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Case report

Papillary meningioma with pleural metastasis: case report and literature review

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Papillary meningiomas are rare meningeal tumors which are associated with a grim prognosis. These tumors usually recur locally and in some cases they metastasize. The clinical, radiological and histopathological features of a case of a papillary meningioma with a pleural metastasis in a 13-year-old boy are presented. The literature on metastasizing papillary meningiomas is reviewed. Up to now, 131 cases of papillary meningioma have been reported in the literature. Only 8 cases gave rise to metastases outside the central nervous system. The preferential site of metastasis appeared to be the lung. This is the first report of a papillary meningioma giving rise to a metastasis in the pleura.

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The rarely occurring papillary variant of meningioma is known to recur quickly and potentially metastasize (1–9). Therefore, papillary meningiomas have been included in the group of malignant meningiomas (4, 10). So far, 131 cases of papillary meningioma have been recorded in the literature. In only 8 of these cases, metastases outside the CNS were reported (1, 3, 11–13). In children, meningiomas represent only 1 to 4% of all intracranial tumors (14). No less than half of these cases are histologically and clinically malignant (15). In the present report, a 13-year-old boy with the papillary meningioma variant with pleural metastases is described. Pleural metastases of papillary meningiomas have not been recorded before. The literature on papillary meningiomas with metastatic behavior is reviewed.

Case report

Clinical history

A 13-year-old boy was admitted with a 5-month history of intermittent headache and vomiting. He also complained of dizziness and his gait was unsteady. Physical examination revealed signs of meningeal irritation and left-sided otalgia. A blue–reddish mass was protruding in the left external auditory canal. CT and MRI scans revealed a large tumor in the posterior fossa extending into the medial fossa causing destruction of the petrous bone (Fig. 1A). The tumor was heterogeneous on CT imaging and showed intense contrast enhancement. A biopsy was obtained through retro-auricular craniotomy. The diagnosis of malignant papillary meningioma was made (Fig. 2A). Clinical hallmarks

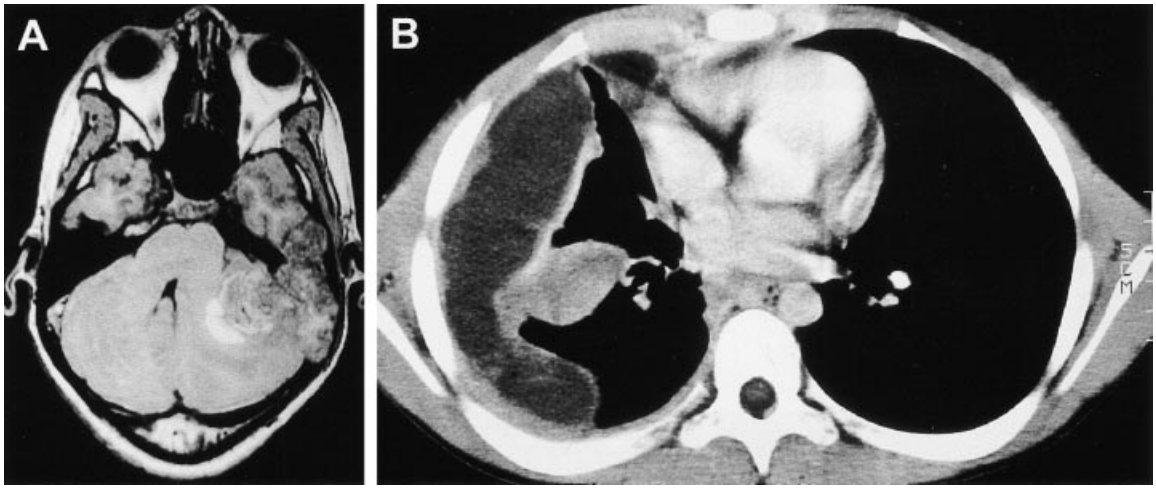

Fig. 1. (A) MRI of head. A large tumor destroying the petrous bone and invading the cerebellum. (B) MRI of thorax. A large right-sided mass is seen, thickening the pleura and compressing lung tissue.

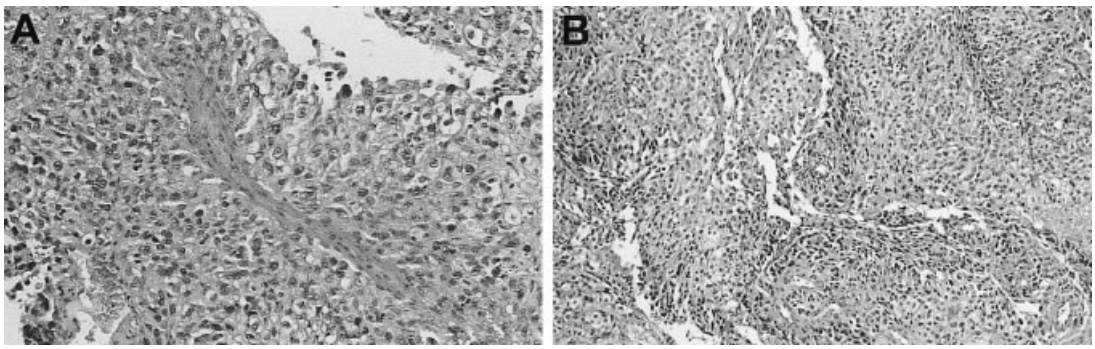

Fig. 2. (A) Primary meningeal tumor. The tumor tissue is composed of papillary structures with highly pleomorphic and spindle-shaped cells, with scattered mitoses and infiltration in blood vessels. Not shown: the tumor cells are immunopositive for vimentin, though negative for epithelial membrane antigen, while immunohistochemistry for glial fibrillary acidic protein, cytokeratin, S100, HMB-45, alpha-fetoprotein, β-human choriogonadotrophin, desmin, smooth muscle actin and lymphocytic markers remains negative. Ultrastructurally, the presence of cytoplasmic intermediate filaments, cell–cell articulation and desmosome formation were appreciated, corroborating the meningeal origin of the tumor (H&E, ×400). (B) Pleural metastasis of the papillary meningioma (H&E, ×250). The histology is essentially identical to that seen in Fig. 2A, and so is the immunohistochemical profile (not shown). There are papillary structures composed of cells with nuclear pleomorphism and scattered mitoses.

of neurofibromatosis type 2 were absent. Through retro-auricular craniotomy and left-sided partial mastoidectomy, debulking of the tumor mass was achieved. The transverse and sigmoidal sinuses were displaced and occluded by the tumor mass and the cerebello-pontine angle was invaded. Radical removal of the tumor was not possible. Postoperatively the patient suffered from palsy of the left facial nerve. Chemotherapy consisting of vincristine, lomustine, procarbazine, hydroxyurea, cisplatin, cytosine-arabinoside and cyclophosphamide was administered and combined with localfield radiotherapy at a total dose of 57.6 Gy. A remarkable shrinkage of the tumor mass was achieved. About 1 year after conclusion of the therapy the patient presented with mild dyspnea, cough and chest pain associated with respiration. Physical examination and plain films of the thorax revealed massive right-sided hydrothorax. CT scanning showed irregular thickening of the right pleura, suggestive of tumor. No lung involvement was noted (Fig. 1B). A biopsy of this pleural mass revealed metastasis of the intracranial tumor (Fig. 2B). The patient was subsequently treated with chemotherapy including ifosfamide, vincristine and actinomycin-D. Adjuvant radiotherapy with a total dose of 51.0 Gy was administered to the right hemithorax. The response to therapy was excellent and complete remission was achieved. Four months later, however, the patient was admitted again with right chest pain. CT scans showed tumor recurrence in the right lower part of the pleura, while no recurrence of the primary intracranial tumor was noticed. The patient died 3 months later from progressive disease.

Discussion

Generally, the term papillary meningioma is used for meningiomas which show, either exclusively or in part, papillary structures. Papillary histology may occur in low-grade and in anaplastic meningiomas. Cytology of squash preparations may be helpful in tracing the papillary formations (8). The clinical outcome of low-grade meningiomas with papillary foci is not necessarily unfavorable, whereas pure papillary meningiomas definitely should be considered malignant tumors (2–5, 8, 12). Up to now, 131 cases of papillary meningiomas have been reported in the literature. Most cases of papillary meningioma concerned adult males with a median age of 34 (28–40.8) years. Remarkably, a large proportion of tumors was located in the posterior cranial fossa, and locations rather unusual for meningioma have been described (5). About 55% of cases recurred, 75% of cases showed brain invasion, 20% metastasized and at least 50% of patients died from the disease (1, 3, 11–13). In most cases the tumors were surgically treated, ranging from debulking to radical removal. The median survival was 8 years, illustrating the malignant propensity of these tumors. Metastases of papillary meningiomas are preferentially located in the lung. Regarding non-papillary malignant meningiomas, only 79 extracranial metastases have been reported in the literature (16). Metastases involving the pleura are rarely seen (17). Recently, aggressive surgery followed by adjuvant radiotherapy was advocated in cases of malignant meningioma (18).

Meningiomas of higher malignancy grade show, in addition to loss of chromosome 22 or mutation of the NF2 gene, aberrations of various chromosomes, such as losses on 1p and 22q (19, 20). Meningiomas in childhood mostly affect boys, may occur in the setting of NF2, and approximately half of the cases behave in a malignant fashion (14, 15, 21–23). In a recent case report on a papillary meningioma occurring in the posterior fossa of a 3-year-old boy with dissemination through the CSF, partial trisomy 1q was observed as result of an unbalanced translocation 1;15 (p13;q11) (23). By now, data on the genetic idiosyncrasies of malignant meningioma, the papillary variant in particular, are too scanty to be helpful in specifically making the ominous diagnosis.

References

1. Cushing H, Eisenhardt L. The story of Dorothy May Russell. In: Meningiomas: their classification regional behavior, life history and surgical end results. Springfield, IL: Charles C Thomas, 1938:692–719.
2. Stefanko SZ, Mackay WM. Papillary meningioma. Acta Neuropathol (Berl) suppl 1981;**7**:126–8.
3. Fukushima T, Tsugu H, Tomonaga M, Shirakusa T. Papillary meningioma with pulmonary metastasis. Case report. J Neurosurg 1989;**70**:478–82.
4. Scheithauer BW. Tumors of the meninges: proposed modifications of the World Health Organization classification. Acta Neuropathol 1990;**80**:343–54.
5. Shuangshoti S. Primary papillary meningioma of the optic nerve sheath: A case of unique location and benign pathology. Surg Neurol 1993;**39**:200–3.
6. Radhakrishnan VV, Saraswathy A, Rout D. Papillary meningioma – a clinicopathological study of six cases. Indian J Cancer 1993;**30(4)**:164–8.
7. Lirng JF, Enterline DS, Tien RD et al. MRI of papillary meningioma in children. Pediatr Radiol 1995;**25(S)**:9–13.
8. Kobayashi S, Haba R, Hirakawa E, Yamadori I, Miki H, Ohmori M. Cytology and immunohistochemistry of anaplastic meningiomas in squash preparations. A report of two cases. Acta Cytol 1995;**39**:118–24.
9. Prayson RA. Malignant meningioma. A clinicopathological study of 23 patients including MIB1 and p53 immunohistochemistry. Am J Clin Pathol 1996;**105**:719–26.
10. Kleihues P, Burger PC, Scheithauer BW. The new WHO classification of brain tumours. Brain Pathol 1993;**3**:255–68.
11. Kepes JJ, Goldware S, Leoni R. Meningioma with pseudoglandular pattern. J Neuropathol Exp Neurol 1983;**42**:61–8.
12. Ludwin SK, Rubinstein LJ, Russell DS. Papillary meningioma: a malignant variant of meningioma. Cancer 1975;**36**: 1363–73.
13. Pasquier B, Gasnier F, Pasquier D, Keddari E, Morens A, Couderc P. Papillary meningioma. Clinicopathologic study of seven cases and review of the literature. Cancer 1986;**58**:299–305.
14. Deen HG Jr, Scheithauer BW, Ebersold MJ. Clinical and pathological study of meningiomas of the first two decades of life. J Neurosurg 1982;**56**:317–22.
15. Hope JK, Armstrong DA, Babyn PS et al. Primary meningeal tumors in children: correlation of clinical and CT findings with histologic type and prognosis. Am J Neuroradiol 1992;**13**:1353–64.
16. Rawat B, Franchetto AA, Elavathil J. Extracranial metastases of meningioma. Neuroradiol 1995;**37**:38–41.
17. Russell WE, Sach E. Fibrosarcoma of arachnoidal origin with metastasis. Report of 4 cases with necropsy. Arch Path 1942;**34**:240–61.
18. Dziuk TW, Woo S, Butler EB et al. Malignant meningioma: an indication for initial aggressive surgery and adjuvant radiotherapy. J Neurooncol 1998;**37**:177–88.
19. Weber RG, Bostrom J, Wolter M et al. Analysis of genomic alterations in benign, atypical, and anaplastic meningiomas: toward a genetic model of meningioma progression. Proc Natl Acad Sci 1997;**94**:14719–24.
20. Lamszus K, Kluwe L, Matschke J, Meissner H, Laas R, Westphal M. Allelic losses at 1p, 9q, 10q, 14q, and 22q in the progression of aggressive meningiomas and undifferentiated meningeal sarcomas. Cancer Genet Cytogenet 1999;**110**:103–10.
21. Schut L, Canady AI, Sutton LN, Bruce DA. Meningeal tumors in children. Pediatr Neurosurg 1994;**20**:207–12.
22. Molleston MC, Moran CJ, Roth KA, Rich KM. Infantile meningioma. Pediatr Neurosurg 1994;**21**:195–200.
23. Bouvier C, Zattara-Canoni H, Daniel L, Gentet JC, Lena G, Figarella-Branger D. Cerebellar papillary meningioma in a 3-year-old boy: the usefulness of electron microscopy for diagnosis. Am J Surg Pathol 1999;**23**:844–8.