Unusual locations of hemangioblastomas

Case illustration

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We present a case of multiple hemangioblastomas of the neuraxis, two of which were found in unusual locations. This 32-year-old woman, with von Hippel–Lindau disease, presented with a few days’ history of nausea and on examination a right cerebellar syndrome. Gadolinium-enhanced T₁-weighted magnetic resonance (MR) imaging demonstrated two right cerebellar nodular cystic hemangioblastomas associated with other nodular lesions of the neuraxis, two of which were of particular interest. The first was located in the intrasellar region, more precisely in the neurohypophysis (Fig. 1 left); the second was located in the tectum of the mesencephalon with exophytic extension into the quadrigeminal cistern (Fig. 1 right). Total resection of the cerebellar hemangioblastomas was performed via a right-sided suboccipital craniotomy, during which the quadrigeminal cistern hemangioblastoma was also resected via a subtentorial supracerebellar approach.

Macroscopically, the quadrigeminal cistern hemangioblastoma had no attachment to the tectum and appeared to arise from the right trochlear nerve, which was encased by the tumor and had to be sectioned to permit total resection of the lesion. Histopathological examination confirmed that the hemangioblastoma had arisen from the trochlear nerve.

We obtained strong radiological and scintigraphic evidence (Fig. 2) suggesting that the intrasellar lesion was also a hemangioblastoma nodule. Only three cases of hemangioblastomas found in this location have been reported.⁴ We chose to observe this lesion because of its asymptomatic nature.

This is the first report of a hemangioblastoma involved with a cranial nerve other than the optic nerve.⁵ The discovery of lesions in such unusual locations confirms that they could arise almost anywhere throughout the central nervous system, although cerebellar and spinal locations are far more common.

References