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Intrathecal paragangliomas are an unusual cause of the cauda equina syndrome. Although well documented in the neuropathology and neurosurgery literature, only one case report has appeared in the radiology literature to date [1]. Recognition of the tumor is important because of its biologic characteristics, which differ significantly from ependymoma, the tumor with which it is most commonly confused.

Case Report

A 34-year-old man was initially seen in September 1971 for a 1-year history of intermittent right lower extremity pain. Myelography showed an intradural mass at L2 causing a high-grade obstruction. The CSF protein was elevated. In October 1971, a well-circumscribed tumor, which grossly resembled a neurofibroma or meningioma, was completely excised. Initial pathologic studies indicated that the tumor was an ependymoma. The patient had an unremarkable postoperative course and remained asymptomatic until the summer of 1987 when he began experiencing low back pain radiating into a right S1 dermatome distribution. MR images obtained on a 1.5-T system* showed an intradural lesion separate from the conus but involving the filum terminale. On T1-weighted images, the lesion's signal was approximately equal to that of the conus (Fig. 1A). On T2-weighted images (Fig. 1B), the signal was slightly heterogenous and more intense than CSF. A subsequent myelogram and postmyelogram CT confirmed a high-grade obstruction at the L2–L3 interspace. No significant bony erosion was present.

In September 1987, the patient underwent repeat lumbar laminectomy. Extensive intradural tumor and scar were found enveloping the entire cauda equina and adjacent dura (Fig. 1C). Only incomplete tumor excision with duraplasty could be performed. Postoperative radiation therapy was given.

Microscopic examination of the tumor revealed histologic features of a paraganglioma. The tumor cells had a regular appearance, with round to oval nuclei, few mitoses, and little pleomorphism. The cytoplasm was abundant and eosinophilic with a granular appearance. Vascular trabecular structures segregated the tumor into nests of cells (Zellballen) (Fig. 1D). Electron microscopy showed variable numbers of dense core neurosecretory granules within the cytoplasm. No perivascular pseudorosettes or central canals were seen, and immunocytochemical staining for glial fibrillary acidic protein was negative, thus excluding the diagnosis of ependymoma. Review of the original tumor histology showed characteristics identical to those of the present tumor.

Discussion

Paragangliomas are tumors of the accessory organs of the peripheral nervous system or paraganglia [2]. The adrenal medulla is itself a paraganglion and tumors of it are pheochromocytomas. Extraadrenal paraganglion tumors occur most often in the carotid body, glomus jugulare, mediastinum, and paraaortic regions. CNS locations include the petrous ridge, pineal region, and sella turcica. Although often regarded as benign, paragangliomas of the retroperitoneum and larynx have high morbidity and eventual mortality [3]. Those tumors occurring in their usual branchiomeric location may be locally invasive. Malignancy is found in 6.5% of all extraadrenal paragangliomas. Local tumor recurrence is described in approximately 12%, with late recurrences even after 30 years. Distant metastases are rare, but the tumor may be multifocal in a small percentage of cases [2].

Paragangliomas of the cauda equina were initially described in 1970 [4]. Only 53 cases have been reported to date. There is a slight male predominance, and the mean age at diagnosis is 46 [5]. The most common presentation is low back pain with associated sciatica. Motor or sensory deficits in the lower extremities, as well as bowel or bladder symptoms, have also been reported [5]. These are typical for cauda equina masses of any type. Frequently findings of neuropeptide production, notably 5HT and somatostatin are present. However, symptoms of 5HT hypersecretion are rare. Only one patient had been reported with a history of flush attacks in association with elevated urinary norepinephrine levels and a paraganglioma at this location [2].

These lesions may arise from paraganglia located in the cauda equina [6]. Gross descriptions stress the vascular encapsulated nature of the tumor. Both cystic and multinodular components have been identified [5]. Because of similar histologic features, the tumor is usually confused with an ependymoma. Indeed the first reported case of a cauda

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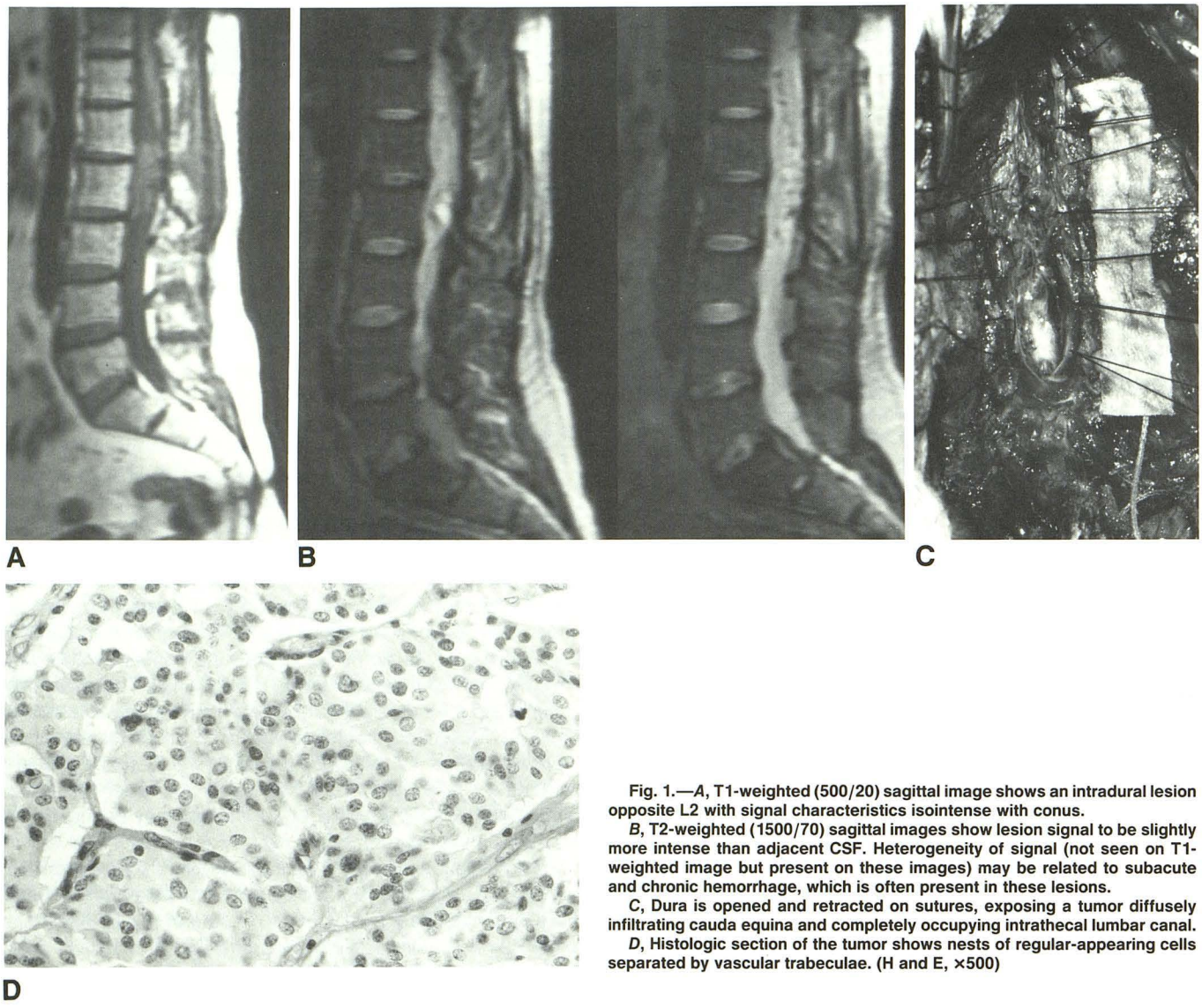

Fig. 1.—*A*, T1-weighted (500/20) sagittal image shows an intradural lesion opposite L2 with signal characteristics isointense with conus.

***B*, T2-weighted (1500/70) sagittal images show lesion signal to be slightly more intense than adjacent CSF. Heterogeneity of signal (not seen on T1-weighted image but present on these images) may be related to subacute and chronic hemorrhage, which is often present in these lesions.**

***C*, Dura is opened and retracted on sutures, exposing a tumor diffusely infiltrating cauda equina and completely occupying intrathecal lumbar canal.**

***D*, Histologic section of the tumor shows nests of regular-appearing cells separated by vascular trabeculae. (H and E, ×500)**

equina paraganglioma was initially described as a "secretory ependymoma" [4]. However, the presence of neurosecretory granules mitigates against ependymal origin, and later review of the case confirmed the diagnosis of paraganglioma. Similarly, our case was initially misdiagnosed as an ependymoma.

Biologically the neoplasm is quite different from an ependymoma. Most spinal ependymomas are located in the cauda equina and originate from the conus medullaris or filum terminale. These tumors may invade and expand cauda equina nerve roots and may invade meninges and bone. Slooff et al. [7] found that the average postoperative survival time for ependymoma of the cauda was 14 years; for ependymoma of the spinal cord they found the average postoperative survival time was 9 years. Their results also suggest that ultimate prognosis is improved with more extensive forms of therapy. Distant metastases also have been reported. Paragangliomas seem to grow more slowly. They are usually well-encapsulated intradural lesions without a dural attachment [5]. Most tumors are limited to the filum terminale, although secondary involvement of the conus and caudal nerve roots can occur [5]. Because paragangliomas are encapsulated, such tumors are generally excised totally at surgery. Distant metastases have not yet been reported. In a recent review of cauda equina paragangliomas by the Mayo Clinic, 26 of 31 patients undergoing total excision of their tumor remain disease free. Of four patients having subtotal excision, two remain asymptomatic after radiation therapy. A third patient also receiving radiation therapy for partial resection presented after 1 year with osseous invasion and retroperitoneal involvement. Twenty years later, the patient is paraplegic but alive. The fourth patient died of tumor-related complications [5].

Preoperative radiologic evaluation of these tumors has been done primarily by myelography. All of these myelograms have shown some degree of obstruction, either partial or total. Elevated CSF protein as high as 7000 mg/dl has been reported at the time of myelography. Although elevated CSF

protein levels are often seen in the presence of a subarachnoid block, the elevated levels in paragangliomas seem to be disproportionately high in comparison with the level or degree of block [8]. Plain films have generally been normal, with the exception of degenerative changes. In a few cases, bone erosion thought to be caused by compressive effects of the tumor has been described [9]. Two patients have had preoperative angiography showing a highly vascular tumor [1, 2]. One underwent preoperative embolization of the tumor. The authors of this report advocated preoperative angiography in patients with an appropriately located spinal tumor and elevated urinary biogenic amines [1]. As with other spinal tumors, MR imaging will most likely become the imaging technique of choice.

Adequate therapy requires total surgical removal. Although the value of radiotherapy for incompletely excised tumors has not yet been established, it is still recommended [2]. Prolonged follow-up is also mandatory because of slow tumor growth and late recurrence.

In summary, paragangliomas of the cauda equina are largely benign and encapsulated lesions that present with typical symptoms of a cauda equina lesion. Paraganglioma must be considered in the differential diagnosis of any lesion in this location, particularly when neuropeptide production is elevated. MR will be the imaging technique of choice in evaluating the lesions. Myelography, CT, and angiography may be helpful as adjunctive studies. Total surgical resection is considered optimum therapy.

REFERENCES

1. Solymesi L, Ferbert A. A case of spinal paraganglioma. *Neuroradiology* **1985**;27:217–219
2. Boker D-K, Wassmann H, Solymosi L. Paragangliomas of the spinal canal. *Surg Neurol* **1983**;19:461–468
3. Anderson J, Gullan R. Paraganglioma of the cauda equina: a case report. *J Neurol Neurosurg Psychiatry* **1987**;50:100–103
4. Miller CA, Torack RM. Secretory ependymoma of the filum terminale. *Acta Neuropathol* (Berl) **1970**;15:240–250
5. Sonneland P, Scheithauer B, LeChago J, Crawford B, Onofrio B. Paraganglioma of the cauda equina region: clinicopathologic study of 31 cases with special reference to immunocytology and ultrastructure. *Cancer* **1986**;58:1720–1735
6. Binkley W, Vakili ST, Worth R. Paraganglioma of the cauda equina: case report. *J Neurosurg* **1982**;56:275–279
7. Slooff JL, Kernohan JW, MacCarthy CS. *Primary intramedullary tumors of the spinal cord and filum terminale*. Philadelphia: Saunders, **1964**
8. Soffer D, Pittaluga S, Caine Y, Feinsod M. Paraganglioma of cauda equina: a report of a case and review of the literature. *Cancer* **1983**;51:1907–1910
9. Reyes MG, Toores H. Intrathecal paraganglioma of the cauda equina. *Neurosurgery* **1984**;15:578–582