A Case of Spinal Meningeal Melanocytoma

Spinal Meningeal Melanositom Olguşu

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Melanocytoma is a well differentiated neoplasm arising from leptomeningeal melanocytes. It was first described by Limas and Tio in 1972 (1). Leptomeningeal melanocytes are derived from the neural crest and they are found at highest density underneath the medulla and along the cervical spinal cord (2,3). Most tumors occur in the posterior fossa and the spinal cord (4). The prognosis of melanocytoma patients is usually favorable, however local recurrence is not uncommon. Rarely progression to malignant melanoma, leptomeningeal spread and even distant metastases have been reported (4-6).

Clinical Summary

66 year-old women patient was admitted to the Istanbul Education and Research Hospital. She had a pain in the right flank and both legs about 3 months before admission. Neurological examination was normal. Magnetic resonance imaging (MRI) showed a mass located posterior to the left side of the cord within the intradural space at the T11 level (Figure 1). Laminectomy revealed a T11 intramedullary tumor with a cystic cavity. Tumor was 10x7 mm in size. Total excision had been made. After 3 year follow up there was no recurrence.

Pathological Findings

They had monomorphic, round to oval vesicular nuclei with inconspicuous nucleoli. The cytoplasmic melanin content was variable in amount. Melanophages are abundant. Mitotic figures were very rare (0-1/ 10 hpf) and necrosis not observed. The MIB-1 index was 2 %. Focal areas of medulla spinalis invasion by tumor cells were showed (Figure 4). Immunohistochemically the neoplastic cells stained S-100 protein and HMB-45. In contrast, immunostaining for GFAP and epithelial membrane antigen (EMA) remained
negative. Based on these histological and immunohistochemical findings, the tumor was classified as a meningeal melanocytoma. A complete physical examination were done in dermatology department to rule out malignant melanoma and to investigate neurocutaneous syndrome. It was normal.

Primary melanocytic neoplasms of the central nervous system consist of a spectrum ranging from well-differentiated melanocytoma to its overtly malignant melanoma. Only a small group of intermediate lesions bridges these extremes with their significant difference in clinical behavior (4). The low-grade lesions have architectural and cytologic features useful in distinguishing them from the high grade ones (i.e., melanomas). Melanocytes disposed in tight nests with more heavily pigmented cells, bland cytologic properties are features of melanocytoma (4). Clinically aggressive melanomas show sheets, loose nests, overtly anaplastic cells, necrosis, increased mitotic activity and CNS invasion (4). In one study MIB-1 index was 2% to 15% for melanomas but only 0% to 2% for melanocytoma (7). Our case had bland cytology, low level mitotic activity and no necrosis, but some features such as microscopic CNS invasion, high MIB-1 index and focal sheet-like growth pattern pointed to intermediate differentiation. Local recurrences have been described in spinal meningeal melanocytoma. Even totally resected, transformation from melanocytoma to melanoma is a possibility (8,9). Melanocytoma can be misinterpreted as meningioma or schwannoma. Meningioma is reactive for EMA and not for markers such as HMB-45. Schwannomas share S-100 reactivity of melanocytic tumors, but are negative for HMB-45.

Spinal meningeal melanocytoma is a rare tumor of central nervous system. Histological examination of tumor tissue is mandatory to establish the diagnosis and exclude other tumor entities in particular malignant melanoma, melanotic schwannoma and meningioma. Even after complete resection, a through follow-up is necessary to identify local recurrence.

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