Intrasellar and suprasellar paraganglioma: CT and MR findings.

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Intrasellar and Suprasellar Paraganglioma: CT and MR Findings

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Summary: We present the CT and MR findings of an intrasellar and suprasellar paraganglioma in a 17-year-old girl. Paragangliomas in this location are rare. The lesion had several imaging features characteristic of paragangliomas including marked enhancement after contrast and multiple vascular flow voids.

Index terms: Paraganglioma; Sella turcica, computed tomography; Sella turcica, magnetic resonance; Sella turcica, neoplasms

Case Report

A 17-year-old white girl was first evaluated 5 years before admission after failing visual screening in school. An ophthalmology examination revealed decreased visual acuity in both eyes and bitemporal hemianopsia. The patient noticed difficulty in reading the board at school and also complained of occasional headaches. A computed tomographic (CT) scan at that time reportedly revealed an intrasellar and suprasellar mass with some calcification. Magnetic resonance (MR) was reported to have demonstrated vessels within the lesion. A physical examination was remarkable only for vertical nystagmus. An arteriogram reportedly showed a highly vascular sellar and suprasellar tumor. A right subfrontal approach was used to explore the lesion, but because of excessive bleeding from the tumor, only a biopsy was accomplished. The pathologic diagnosis was paraganglioma.

The patient was referred to our institution 4 years later at age 17 after 4 months of increasing visual complaints and severe intermittent frontal headaches. A neurologic examination showed that she had visual acuity of 20/40 bilaterally and a complete bitemporal hemianopsia with macular involvement. There were no other focal neurologic deficits. Endocrine testing revealed elevated 3,5,3′-triiodothyronine and thyroxine levels and decreased thyroid-stimulating hormone levels. Luteinizing hormone–releasing hormone provocative testing showed a prepubertal gonadotropin response. Her bone age was 15, which was less than two standard deviations from the mean. An unenhanced CT revealed a large, homogenous mass within the sella and suprasellar cistern with a focal hyperdensity possibly representing calcification at its apex. After contrast administration, the mass enhanced intensely and homogeneously. The sellar floor was thinned and remodeled, and neither the infundibulum nor the optic chiasm could be identified (Fig 1A). The third and lateral ventricles were mildly enlarged. MR was performed on a GE 1.5-T magnet (General Electric, Milwaukee, WI). The mass was slightly hyperintense to white matter on short spin-echo (500/25/2) (repetition time/echo time/excitations) images and contained multiple linear and punctate signal voids (Fig 1B). Some of the linear flow voids had a branching pattern. Flow was confirmed in these areas by the use of gradient-echo techniques (Fig 1C). The lesion was hyperintense to gray matter on proton-density images (2700/30) and isointense to gray matter on long spin-echo images (2700/90) (Fig 1D and E). Diffuse enhancement was demonstrated after gadolinium administration (Fig 1F and G). The mass touched the superior margin of the left cavernous carotid artery, but narrowing of the vessel was not present.

The tumor was resected through a subfrontal approach. It was encapsulated and highly vascular.

Multiple fragments of soft, reddish-brown tissue were available for histologic analysis. Microscopically, the neoplasm was composed of lobulated clusters of cells, suggestive of a "zellballen" pattern, occasionally surrounded by a fine capillary network (Fig 2). Numerous slitlike vascular spaces were present. Nuclear pleomorphism was more prominent in our specimen as compared with the slides from the original biopsy. Mitoses were rare, and necrosis was absent. Most cells stained positively with chromogranin. The histopathologic features were similar to those obtained from the previous biopsy. An ultrastructural examination revealed numerous dense core granules and junctional complexes. The ultrastructural findings and the positive chromogranin stain are characteristic of neoplasms of neural crest origin and support a histopathologic diagnosis of paraganglioma.

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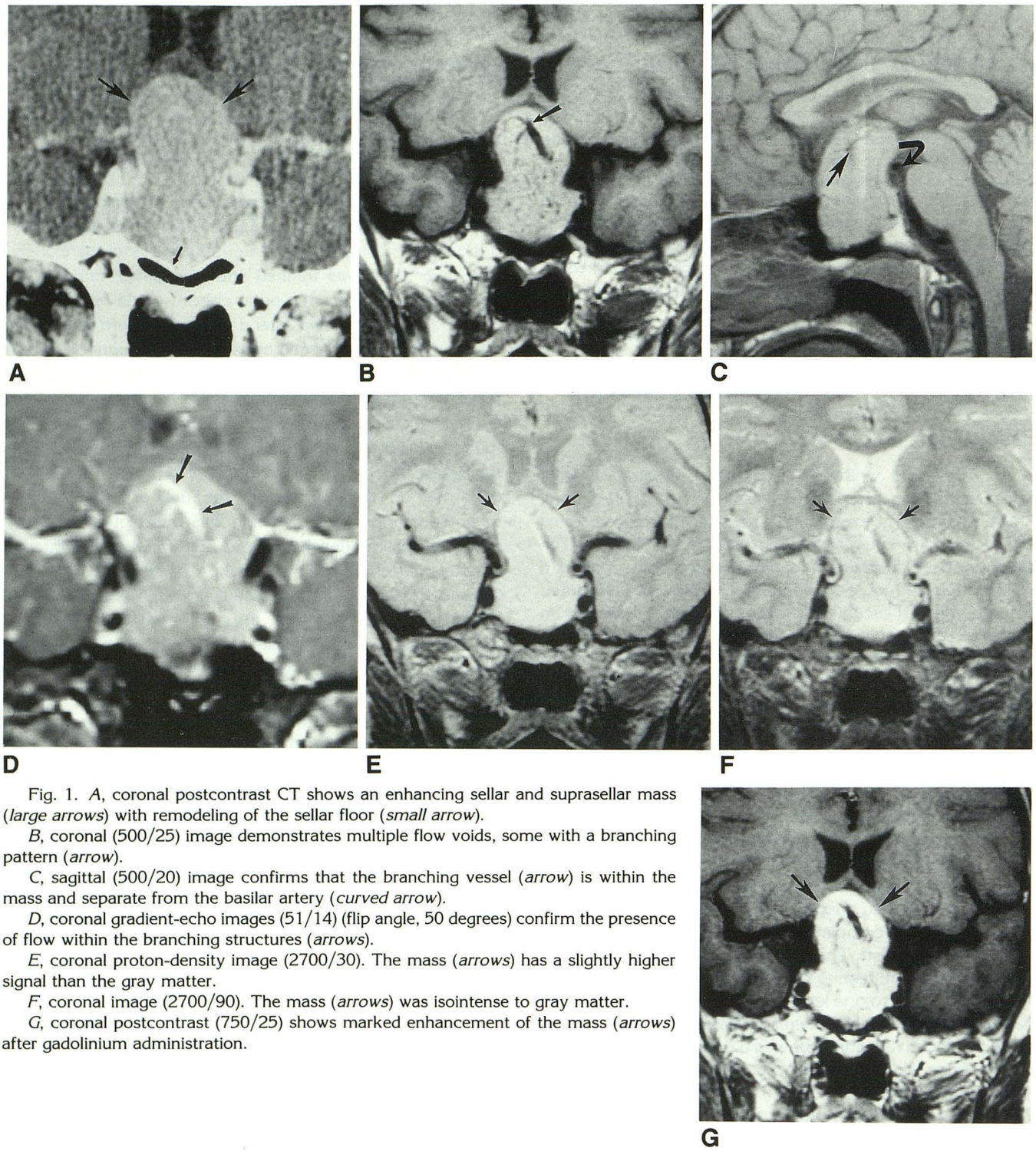

Fig. 1. *A*, coronal postcontrast CT shows an enhancing sellar and suprasellar mass (*large arrows*) with remodeling of the sellar floor (*small arrow*).

B, coronal (500/25) image demonstrates multiple flow voids, some with a branching pattern (*arrow*).

C, sagittal (500/20) image confirms that the branching vessel (*arrow*) is within the mass and separate from the basilar artery (*curved arrow*).

D, coronal gradient-echo images (51/14) (flip angle, 50 degrees) confirm the presence of flow within the branching structures (*arrows*).

E, coronal proton-density image (2700/30). The mass (*arrows*) has a slightly higher signal than the gray matter.

F, coronal image (2700/90). The mass (*arrows*) was isointense to gray matter.

G, coronal postcontrast (750/25) shows marked enhancement of the mass (*arrows*) after gadolinium administration.

Discussion

Paragangliomas are highly vascular, usually benign tumors that are derived from paraganglion cells of neural crest origin. They most typically arise at the carotid bifurcation, the jugular bulb, and the middle ear and from the perineurium of the vagus nerve below the skull base. They have been termed, respectively, carotid body, glomus jugulare, glomus tympanicum, and glomus vagale tumors. The CT and MR appearances of paragan-

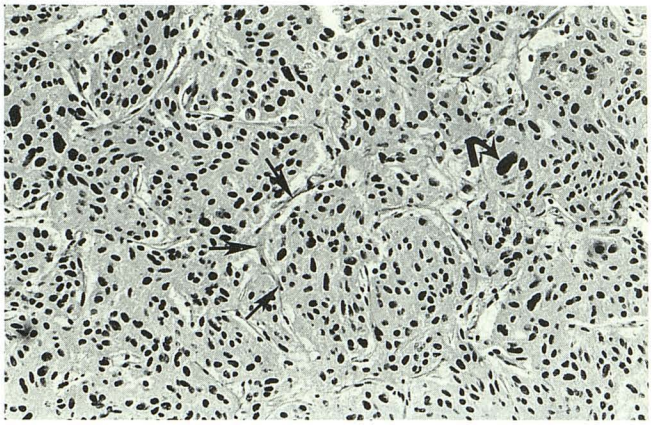

Fig. 2. Photomicrograph (light microscopic photograph) showing zellballen (*straight arrows*) and nuclear pleomorphism (*curved arrow*). This is characteristic of paraganglioma (hematoxylin and eosin, ×110).

gliomas in these locations have been well described (1–4). Suprasellar paragangliomas are unusual, although they have been mentioned in the pathology literature (5, 6).

"Paragangliomas of uncertain cell of origin" occur in the sella, cauda equina, pineal gland, duodenum, and uterine endometrium (4). Bilbao et al have suggested that neural crest or paraganglionic tissue may be included in the developing adenohypophysis, on the basis of avian embryo pituitary studies (5). By light microscopy, a lobulated pattern of cell clusters termed zellballen is typical of paragangliomas, as is prominent vascularity (4) (Fig 2).

The vascular flow voids and intense enhancement seen in this case are typical of paragangliomas in more conventional locations. Meningiomas and choristomas are other highly vascular tumors that occur in the sella and suprasellar region and might be confused with this lesion (7). Pituitary adenoma or craniopharyngioma could have a similar appearance to the tumor described in this case, but the multiple flow voids indicate marked vascularity, which would be unusual for these lesions. A vascular metastasis is also a possibility. On the CT scan, there was a small, peripheral hyperdensity within the mass. This was initially interpreted as calcification, but no calcium was detected on histologic sections. This is interesting, because calcification within paragangliomas is an uncommon finding. The evidence of multiple flow voids was the most important feature of this tumor because it alerted the surgeon to the highly vascular nature of this tumor preoperatively.

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