CASE REPORT

Papillary (Malignant) Meningioma of the Foramen Magnum Presenting as a Posterior Neck Mass

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Abstract—Meningiomas and neurofibromas are the most common extramedullary tumors in the region of the foramen magnum. The clinical symptoms are variable, including headache, occipitocervical pain, and hoarseness. Neurological symptoms are present only in 40% of cases. The age group ranges from 4 to 56 years. A number of patients are misdiagnosed as multiple sclerosis, syringomyelia, and cervical disc disease. The overall incidence of these tumors vary from 1.6 to 2.4%. The majority of meningiomas are histologically benign. Papillary (malignant) meningiomas are even rarer in this location. The present report is that of a 46-year-old man who developed a painless mass in the nape of his neck, without any neurological symptoms. Following initial biopsy at another hospital, a definitive resection was undertaken at Loyola University Medical Center. The diagnosis of a malignant meningioma was made after extensive immunohistochemical and electron microscopic studies. The patient developed a positive cervical lymph node 1 year later, which was excised and showed identical histopathologic features (*Skull Base Surgery*, 4(3):164–168, 1994)

Meningiomas are the second most common intracranial primary neoplasm, accounting for about 18 to 20% of all brain tumors. The majority are located supratentorially along the cerebral convexity. Some 80 to 90% of meningiomas occur in the anterior half of the cranial cavity, and only 2 to 3% are found in the region of the foramen magnum.

The first systematic evaluation of foramen magnum tumors was given by Elsberg in 1925,¹ followed by two additional reports in 1929.^{2,3} The first extensive review of benign and malignant tumors of the foramen magnum was from Love et al.⁴ Then, Meyer et al.⁵ reviewed 102 documented cases of foramen magnum tumors and reported an incidence of 76% for meningiomas.

Foramen magnum tumors are difficult to diagnose clinically because they mimic other neurological diseases, including multiple sclerosis. As many as 40% of patients⁶ may have a normal neurological examination. Meyer noted that an association with Von Recklinghausen disease was reported in about 10% of patients. The majority of meningiomas (60%) were located anterior to the cord,

with 20% lateral and 20% posterior. The mean age was 49 years. Suboccipital neck pain was present in 65% of symptomatic patients.

CASE HISTORY

J.T., a 46-year-old right-handed white male priest, noticed a painless posterior midline neck mass unassociated with any other clinical symptoms for about a year. Physical examination revealed a well-developed, well-nourished white male with very broad shoulders. A large posterior midline, firm, hypomobile mass extended from the occipital protuberance to the middle of the posterior neck. The remainder of the physical examination, including otologic, neuro-otologic, and cranial nerve evaluation, was normal. A computed tomography scan showed a large lesion eroding through the posterior occipital bone adjacent to the medulla and extending inferiorly to the posterior portion of the body of the first cervical vertebra.

Figure 1. Magnetic resonance image showing the large tumor with involvement of the posterior fossa and upper cervical spinal canal.

The magnetic resonance imaging film (Fig. 1) revealed a large bulky occipital tumor extending into the foramen magnum, with intrathecal nodular infiltrate extending to C-1 and erosion of the skull base. A four-vessel angiogram showed no evidence of vascular supply from the vertebral arteries. An extensive workup for metastases, including bone scans, was negative. The tumor was excised along with the posterior musculature down to and including the dura of the spine. The tumor involved the midposterior of the occipital bone, the arch of C-1 ver-

tebra, through the foramen magnum and laterally between the mastoid tips. The right vertebral artery also was involved. Duraplasty was performed with fascia lata graft, and the surgical defect was repaired with a large trapezius myocutaneous flap.

PATHOLOGY

An incisional biopsy had been performed at an outside hospital and reported as glomangiosarcoma before the patient was referred to Loyola and underwent an extensive local resection with removal of the bone with the dura in the region of the foramen magnum and upper cervical spinal canal. The resected gross specimen was submitted to pathology in four separate containers. The first contained the main tumor mass with the attached skeletal muscle tissue, skin, and fascia. This specimen measured $15 \times 12.5 \times 5$ cm overall. The tumor nodule itself was circumscribed, firm, grey-tan in color, and focally hemorrhagic, and it was situated deep in all of the skeletal muscle tissue (Fig. 2). Portions of the dura and the occipital bone were submitted as separate specimens, and both showed gross involvement by tumor. The specimen from the left lateral neck margin also showed a large nodular tumor mass measuring 1.8 cm in diameter.

Histopathologically, the tumor was composed of highly vascular malignant papillary tumor, with areas of necrosis and increased mitotic activity. The nuclei were fairly uniform with slight vesicular chromatin (Figs. 3 and 4). The tissue was submitted for electron microscopic studies and immunohistochemical studies, and the results are reported as follows. Psammoma bodies were not seen.

Cytokeratins stains (CAM4) (Fig. 5) and neuron-

Figure 2. Gross photograph of the resected specimen. Note the location of the tumor, deep in the muscles (arrow).

Figure 3. Hematoxylin and eosin-stained sections showing papillary tumor with vascular stroma and uniform vesicular nuclei. Original magnification $\times 200$.

Figure 4. Note the uniform vesicular nuclei and hemorrhage in the background. Original magnification $\times 400$.

Figure 5. Immunoperoxidase stain showing focal positivity to cytokeratins. Original magnification $\times 200$.

Figure 6. Electron microscopic photographs showing the interdigitating cell membranes, incomplete basal lamina, and cell junctions. Original magnification $\times 37500$.

specific enolase stains were focally positive. S-100, chromogranin, Vimentin, thyroglobulin, factor 8, epithelial membrane antigen, desmin, synaptophysin, and muscle-specific actin were negative. Repeat immunostains were done at the Mayo Clinic and showed a similar reaction pattern. Progesterone receptors were reported as 2+ positive.

Electron microscopic studies revealed fairly uniform cells with complex interdigitating cell membrane, absent or incomplete basal lamina, and presence of cell junctions. No intracytoplasmic lumen or microvilli characteristic of adenocarcinoma were seen (Fig. 6). A diagnosis of malignant papillary tumor most consistent with malignant meningioma was made based on the electron microscopic studies.

The main differential diagnoses include metastatic adenocarcinoma, amelanotic melanoma, mesothelioma, synovial sarcoma, alveolar rhabdomyosarcoma, chondroma, and, occasionally, other sarcomas. In our patient, the battery of immunohistochemical stains was used to exclude all the differential diagnoses except metastatic carcinoma, which was excluded on the basis of extensive clinical workup. The electron microscopic studies provided the definitive diagnosis.

A radical neck dissection yielded 22 lymph nodes, one of which contained metastatic tumor. In January 1994 the patient developed a liver nodule that was diagnosed by fine-needle aspiration cytology as papillary metastatic tumor. The morphology was identical to the original tumor. The immunoperoxidase stains (S-100 and Vimentin) were reported positive in the liver metastasis.

DISCUSSION

Meningiomas arise from the "arachnoid cap cells" on the external surface of the arachnoid membrane, which are unique in structure and function. Arachnoid cap cells have characteristics similar to the lining cells of the peritoneum or pleura—namely, mesothelial cells and the synovial cells lining the joint spaces. Tumors arising from these traditionally "nonepithelial" cells have epithelial features and present a biphasic pattern. The epithelial features of the cells are demonstrated on immunohistochemical stains as well as ultrastructurally. Meningiomas also exhibit well-demonstrated epithelial and mesenchymal features, particularly in the so-called "secretory" meningiomas.⁷ The dual nature of the meningothelial cells expressed variably led Cushing and Eisenhardt⁸ to identify nine major types and 22 subtypes of meningiomas on a histological basis alone.

Papillary meningiomas are uncommon and constitute only about 10% of all malignant meningiomas.⁹ Brignolio and Favero⁶ reported an incidence of less than 2% of 750 meningiomas. Pasquier¹⁰ reported a 2.5% incidence of the papillary type in 277 meningiomas; the age ranged in his series was 3 to 69 years, with a median age of 36.5 years for papillary meningioma.

The papillary pattern in any meningioma is associated with more aggressive behavior, regardless of the presence of other histological patterns.¹¹ Moreover, distant metastases are seen in 30% of patients.¹² Papillary meningiomas occur more frequently in children. In a series of 46

patients published by Pasquier et al,¹⁰ 37 were adults and 9 were children under the age of 18 years. Three tumors were located in the posterior fossa, 3 in the spinal cord, and the rest had a supratentorial location. There were no papillary meningiomas in one series of 78 cases within the foramen magnum.¹³

As many as 40% of patients with foramen magnum meningiomas are asymptomatic. When symptoms occur, the most frequent presenting complaints are suboccipital neck pain, dysesthesias, gait disturbances, weakness, and hand clumsiness. The most common neurological findings include hyperreflexia, hand weakness, Babinski sign, spastic gait, sensory loss, and 11th cranial nerve involvement. However, none of these signs or symptoms are pathognomonic for foramen magnum tumors. Nuclear magnetic resonance scanning appears to be the most useful, noninvasive means of evaluating tumors in the foramen magnum region.¹⁴

At surgery, these tumors are often necrotic and usually massive, measuring up to 12 cm in diameter, as was the case with our patient. Histologically, this patient demonstrated a predominant papillary pattern (80%), with some areas showing syncytial arrangement. The fibrovascular stroma usually contain hyalinized blood vessels. Psammoma bodies are usually absent.¹⁰ Similarly, whorl formation is also rare in papillary meningiomas. The papillary pattern is most evident in the hemorrhagic and necrotic areas. Mitoses are frequently noted. Infiltration of the adjacent structures is seen in more than 75% of cases, as was true with this patient.

Local recurrence is considerably higher in papillary meningiomas—56.5%, as opposed to the usual rate of 10 to 20% with conventional meningiomas. Pasquier reported the presence of distant metastases in 21.7% of his series of 46 cases.¹⁰ Twenty-three of the 46 patients died within 0.1 to 16 years of the original operation, and 14 more were dead within 5 years.

The rate of metastases for all meningiomas is reported to be about 1%, with the most common sites being lungs, lymph nodes, liver, and kidneys. In our patient, the lymph node and liver metastasis showed identical papillary formations.

Various immunohistochemical markers are reported as positive, but none is specific for meningioma. Epithelial membrane antigen is reported to be positive in up to 80% of meningiomas. S-100 is reported variably, from none to as high as 65%. Cytokeratins (epithelial marker) and Vimentin are also expressed in meningiomas. The Vimentin stains the abundant intermediate filaments in the cytoplasm of meningiomas. Immunohistochemical demonstration of estrogen and progesterone receptors has been reported in fresh tissues. The use of monoclonal antibodies in formalin-fixed tissue is of questionable value, particularly in estrogen receptors. It has been proven beneficial in demonstrating progesterone receptors in meningiomas. The present case had positive progesterone receptors, supporting the diagnosis of meningioma.

Characteristic electron microscopic features described in meningiomas are (1) complex folding and interdigitating plasma membrane, (2) intracytoplasmic filaments, and (3) hemidesmosomes and absent or incomplete basal lamina. All features were present in this case.

Because diagnostic problems are so common with a predominant or exclusively papillary pattern some investigators question the meningiomatous nature of papillary meningioma. The main diagnostic difficulties are encountered with metastatic adenocarcinoma. Other neoplasms with papillary formations include mesotheliomas and synovial sarcomas. In the present case, extensive clinical workup failed to reveal any other primary sites of carcinoma, and the diagnosis was confirmed by electron microscopy.

In summary, a 46-year-old male patient presented with an unusual papillary malignant tumor that likely arose in the region of the foramen magnum posteriorly, infiltrated the occipital bone, and presented as a painless posterior neck mass. The final diagnosis of malignant papillary meningioma was made after extensive immunohistochemical studies and excluding other possible primary sites, and was confirmed finally by electron microscopy. To our knowledge, this is the first reported case of a malignant papillary meningioma in the foramen magnum area.

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