CASE REPORT

Challenges for the Endocrine Surgeon in the Decision-making in Managing Head and Neck Paragangliomas

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Abstract

Head and neck paraganglioma tumors present with a lot of challenges in decision-making for choosing the most appropriate management option for particular patients. We present a young patient who is a 22-year-old lady with neck mass for 7 years duration with a gradual increase. There was a significant family history of a sister dying at 18 years of age due to an unknown cause. She was normotensive, nonfunctional, and had a large neck mass reaching up to the base of the skull. As it was considered inoperable, we explored other treatment options and gave her radiation therapy. The tumor mass reduced 60% in 8 months duration. We have kept her in close follow-up with repeat imaging at 6 months intervals.

Conclusion: We need to a tailor-made treatment option for each patient with neck paraganglioma and chose wisely among surgery with or without preoperative embolization and radiation therapy.

Keywords: Endocrine, Paraganglioma, Succinate dehydrogenase.

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Case Description

A 22-year-old lady presented in our outpatient clinic with a history of (h/o) gradually progressive swelling on the right lateral aspect of the neck since the past 7 years, associated with continuous, dull aching nonradiating pain. There was no other significant history related to the swelling. She was normotensive.

She had h/o sputum-positive pulmonary tuberculosis at the age of 6 years with reactivation 2 years back, managed with antitubercular treatment (ATT). There was no h/o change in the size of the neck swelling after completion of ATT.

In family history, there was h/o sudden death of her younger sister at 18 years of age (exact cause of death not known).

On examination, she had 8 × 9 cm swelling in the right lateral aspect of the neck, just below the angle of the mandible, along with the upper and middle third of the anterior border of the right sternocleidomastoid muscle (Fig. 1). The upper and lateral borders of the swelling were not well defined, firm in consistency, with restricted mobility, slight tenderness on palpation with transmitted pulsations, and no bruit on auscultation.

Contrast-enhanced computed tomography (CECT) with angiogram of the neck, revealed a well-defined, intensely enhancing lobulated soft tissue lesion 50 × 28 × 70 mm in the right carotid space, extending from the base of the skull adjacent to the jugular foramen and foramen magnum, extending up to the cricoid cartilage, deep to the sternocleidomastoid muscle. The lesion was partially encasing and splaying the external and internal carotid artery and inferiorly indenting the common carotid artery. The right internal jugular vein seems to be infiltrated by the mass. The mass received multiple arterial feeders from the right external carotid artery (Fig. 2).

Fine needle aspiration cytology revealed follicular pattern and spindling of cells, with a round to oval nuclei with nuclear pleomorphism, speckled chromatin with indistinct cell borders with few eosinophilic granules suggestive of malignant neoplasm, likely paraganglioma. Biochemical workup revealed normal 24 hours urinary metanephrines and nor-metanephrines. Due to financial constraints, we could not do the genetic workup for this patient. However, we would have preferred to do succinyl dehydrogenase (SDH) mutations for her.

As the mass was involving the base of the skull and encasing the carotids, it was considered inoperable. Hence, we explored other treatment options for her. The patient was managed with intensity-modulated radiotherapy, 59.3 Gy, delivered in 33 fractions. The patient was under continuous follow-up and after 8 months postradiotherapy, her swelling appreciably reduced in size to approximately 4 × 4 cm (Fig. 3) and became soft to firm in consistency. Repeat CECT reveals a mass of size 3.9 × 3 × 3.8 cm, abutting and partially encasing the right internal carotid artery with focal indistinct fat planes, also encasing the origin of the right external carotid artery and its branches, without the involvement of the base of the skull (Fig. 4).

In view of significant response to radiation, we as clinicians planned to keep her under observation by 6 monthly follow-up and annual imaging of the neck. Surgery is associated with a lot of major morbidities like stroke, nerve injuries, bleeding, salivary gland injury, and wound-related issues postradiation. Hence, radiation therapy (RT) alone is useful in such situations and surgery is reserved for a select group of patients where the tumor is resectable.

Discussion

Paragangliomas are highly vascular and mostly benign tumors. These tumors originate from the neural crest cells in the

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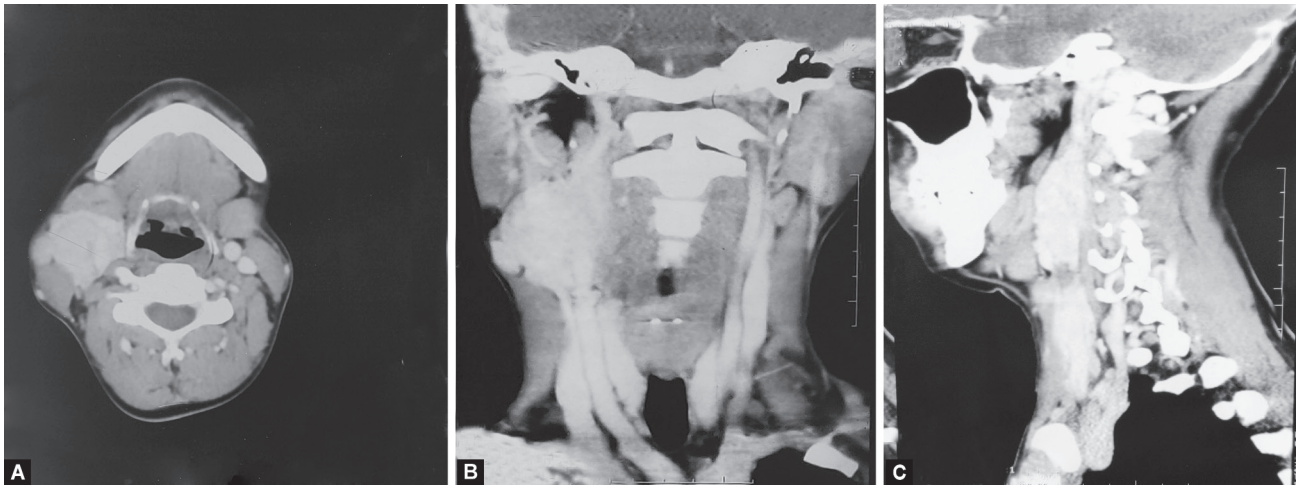
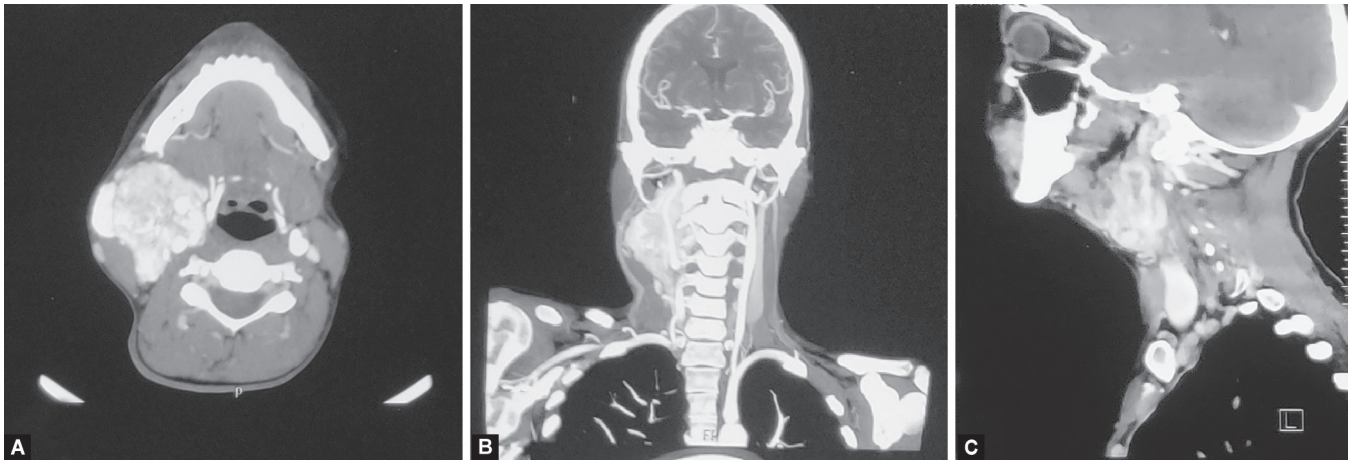
Figs 1A and B: A swelling of 8 × 9 cm in the right lateral aspect of neck, just below the angle of mandible, along the upper and middle third of sternocleidomastoid



Figs 2A to C: CECT (axial, coronal and sagittal sections) of the neck showing an intensely enhancing lobulated soft tissue lesion 50 × 28 × 70 mm, extending up to the base of the skull, partially encasing and splaying the external and internal carotid artery

Figs 3A and B: Post-radiotherapy clinical picture showing significant reduction in the size of the swelling (4 × 4 cm)

sympathetic or parasympathetic chain. In head and neck regions, mostly these tumors are nonfunctional and originate from the parasympathetic chain in the vagus, carotid or jugular bulb, and tympanic plexus (Fig. 5). Usually, patients present with a neck mass and may have a family history of paragangliomas or multicentric tumors. Multicentricity is present in up to 85% of patients having genetic predispositions and up to 10% in sporadic cases.[1] In carotid body tumors, 20% of the times there may be a second tumor on



Figs 4A to C: Post-radiotherapy CECT (axial, coronal and sagittal sections) showing a mass of 3.9 × 3 × 3.8 cm, partially encasing the external and internal carotid artery, without involvement of the base of the skull

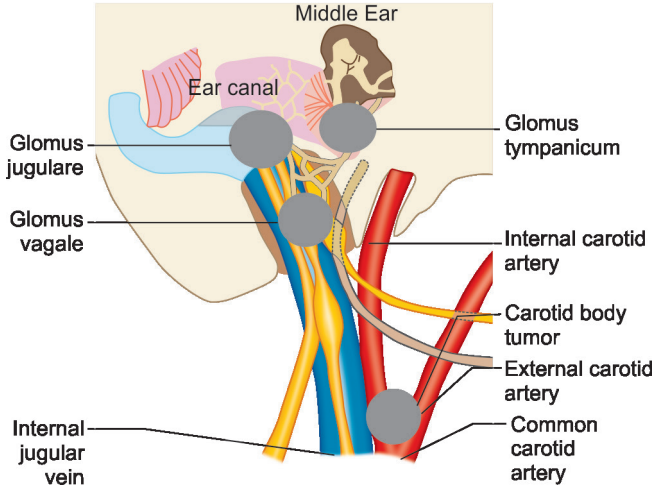


Fig. 5: Various locations at which head and neck paraganglioma massed may arise

the other side of the neck.[2] These tumors can be synchronous or metachronous. Hence, lifelong follow-up in these patients is recommended.

Rarely these tumors may be malignant and may be associated with lymph nodal metastases or distant metastases. In patients having SDH-B mutations, there may be up to 30 to 70% rate of malignant tumors.[3] Mostly, orbital or laryngeal paragangliomas are associated with malignancy.

Workup includes imaging in form of CECT angiography of the head and neck regions or MRI. In bilateral tumors at the carotid bulb region, the patency of the circle of Willis also has to be checked. CECT shows characteristic patterns of the tumor displacing the internal and external carotid arteries as seen in our patient also. Biochemical workup includes 24 hours urinary metanephrine and nor-metanephrine levels to rule out the functional tumor and any associated abdominal pheochromocytomas. In jugular-tympanic tumors, an otoscopic examination is needed as patients may present with tinnitus and hearing loss.

Treatment options include surgery, RT, or embolization.[4] Surgery is recommended in small to moderate size tumors that are resectable. In patients having bilateral tumors in the neck, both tumors need to be resected. However, postbilateral tumor excisions, there may be a risk of baroreceptor function loss and deficits in the cranial nerves resulting in labile hypertension. In selected patients, embolization may be done to reduce vascularity and surgery has to be planned within 48 hours of the embolization procedure.

RT is indicated in large unresectable tumors extending in the base of the skull, in elderly or debilitated patients. RT is associated with low morbidity and usually high tumor response rates.

Hence, treatment strategy has to be planned on an individual basis for each patient and tumor characteristics. In our patient, RT helped in reducing the tumor mass and we could avoid surgery and surgery-related major morbidities also. However, we have kept our patient in close follow-up and may plan surgery whenever indicated.

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