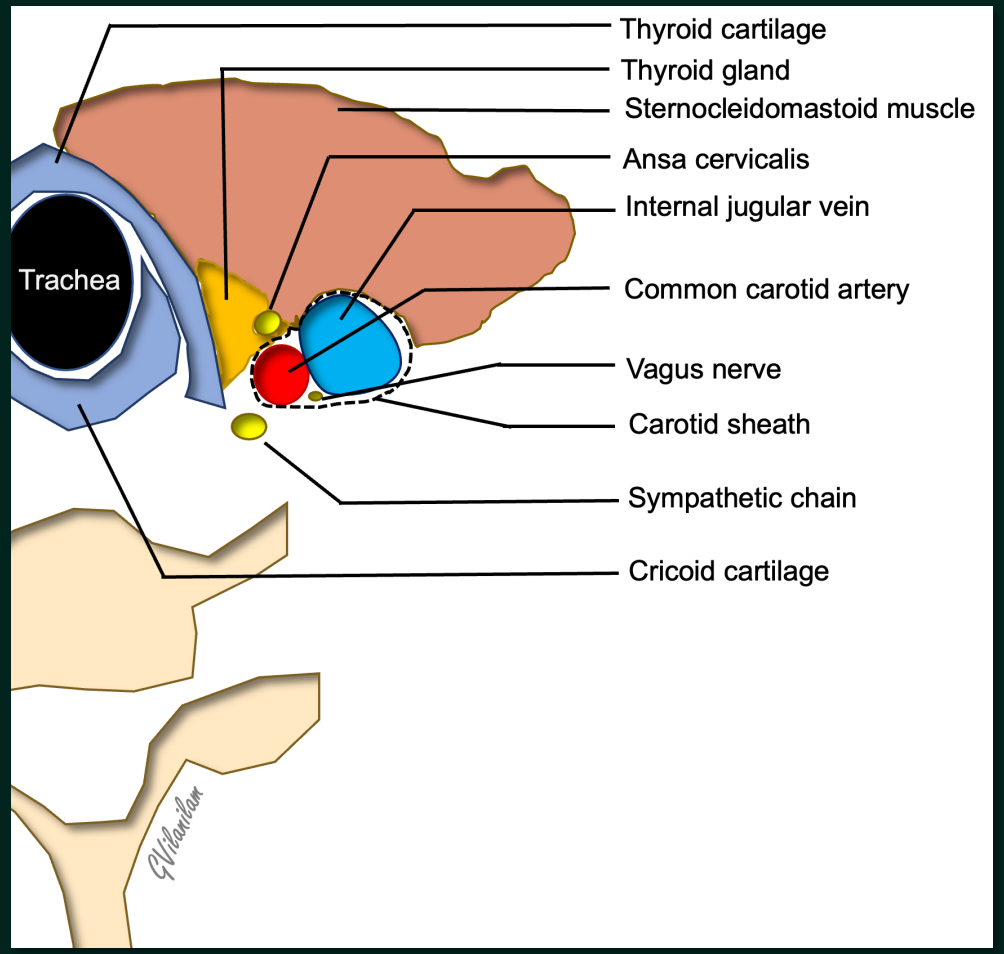
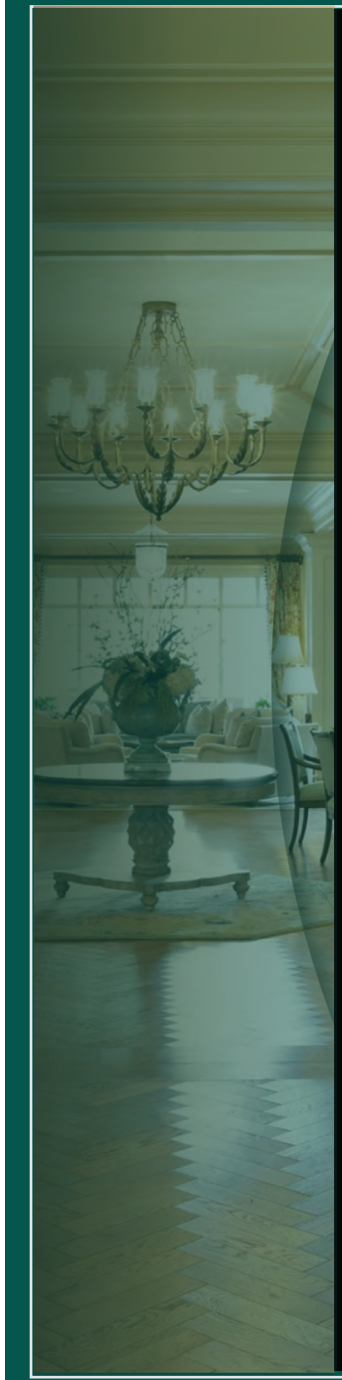
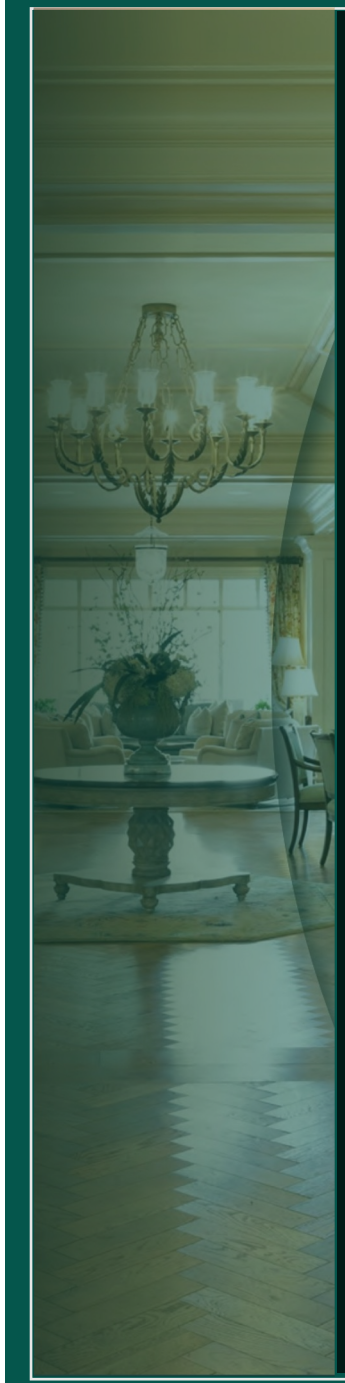


Illustration of the infrahyoid neck at the level of the cricoid cartilage demonstrates a normal carotid sheath containing the common carotid artery, internal jugular vein, and vagus nerve. The sympathetic chain lies posterior to the carotid sheath.

1. A carotid sheath mass involving the vagus nerve displaces the common carotid artery anteromedially and the internal jugular vein posterolaterally.
2. A mass originating from the sympathetic chain will displace the common carotid artery and the internal jugular vein together anterolaterally.



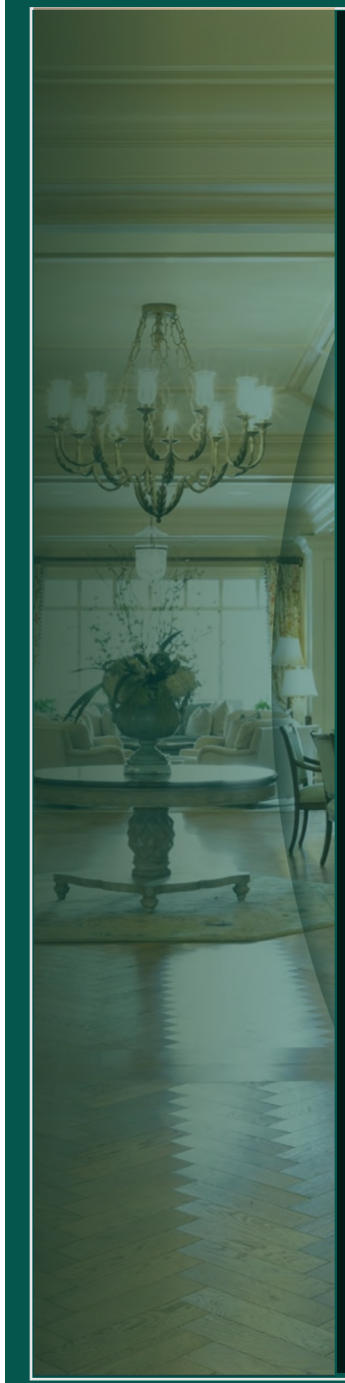
Diagnosis: Biopsy proven recurrent sporadic malignant Triton tumor (MTT); a rare and aggressive variant of MPNST



Discussion and learning points

History and epidemiology: MTT is a rare and aggressive variant of MPNST, accounting for <10% of all MPNSTs. Histologically, it is a malignant soft tissue sarcoma of the peripheral nerves characteristically with rhabdomyoblastic differentiation. It was named after the triton salamander which has the ability to regenerate skeletal muscle from neural tissue.

Diagnostic criteria: Tumors arising from a peripheral nerve or in a patient with neurofibromatosis or representing metastasis from such a tumor, having the growth characteristics of Schwann cells, and the presence of contained rhabdomyoblasts. Sporadic cases with the latter two criteria were later also included.

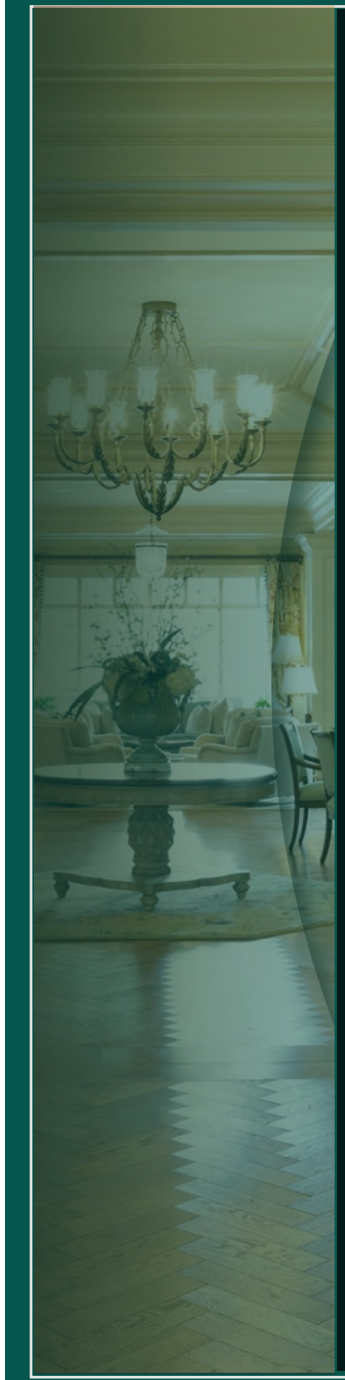


Discussion and learning points

Etiology: Classically associated with Neurofibromatosis, with 60%–70% of cases occurring in adults with neurofibromatosis 1 (NF1). In NF1 patients, it occurs more commonly in males under 35 years. Sporadically, it is extremely rare with the estimated incidence of 1.46 per 1,000,000 individuals and is seen to occur in older individuals. MTT can also be associated with radiation exposure.

Clinical features: Rapidly growing mass that can cause neurological symptoms depending on its location. The size of the tumor at presentation is usually greater than 5 cm, and up to 50% of patients present with metastasis to the lung.

Location: Near peripheral nerves in the head and neck, as well as in extremities such as the sciatic nerve, brachial plexus, cervical plexus, and cervicosympathetic nerve.

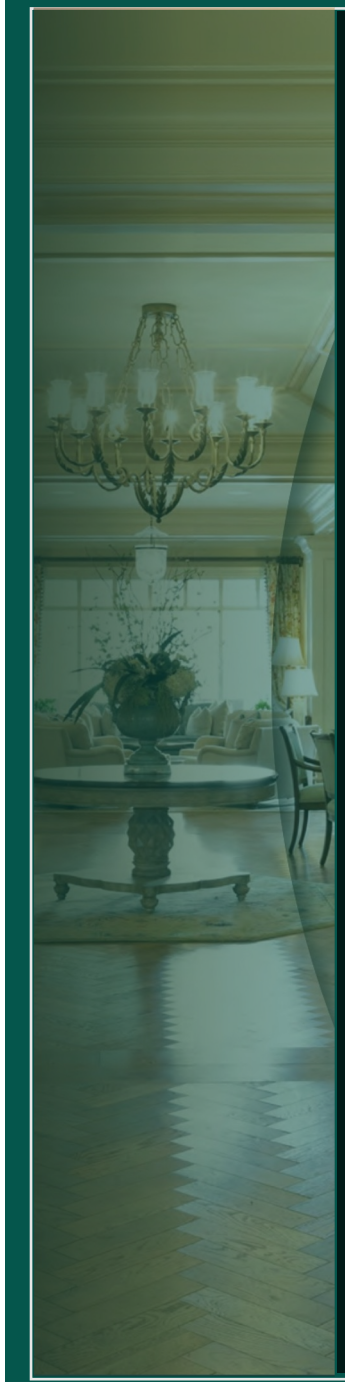


Discussion and learning points

Clinical pearl: In patients with NF, it is important to monitor for the development of new or worsening pain in existing lesions, as this may indicate MTT

Utility of imaging:

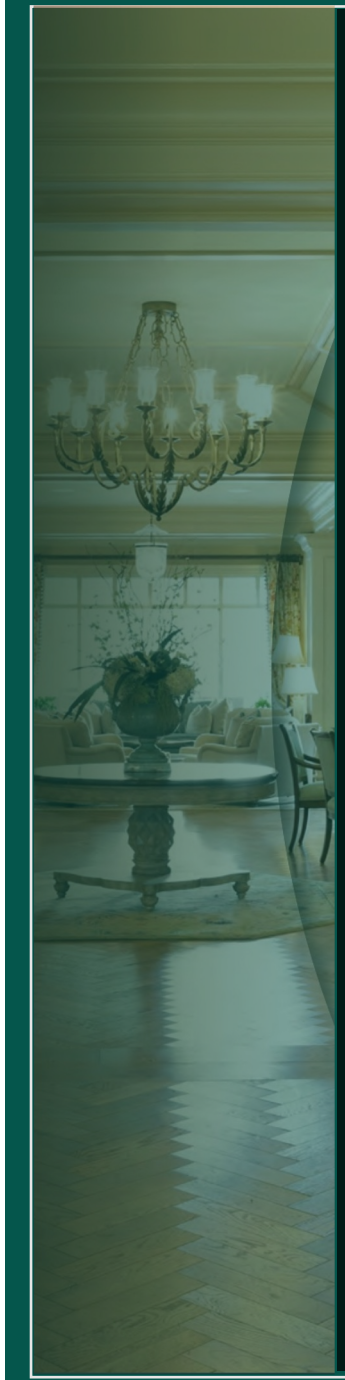
- Ultrasound: MTTs present as large, elongated, and hypoechoic masses.
- CT and MRI: Highly useful in characterizing the mass and delineating the location of the MTT.
- CT: Reveals low attenuation masses usually due to fat entrapment, high lipid content, and cystic areas.
- MRI: MPNSTs (and MTTs) appear isointense to muscle on T1-weighted images and hyperintense on T2-weighted images with fascicular appearance
- FDG-PET: effective for assessing prognosis and detecting metastases.



Tips for localization of mass with CT and MRI:

1. In the infrahyoid neck, the carotid sheath normally contains the common carotid artery, the internal jugular vein, and the vagus nerve. The sympathetic chain is situated posteriorly and outside of the carotid sheath. (Figure in slide 11).

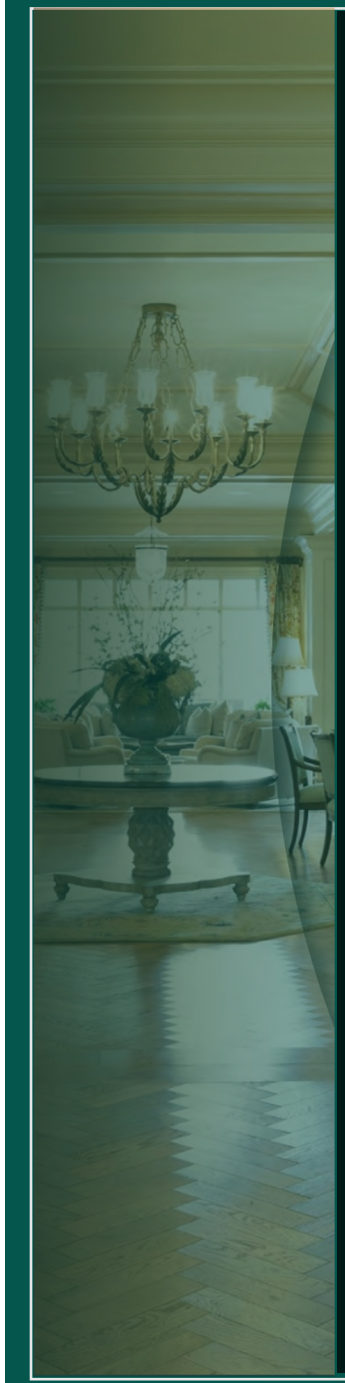
2. Distinguishing between a mass arising from the carotid sheath involving the vagus nerve and a mass originating from the posterior sympathetic chain: Assess the mass effect on the common carotid artery and the internal jugular vein; if the mass originates from the carotid sheath involving the vagus nerve, the common carotid artery is displaced anteriorly and medially, whereas the internal jugular vein is displaced posteriorly and laterally. Conversely, if the mass arises from the sympathetic chain, it displaces the contents of the carotid sheath anterolaterally (figure in slide 11).



Discussion and learning points

Histopathology: Definite diagnosis is possible only after excision and biopsy. Histologically, MTTs typically consist of monotonous spindle cells arranged in fascicles with variable hypo- and hypercellular areas.

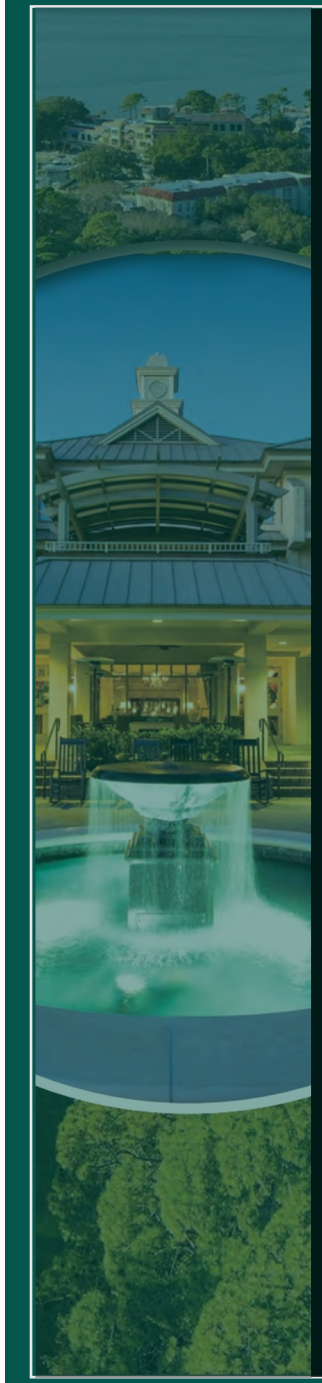
Immunohistochemical staining is crucial in identifying the origin of tumor cells. S-100 protein positivity confirms nerve sheath differentiation, while positivity for desmin, actin, and myogenin confirm rhabdomyoblastic differentiation, as in our patient.



Discussion and learning points

Treatment: Complete surgical resection with negative margins is the treatment of choice for MTT. Tumor respectability is the most important prognostic factor for local MSPNTs. Adjuvant radiotherapy may also provide some benefits, but it needs further research.

Prognosis: Generally poor, with larger tumor size (>5 cm), association with NF-1 and history of radiation therapy associated with worse outcomes. The 5-year survival has been reported around 14% with an overall local recurrence rate of around 50%. Despite the challenges in treating these tumors, early detection and intervention can improve patient outcomes.

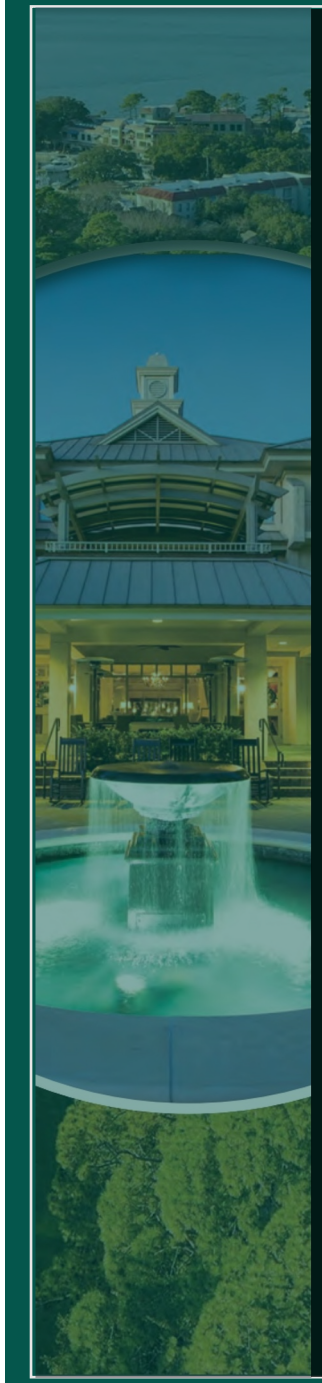


Conclusion and take-home points

1. We have described an uncommon instance of recurrent sporadic MTT within the carotid sheath in a patient without any personal or familial history of neurofibromatosis.

2. Accurate diagnosis of this condition relies on a comprehensive evaluation of the patient's clinical history, including symptoms, neurofibromatosis history, prior radiation therapy, imaging characteristics, location, size, and biopsy findings.

3. Prompt and complete surgical resection of the tumor is essential for optimizing patient outcomes in such cases.



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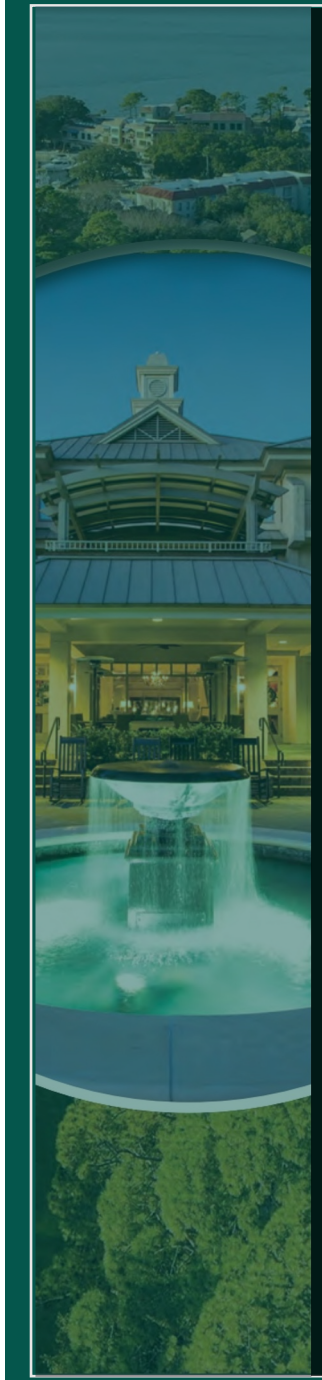

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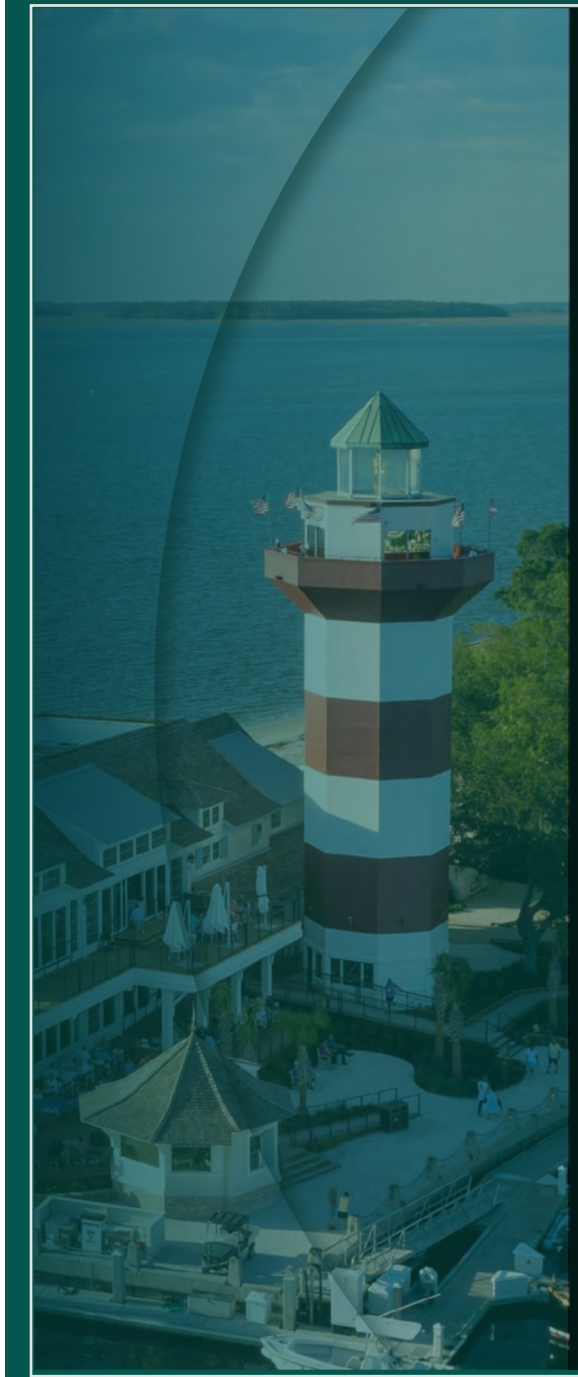

Thank you

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