Neurosciences Quiz

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Elderly male with progressive dysphagia and ataxia

Case Presentation

A 63-year-old male presenting with progressive dysphagia, ataxia, hoarseness of voice, and right sided deafness.

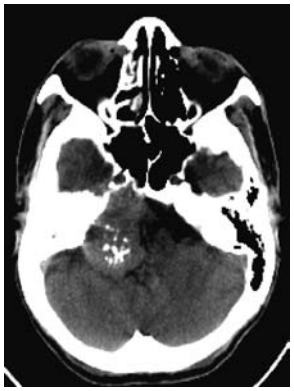

Figure 1 - Axial cut, CT (brain window) through posterior fossa.

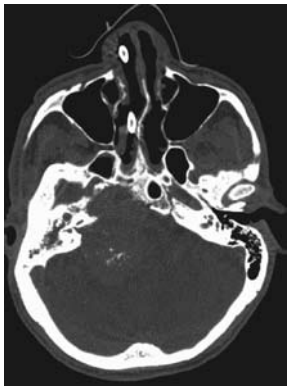

Figure 2 - Axial cut, CT (bone window) through posterior fossa.

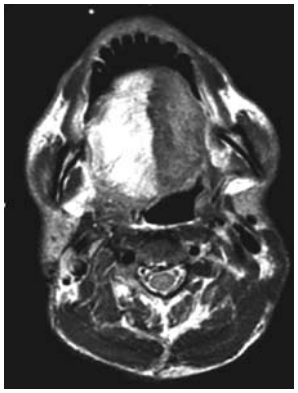

Figure 3 - Axial cut, T2 weighted MRI at the mandible level.

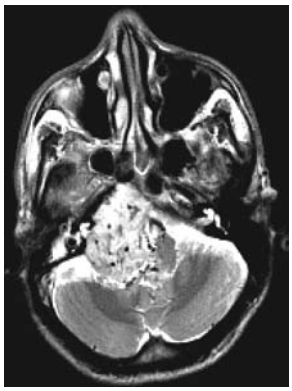

Figure 4 - Axial cut, T2 weighted MRI internal auditory canal level.

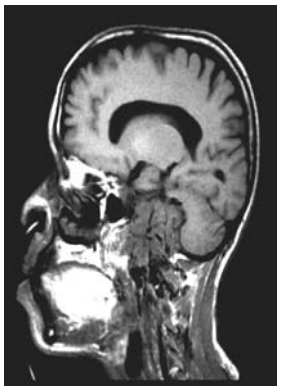

Figure 5 - Sagittal cut, T1 weighted MRI.

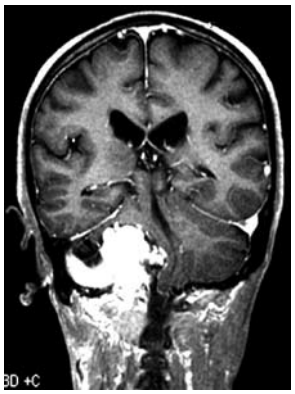

Figure 6 - Coronal cut, post contrast T1 weighted MRI.

Questions

1. What are the radiographic abnormalities?
2. What is the differential diagnosis?
3. What is the diagnosis?

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Radiological findings

Figure 1 - Axial cut; CT demonstrates an extra-axial isodense to hyperdense lesion at the right cerebello-pontine angel (CPA) region with dense calcification and boney erosion of the petrus apex and clivus.

Figure 2 - Axial cut; CT demonstrates boney erosion of the right petrus apex and internal carotid artery canal.

Figure 3 - Axial cut; T2 weighted MRI demonstrates T2 hyperintense and hypertrophied right side of the tongue. Asymmetry of the oropharynx.

Figure 4 - Axial cut; T2 weighted MRI demonstrates right CPA extra-axial heterogenous T2 hyperintense lesion with multiple tiny foci of signal voids.

Figure 5 - Sagittal cut; T1 weighted MRI. Heterogenous T1 hypointense lesion with multiple tiny foci of signal voids.

Figure 6 - Coronal cut; Post contrast T1 weighted MRI. Homogenous and intense enhancement.

Differential diagnosis: Glomus jugulare, jugular schwannoma, meningioma, chordoma, chondrosarcoma, bony metastases (for example renal cell carcinoma, thyroid cancer).

Diagnosis: Glomus jugulare

Discussion

Glomus jugulare tumors are rare, slow-growing, hypervascular tumors that arise within the jugular foramen of the temporal bone. They are included in a group of tumors referred to as paragangliomas, which are defined according to location (namely, origin at the jugular foramen) rather than an anatomic origin, and may arise from Jacobson's or Arnold's nerves of the jugular bulb.[1,2] Other locations include carotid body, glomus vagale, and glomus tympanicum tumors. Although glomus jugulare is a rare tumor, it is the most common of the jugular fossa tumors. These tumors are seen in adults, typically between 40 and 60 years of age, with a moderate female predilection.[1,2] Due to the insidious onset of symptoms, these tumors often go unnoticed, and delay in diagnosis is frequent. Clinical presentation depends on the degree of middle ear involvement. When significant involvement is present then pulsatile tinnitus and hearing loss can occur.[2] Additionally, a number of patterns of cranial nerve palsies have been described. These include: Vernet syndrome (motor paralysis of cranial nerves IX, X, and XI), Collet-Sicard syndrome (Vernet syndrome plus involvement of cranial nerve XII) and Horner syndrome.[1-3] The CT is most useful at assessing the integrity of the bone and ear structures.[1] The MRI is the best modality to characterize this lesion. Usually, the mass is hypointense in T1 and hypointense in T2 and it demonstrates marked and intense enhancement T1 C+ (Gad).[1-3] A salt and pepper appearance is seen on both T1 and T2 weighted sequences; the salt representing blood products from hemorrhage or slow flow and the pepper representing flow voids due to high vascularity. It should be noted that this appearance is sometimes encountered in other lesions (for example, hypervascular metastases) and is not typically seen in smaller glomus tumors.[1] Angiography demonstrates intense tumor blush, with the most common feeding vessel being the ascending pharyngeal.[1] Early draining veins are also noted due to intra-tumoral shunting.[3] Angiography also has a role to play in preoperative embolization, which is typically carried out 1-2 days prior to surgery. An isotope scan demonstrates positive Indium-111 labelled octreotide. Surgery is the treatment of choice and if complete resection is achieved, a cure can be expected. Complications are however, not uncommon due to the large number of sensitive structures in the region and include: cranial nerve deficits and CSF/endolymphatic leak.[2] Recurrence and local invasion is common, occurring in 40-50% of cases.[1] Malignant transformation is less common, seen in 2-13% of cases.[1,2] Although most cervical paragangliomas (for example, carotid body tumors) are considered relatively radioresistant, base of skull paragangliomas are radiosensitive, and thus large inoperable tumors or tumors in elderly and frail patients are often treated with radiotherapy.

References

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2. Mafee MF, Raofi B, Kumar A, Muscato C. Glomus faciale, glomus jugulare, glomus tympanicum, glomus vagale, carotid body tumors, and simulating lesions. Role of MR imaging. ***Radiol Clin North Am*** 2000; 38: 1059-1076.
3. Som PM, Curtin HD, editors. Head and neck imaging. St Louis (MO): Mosby-Year Book; 2003.