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Familial paraganglioma syndrome

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Case summary

A 54-year-old woman presented with poorly controlled hypertension. Abdominal CT revealed bilateral adrenal hypervascular tumors (Fig. 1). Adrenalectomy was performed and pathology showed pheochromocytoma. After surgery, blood pressure was under control. Four years later, she had left-sided hearing loss. Brain MRI showed a left glomus jugulare (with enhancing tumor and signal voids at the left jugular foramen) and left cerebellopontine angle tumor (Fig. 2). The final diagnosis was paraganglioma syndrome, possibly of familial origin, which combines glomus tumor and pheochromocytoma (1, 2). Familial paraganglioma syndromes can be caused by mutations in the catalytic subunits of succinate dehydrogenase, also known as complex II of the mitochondrial respiratory chain (1, 2).

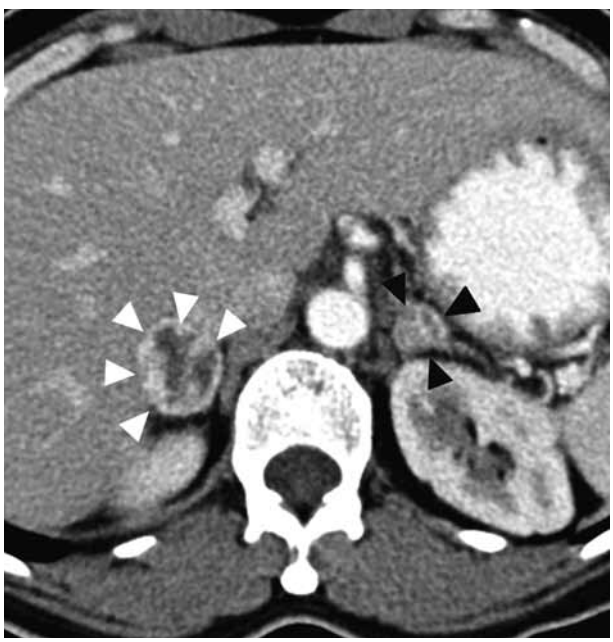

FIG. 1. — CT of the abdomen showing hypervascular nodules in the adrenal glands.

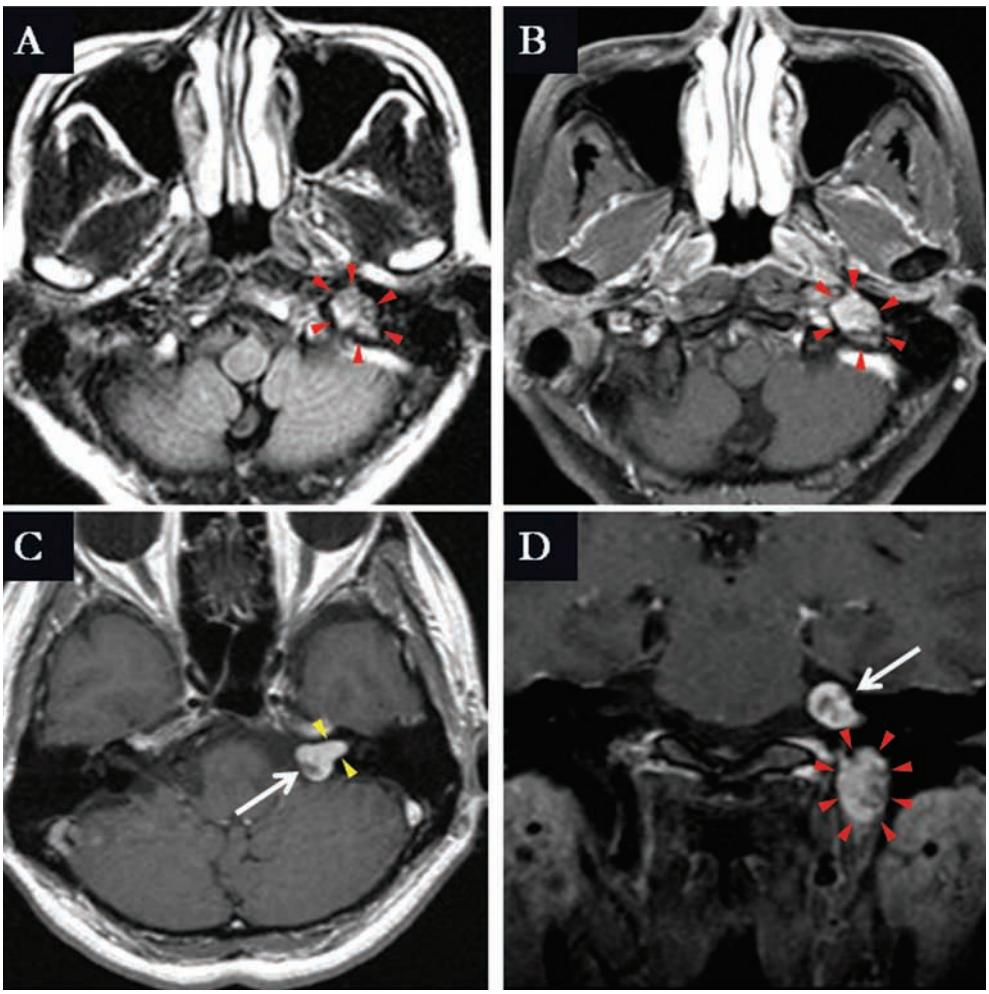

FIG. 2. — MRI of the brain with contrast showing a hypervascular mass at the left foramen jugulare (red arrowhead), and another enhancing mass at the left cerebellopontine angle (white arrow) with extension into the left internal auditory canal (yellow arrowhead).

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