Combined Paraganglioma and Glioma of Conus Medullaris and Cauda Equina

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A mixed paraganglioma and glioma occurred in the conus medullaris and cauda equina of a woman. The tumor was subdural, encapsulated, and vascular. It was first manifested with symptoms and signs of compression of the cauda equina when she was 19 years of age and was excised when she was 24. The tumor was attached to the conus medullaris, the dura mater, and a nerve root of the cauda equina. Striking light and dark cells with an organoid arrangement mingled with a few glial fibrillary acidic protein (GFAP)-positive neuroglia forming a combined paraganglioma and glioma. Twenty-three paragangliomas were reported in the literature including the present case. The sex and age of recorded 19 patients demonstrated ten males and nine females with an average age of 46.3 years. Twenty tumors were subdurally situated at the conus medullaris and cauda equina. Three neoplasms were epidural and thoracic in location.

KEY WORDS: paraganglioma, glioma, mixed paraganglioma and glioma, spinal tumor

INTRODUCTION

Paragangliomas are uncommon neoplasms related to the autonomic nervous system. They occur mostly in the region of the head and neck: Tumors of the carotid body arise from paraganglionic cells at the carotid bifurcation, and of the glomus jugulars in the middle ear from cells in the adventitia of the jugular bulb or along the course of the tympanic branch of the glossopharyngeal nerve. Rarely, they originate from paraganglionic cells in the ganglion nodosum of the vagus nerve. Other sites of paragangliomas include the mediastinum, retroperitoneum, and visceral organs such as the adrenal glands and lungs [1]. The exceedingly rare intracranial paragangliomas have been found at the petrous ridge [2], pineal region [3], and sella turcica [4]. To our knowledge, 22 cases of intraspinal paragangliomas have been recorded [5–17]. A case reported by Gullotta and Helpas [8] was later included by Böker et al in their series of seven cases [17]. We report herein a paraganglioma combined with glioma in the region of the conus medullaris and cauda equina. The complex clinical manifestation delayed proper management.

CASE REPORT

In July 1976, a 19-year-old woman first had hematuria. One month later, low-back pain radiating to the feet and cramping of the lower limbs occurred. In January 1977, she was diagnosed to have flat feet. Special shoes were recommended, but the pain was worse when she used them. Toward the end of 1977, urinary urgency and numbness of the right big toe ensued. Because of suspected urethral stricture, four consecutive urethral dilatations were performed without improvement of the urinary symptoms. In October 1978, she noted atrophy of the right leg. Throughout 1979, she experienced progressive weakness of the right lower limb. In 1980, low-back pain extended from the lumbar region to both hips, and she had difficulty in flexing the thighs. The right lower extremity became increasingly weak. Further investigation including electromyelogram led to a conclu-
sion that she had residual poliomyelitis. Another pair of shoes was prescribed as was a course of physiotherapy. Low-back pain, weakness of the right lower limb, and urinary urgency, however, continued. In April 1981, she had right-foot drop. Roentgenograms of the back now showed an abnormality of the vertebral column. She was then hospitalized, almost five years after the onset of hematuria and low-back pain.

Physical examination revealed normal motion of the back. Kyphoscoliosis and tenderness were absent. The following muscles on the right side were weak: iliopsoas, quadriceps, hamstrings, anterior tibialis, posterior tibialis, and peroneals. The right lumbar region was hypalgesic. The sacral region was analgesic bilaterally. Proprioceptive sense in the big toe and knee and ankle jerks were lost on the right side. The left big toe was weak in flexion and extension.

Urinalysis disclosed 2-plus protein, negative sugar, and 20 erythrocytes and four lymphocytes per high-power microscopic field. The lumbar cerebrospinal fluid contained 922 mg/dl of protein. Roentgenograms of the vertebral column exhibited abnormalities of the first and second lumbar (L1 and L2) vertebrae (Fig. 1). Myelograms showed complete blockage of the radiopaque column at the level of L2 (Fig. 2). These findings indicated an intraspinal mass.

An elongated subdural tumor was encountered upon removal of the thin and bulging laminae of L1 and L2.

The rostral end was attached to the conus medullaris. The right lateral aspect of the middle part of the growth was firmly attached to the dural sac covering the cauda equina. The tumor was surrounded by nerve roots comprising the latter. One nerve root was embedded in the anterorostral aspect of the lesion. The tumor was separated from the conus medullaris by blunt dissection, but it was difficult to free the dura mater and a nerve root. The attached dura mater and nerve root, then, were excised with the lesion, and the dural defect was grafted with a piece of dorsolumbar fascia. Weakness of the right lower limb and urinary symptoms were improved considerably when she was seen 12 and 22 months after operation.

Specimen was 5 × 3 × 2 cm, rubbery, and encapsulated. The light gray cut surface was interspersed by soft and friable foci. Paraffin sections were stained with hematoxylin and eosin (H and E), Mallory's phosphotungstic acid hematoxylin (PTAH), periodic acid-Schiff (PAS), Mayer's mucicarmine, and Gomori's stains for reticulin fibers. The peroxidase-antiperoxidase method was used to localize glial fibrillary acidic protein (GFAP) in formalin-fixed and paraffin-embedded tissue sections [18]. An astrocytoma was similarly processed as a control.

Microscopically, there were abundant polygonal and elongated tumor cells beneath the thick connective tissue capsule of the neoplasm. They were arrayed in nests with richly vascular connective tissue septa between the nests.
Fig. 3 Histopathology. A. Tumor with thick fibrous capsule containing large blood vessel on right margin of photomicrograph. Many connective tissue septa subdivide tumor cells into nests (Gomori’s stain, ×40). B. Nests of tumor cells are surrounded by connective tissue septa creating pseudoalveolar pattern. Note light cytoplasm of several cells (PTAH, ×100). C. Papillary arrangement of tumor cells, mostly light cells is shown. One large light cell is indicated by arrow (H and E, ×100).

Reticulin and collagen fibers seen in Gomori’s stains and PTAH preparations were present around the cellular nests creating a pseudoalveolar (organoid) pattern (Fig. 3A, B). Papillae with cores of scanty connective tissue (Figs. 3C, 4A) and cellular balls formed by envelopment of the tumor cells (Fig. 4B, C) were also scattered in the lesion. In the latter form of arrangement, the polygonal neoplastic cells were often encircled by elongated curvilinear ones. The individual tumor cells had well-defined borders, vesicular round to ovoid nuclei, and deeply staining nucleoli. Nuclear vacuoles were not seen. The fine granular cytoplasm was either pale acidophilic or amphophilic. Many tumor cells were pale, but others were dark, resulting in a striking light and dark appearance of cells (Figs. 3B, C, 4A). Mucicarmine and PAS stains were negative. A few tumor cells were positive for GFAP, indicating their neuroglial nature, astrocytic or ependymal in type or both (Fig. 5). These GFAP-positive neuroglia were deep within the lesion. The pathologic diagnosis was combined paraganglioma and glioma of the conus medullaris and cauda equina.

DISCUSSION

The differential diagnosis of the present neoplasm includes meningioma and glioma. Dural attachment, encapsulation, and high vascularization are features of a meningioma. The cellular balls, moreover, could be viewed as whorls in a meningioma. However, the presence of striking light and dark tumor cells and absence of syncytial appearance are not features of a meningioma.
Furthermore, nuclear vacuoles, additional diagnostic features of a meningioma [19], are not seen. Reciprocally, an organoid pattern of arrangement of the light and dark tumor cells and cellular balls formed by envelopment of the neoplastic cells have been noted in paragangliomas which are also encapsulated and vascular [1]. We are therefore inclined to consider our patient’s tumor as a paraganglioma rather than a meningioma.

GFAP, a component of neuroglial filaments, is often found in neoplastic astrocytes and ependymal cells as well as in certain tumors containing astrocytes and ependymal cells as in glioblastomas and medulloblastomas [20–23]. The presence of GFAP-positive cells in our patient’s tumor suggested it to be a glioma, astrocytoma, or ependymoma or both. Miller and Torack [5] reported a “secretory ependymoma” of the filum terminale, but it was later interpreted as a paraganglioma by others [7,11,13,17]. Horoupian et al [7] regarded their case as a paraganglioma of the cauda equina but admitted that some of its features were reminiscent of an ependymoma. Weidenheim et al [15] studied four paragangliomatous neoplasms of the filum terminale, using histochemistry, immunocytochemistry, and electron microscopy, and found unsuspected glia. Unfortunately, they did not state clearly which method revealed what finding. They concluded that paragangliomatous tumors in this region were ependymal in origin. Based on the presence of GFAP-positive tumor cells, we suggest that our patient’s tumor is a mixed paraganglioma and glioma.

It may be argued that neuroglia in our patient’s tumor are reactive and entrapped. However, their deep position within the tumor is not in accord with their reactive
nature. Reactive glia should be located at the periphery of the lesion. Neurons have been found in some paragangliomas of the conus medullaris and cauda equina, which led to the designation of the lesions as ganglioneuroma-paraganglioma [6], gangliocytic paraganglioma [14], and mixed chemodectoma and ganglioneuroma [16]. Thus, it would be surprising to encounter included neuroglia in paragangliomas, as in the present case. Neurons and neuroglia are intimately related; they are derivatives of the neuroepithelium.

Review of the literature disclosed that of a total of 23 patients with intraspinal paragangliomas including the present one, the sex and age were recorded in 19. There were ten males and nine females. The oldest one was 70-year-old man [17], and the youngest was a 19-year-old woman (present case). The average age was 46.3 years. Twenty tumors were subdurally situated at the conus medullaris and cauda equina [5–17]. Three were epidural and thoracic in location [17]. Disturbed motor and sensory functions of the lower limbs and urinary and rectal dysfunction were produced by tumors located in the region of the conus medullaris and cauda equina. These symptoms and signs are typical of any mass lesion in this region [24,25]. The duration of symptoms ranged from two months [17] to 15 years [5]. Attachments of the tumors to nerve roots occurred in four [5,9,12, present case], to dura mater in two [17, present case], to conus medullaris in one (present case), and to bone in one [17]. Partial removal was done in three patients [11,17]. In one of these patients, combined treatment of 6,000 rads of cobalt therapy and electron therapy has resulted in regression of the remaining tumor [11]. Recurrence of the tumors occurred in two cases, eight months and 12 years after extirpation, respectively [17]. One epidural thoracic paraganglioma occurred 15 years after removal of a tumor at the left carotid bifurcation [17]. Generally, the tumors were circumscribed, encapsulated, and vascular.

REFERENCES

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