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Rare presentations of paragangliomas

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Background: Paragangliomas are benign, hypervascular neoplasms accounting for 0.012% of all tumors and 0.6% of all neoplasms in the head and neck region. These neoplasms are insidious in onset, making the diagnosis more challenging. In this report, we have presented three unique cases of paragangliomas.

Case Report: Case 1: A 65 year-old woman presented with hoarseness and hearing loss of two months duration. On physical examination, she was found to have left cranial nerve VIII, IX and X paralysis. Laryngoscopy also revealed left vocal cord paralysis. MRI of the brain was consistent with Glomus Vagale Tumor. Pheochromocytoma work up was negative. She was successfully treated with IR guided embolization and surgical debulking of the tumor.

Case 2: A 31 year-old woman with a history of recurrent left ear canal polyps of four year duration, presented with bloody otorrhea and pulsatile tinnitus. MRI showed a left middle ear mass extending through the jugular foramen consistent with Glomus Jugulotympanicum Tumor. She underwent IR guided embolization followed by surgical debulking of the tumor with complete resolution of the symptoms.

Case 3: A 50 year-old woman with a history of pulsatile tinnitus in the left ear, presented with left sided facial numbness. MRI revealed a mass which on biopsy showed Glomus Jugulare Tumor. Her symptoms resolved after embolization and surgical debulking. However, tumor relapsed requiring radiotherapy.

Discussion: Paragangliomas are rare, slow growing tumors, arising from paraganglionic tissue of neural crest origin. They usually presents between the 5th and 6th decade of life with a female predominance. Common presentations include a mass in the middle ear (Glomus Tympanicum) causing pulsatile tinnitus and conductive hearing loss, or as a pulsatile painless mass in the neck (Carotid Body Paraganglioma). Our second case was diagnosed with Paraganglioma in the second decade of life, which is very unusual. She presented with recurrent external ear polyps which is a unique finding. In 2.5% of cases, paragangliomas present with hoarseness and vocal cord paralysis (Glomus Vagale) as mentioned in our first case. The unique feature of the third patient, unlike the first two, was the recurrence of the tumor after resection reaching the same initial size in 7 months. This suggests the possibility of a malignant Paraganglioma, which occurs only in 1-5% of Paragangliomas.

Conclusion: We present a broad spectrum of clinical presentations of this extremely rare tumor. These presentations include dysphagia, hearing loss, pulsatile tinnitus, hoarseness, pain and cranial nerve palsies. A high index of suspicion and clinical awareness could help in early diagnosis and with improved outcomes in the treatment of paragangliomas.

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