Pearls & Oysters: Lumbar paraganglioma

Can you see it in the eyes?

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PEARL

- Paragangliomas of the CNS are uncommon and only a few cases of paragangliomas in the lumbar region have been reported in the English literature.^{1,2} Accompanied by clinical signs of raised intracranial pressure, this condition is extremely rare.

OYSTER

- A lumbar tumor with papilledema as the only clinical sign can be easily missed. When there are clues for a lumbar tumor, e.g., history of back pain, MRI of the spine is mandatory.

CASE REPORT A 34-year-old man was referred by his ophthalmologist because of bilateral papilledema and a 2-month history of transient obscurations in his left eye. These obscurations lasted for several minutes and occurred approximately twice a day. The patient denied complaints of headache, nausea, vomiting, lower back pain, or leg weakness. The physical examination was unremarkable. The patient was not obese and had a normal blood pressure. The neurologic examination revealed bilateral papilledema, worse on the left than the right (figure, A), and there was no visual disturbance. The examination was otherwise normal.

Cranial CT and MRI of the head showed no abnormalities. A lumbar puncture was performed in order to obtain CSF and measure intracranial pressure, but was unusually difficult, requiring several attempts before succeeding at level L3-L4. The lumbar puncture revealed an opening pressure of 16 cm H₂O. Several hours after the lumbar puncture the patient started to complain of severe lower back pain radiating toward both legs. An MRI of the lumbosacral spine was performed which revealed an intradural mass at the L5-S1 level, isointense on T1- and T2-weighted images, which filled the entire spinal canal. There was homogenous enhancement after gadolinium (Gd) injection (figure, C and D).

The patient was referred to a neurosurgeon and underwent a laminectomy to gain access to the tumor. After dural opening, a well-encapsulated and vascularized tumor was seen attached to several nerve roots. The tumor was removed completely. Microscopy of the tumor revealed a classic “Zellballen” pattern (figure, E), and the tumor was classified as a paraganglioma WHO grade 1.

Four months after surgery, the visual symptoms and the papilledema had resolved completely (figure, B), and the back pain had resolved.

DISCUSSION The most common presenting symptom in a patient with a lumbar paraganglioma is low backache with radiation toward the legs.³ Papilledema as the presenting symptom is rare, and only 7 prior cases have been described in English literature.^{1,2,4,5} Of these 7 patients, 6 (86%) also had lower back pain, sciatic pain, or weakness of the legs at presentation. Increased intracranial pressure (ICP) and papilledema have long been associated with spinal cord tumors. Ependymomas are responsible for more than 50% of such cases.⁶

The mechanism by which lumbar tumors cause elevated intracranial pressure or papilledema is a subject of discussion. One hypothesis is that elevated CSF protein or recurrent episodes of subarachnoid bleeding cause malabsorption by clotting the CSF outflow canals.^{1,6} Another hypothesis is that a lumbar tumor disturbs the “elastic reservoir” of the lumbar part of the spinal sac, thereby decreasing its ability to maintain constant intracranial pressure.⁶ Interestingly, in the case presented, a normal opening pressure was found, suggesting the presence of a separate pathophysiologic mechanism causing papilledema without increased intracranial pressure. It is also possible that the intracranial pressure dropped after several unsuccessful attempts to perform the lumbar puncture resulting in CSF leakage.

The diagnostic procedure of choice to reveal a lumbar paraganglioma is MRI, although MRI findings are nonspecific. The tumor can be hypointense,

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Figure Ophthalmoscopic, radiologic, and pathologic imaging

(A) Preoperative fundusoscopic images show bilateral papilledema. (B) Postoperative fundusoscopic images show resolved papilledema. (C) MRI of the lumbar spine, sagittal T1 images with gadolinium show homogenous enhancement of the lumbar tumor. (D) MRI of the lumbar spine, sagittal T2 images show slightly hyperintense tumor signal and dilated intradural veins above the tumor. (E) Photomicrographs of a lumbar paraganglioma show monomorphic cells organized in nests (Zellballen) (hematoxylin & eosin, original magnification $\times 20$).

isointense, or hyperintense on T1- and T2-weighted images and gadolinium enhancement may be either present or absent.²

Specific pathologic findings of a lumbar paraganglioma are similar to paragangliomas in other re-

gions. The classic pattern of cells organized in nests (Zellballen) is often not present.⁷

Treatment is total resection of the tumor. If the tumor is resected completely, prognosis is excellent.

Although a paraganglioma has its origin in neuroepithelial cells, functional hormonal activity in lumbar paragangliomas is rare. Only 3 cases have been previously described.² Therefore, no further hormonal evaluation is necessary before tumor resection if there are no symptoms of increased catecholamine release such as high blood pressure, flushing episodes, or tachycardia.

Lumbar paragangliomas presenting with papilledema are extremely rare. If a patient with bilateral papilledema complains of lower back pain, MRI of the lumbar region should be included in the diagnostic evaluation. However, the presented case shows that a spinal tumor may also be considered as a possible cause of papilledema even in the absence of lumbar pain.

AUTHOR CONTRIBUTIONS

Dr. Adriani: drafting/revising the manuscript, acquisition of data. Dr. Stenvers: drafting/revising the manuscript, analysis or interpretation of data, acquisition of data, statistical analysis. Dr. Imanse: drafting/revising the manuscript, study supervision.

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REFERENCES

1. Haslbeck KM, Eberhardt KEW, Nissen U, et al. Intracranial hypertension as a clinical manifestation of cauda equina paraganglioma. *Neurology* 1999;52:1297–1298.
2. Gelabert-Gonzalez M. Paragangliomas of the lumbar region: report of two cases and review of literature. *J Neurosurg Spine* 2005;2:354–365.
3. Singh NG, Sarkar C, Sharma MC, Gaikwad SG, Shashank SK, Mehta VS. Paraganglioma of cauda equina: report of seven cases. *Brain Tumor Pathol* 2005;22:15–20.
4. Bannykh S, Strugar J, Baehring J. Paraganglioma of the lumbar spinal canal. *J Neuro-Oncol* 2005;75:119.
5. Sankhia S, Khan GM. Cauda equina paraganglioma presenting with intracranial hypertension: case report and review of the literature. *Neurol India* 2004;52:243–244.
6. Ghazi NG, Jane JA, Lopes BS, Newman SA. Capillary hemangioma of the cauda equina presenting with radiculopathy and papilledema. *J Neuro-Ophthalmol* 2006;26:98–102.
7. Tischler AS. Pheochromocytoma and extra-adrenal paraganglioma. *Updates Arch Pathol Lab Med* 2008;132:1272–1284.

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