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VAGAL PARAGANGLIOMA – A CASE REPORT ILLUSTRATING COMPLETE IMAGING WORK-UP IN A RARE PATHOLOGY.

Radiodiagnosis

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ABSTRACT

Vagal paragangliomas are rare tumors . Most of them are non-functional and present with an asymptomatic masses, few may present with hoarseness of voice, mild pain and discomfort. Few may be functional and may be associated with hypertension and tachycardia. Timely treatment is based on early diagnosis. Imaging plays a critical role in diagnosis with CT and MRI angiography being the investigations of choice with characteristic findings. In this case report we present a case of Vagal paraganglioma with complete imaging workup

KEYWORDS

INTRODUCTION

Vagal Paragangliomas are rare tumors arising from paraganglionic cells in the vagal nerve perineurium just below or at the level of ganglion nodosum(1). They comprise only 3% of all head and neck paragangliomas(2). These have an incidence much less as compared to carotid body tumors and other glomus tumors(3). Common clinical presentation is usually as swelling in the neck and hoarseness of voice. However uncommonly, vagal paragangliomas may present with hypertension, tachycardia, flushing, as they contain epinephrine and norepinephrine precursors. Lying between the jugular vein and internal carotid artery, they displace the artery medially. They do not splay the carotid bifurcation which is a feature seen with carotid body tumors(4,5). In this case report we present complete imaging work-up in a case of non-functioning Glomus vagale in a 16 year old female. She presented with a lump in the posterolateral neck and was diagnosed on ultrasound, further characterized with CT angiography and MRI , treated with embolization followed by surgical resection.

Case Report :

A 16 year old patient presented with a swelling in posterolateral neck since a year. It was associated with mild pain and discomfort. She did not have any hypertension, tachyarrythmias or flushing. Swelling did not subside with antibiotics but rather increased in size. On examination there was a deep seated fusiform swelling in the right upper lateral neck along the vertical length of the sternocleidomastoid muscle. It was approximately 2x3 cm in size. It was non-tender and firm in consistency. The skin above the swelling showed no changes. There was no associated neck lymphadenopathy.

B mode ultrasound evaluation revealed a hypo to isoechoic, well defined oblong lesion. On colour doppler evaluation the internal carotid artery and jugular vein were seen splayed around the lesion. The lesion itself also appeared vascular(Fig.1).

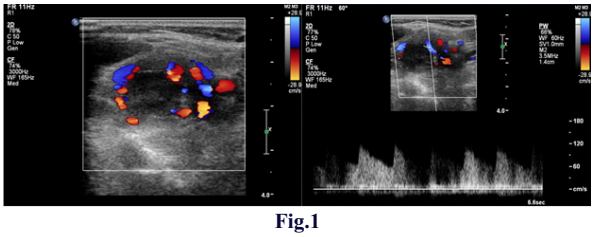


Fig.1

A contrast enhanced CT angiogram was performed. It revealed an intensely enhancing mass extending into the carotid space. The mass splayed the internal carotid artery medially and the internal Jugular vein along its lateral margin. (Fig.2)

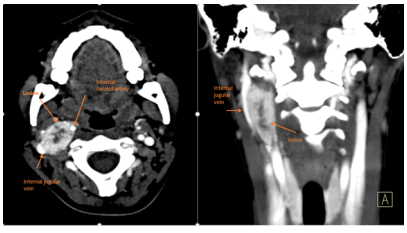


Fig.2 - Contrast enhanced CT angiography

MRI scan was also performed for better characterisation of the lesion and to rule out possibility of other paragangliomas. 3 Tesla GE 750 W wide bore scanner was used. MRI was performed using T2, T1 , T2 fat sat and DWI sequences in multiple planes. High resolution post contrast images were obtained using time resolved imaging of contrast kinetics. A lobulated well-circumscribed, heterogeneous signal intensity mass lesion was identified which extended from above the carotid bifurcation to just below the skull base. It measured 2 x 2.7 x 3.5 cm in anterio-posterior, transverse and supero-inferior extents. Multiple flow voids were observed which gave a positive “salt and pepper” appearance, characteristic of paragangliomas. The location is typical for glomus vagale. The lesion did not splay the carotid bifurcation distinguishing it from carotid-body tumor. (Fig.3)

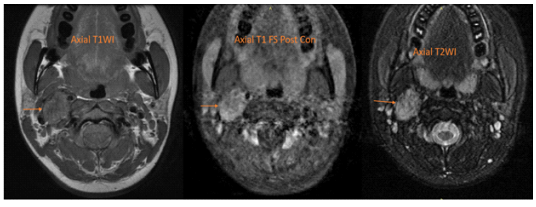


Fig. 3: Axial T1WI, Axial T1 FS post contrast, Axial T2 WI.

Magnetic Resonance Angiography demonstrated the presence of multiple vascular twigs within the lesion.(Fig 4a)

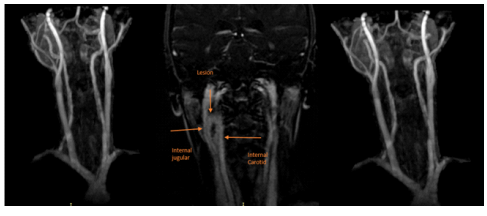


Fig. 4a- MR angiography

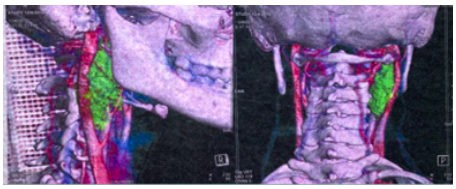
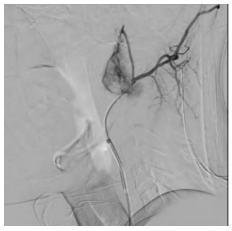


Image 4b- 3D CT angiogram reconstruction

In view of intense vascularity DSA and endovascular embolization was performed before surgical excision. On embolization, the feeders from ascending pharyngeal artery were embolized.(Fig 5a and 5b)



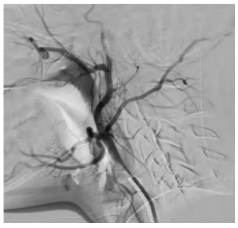


Image 5a- Pre op embolization – feeder from ascending pharyngeal artery. **Image 5b – post embolization.**

DISCUSSION:

Paragangliomas are neoplasms that arise from chemoreceptor paraganglial tissues, distributed throughout the body. 90 % arise in the adrenals and 3% arise in the neck(5). Paragangliomas in the head and neck can be found in four primary locations which include the jugular bulb, the middle ear cavity, the vagus nerve and the carotid body. The majority of paragangliomas are benign and malignant behavior is seen in 10% of cases(6). The male to female ratio is 2:1 with a mean age of diagnosis 40 years (7). In our case however the age was 16 years which is an unusual age of presentation.

Vagal paragangliomas are rare tumors that originate in retro-styloid portion of the parapharyngeal space. The closest differential is carotid body tumor which is differentiated by its hallmark feature – of splaying the carotid vessels. Presentation is common as an asymptomatic swelling at the angle of mandible (8). It may present as hoarseness, vocal cord paralysis and horner's syndrome owing to its relationship with nerves at the skull base (9).

Imaging establishes the diagnosis in almost all of the cases. On ultrasound, it is seen as a solid heterogeneously hypoechoic lesion comprising of small vascular structures. However, it can not be differentiated from other potential lesions in this location on ultrasound alone. Contrast-enhanced cross-sectional imaging with angiography is the investigation of choice. On contrast-enhanced CT, the lesion is seen as an intensely enhancing mass which splays the internal carotid artery and internal jugular vein. Similar appearance was seen in our case, which suggested a diagnosis of vagal paraganglioma. However, CT has limitations in imaging skull base lesions.

MRI plays the most significant role in the diagnosis of head and neck paragangliomas. These tumors appear hyperintense on T2WI and show contrast enhancement on T1WI. On T1WI these tumors show ovoid flow voids within high signal giving a salt and pepper appearance. The flow voids represent vascular structures within a hyperintense matrix, this appearance may sometimes be seen on T2WI. MRI also helps to delineate the extent of the tumor and its relationship to carotid vessels. Furthermore, MR angiography helps to rule out other paragangliomas in the head and neck (10). On MR Angiography splaying of the internal carotid and internal jugular vein is seen. In our case the lesion shows intense enhancement on post-contrast images, MR angiography showed anteromedial displacement of the internal Carotid Artery and posterolateral displacement of the internal Jugular Vein.

Some larger tumors need pre-embolization before surgery to decrease the vascularity of tumor. Digital subtraction angiography provides an arterial map and endovascular access for embolization of tumor (11). In our case tumor flush was visualized supplied by a vascular twig from the ascending pharyngeal artery. Post embolization the tumor vascularity was significantly reduced.

The differential diagnosis of Vagal Paragangliomas include, (i) Carotid body tumor: it splays the internal and external carotid arteries unlike a Vagal paraganglioma; (ii) Vagal Schwannoma which is a fusiform mass in carotid space, less vascular in comparison to paraganglioma with no increase in the number of feeding vessels seen on angiography; (iii) Meningioma: Arises from the jugular foramen and is less vascular than paraganglioma; (iv) Glomus jugulotympanicum which is epicentred at the jugular foramen.

CONCLUSION:

Vagal paragangliomas are rare tumors . Most of them are non-functional and present with an asymptomatic masses, few may present with hoarseness of voice, mild pain and discomfort. Few may be functional and may be associated with hypertension and tachycardia. Timely treatment is based on early diagnosis. They are highly vascular lesions and present in close relation with the neck vessels. Imaging plays a critical role in diagnosis with CT and MRI angiography being the investigations of choice with characteristic findings. Endovascular embolization plays key role in treatment plan to limit intraoperative hemorrhage.

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