# Correspondence and Short Communications

*Comments on published articles, short communications of a preliminary nature, case reports, technical notes and the like are accepted under this heading. The articles should be short and concise and contain a minimum of figures, tables and references.*

## PRIMARY MENINGEAL MELANOMA

Melanocytic cells are normally found in the meninges of the central nervous system (CNS). They are numerous only on the ventral aspect of the lower part of the medulla oblongata, and a slight pigmentation is macroscopically visible in this region (1). Diffuse or circumscribed primary tumours derived from meningeal melanocytic cells are rare. We report a case of primary meningeal melanoma from our hospital and the total number of 6 in the files of the Norwegian Cancer Registry during a 30-year period (1955–1984). The database of the registry from the period 1955–1984 contained 9 cases registered as primary CNS-tumours derived from melanocytes. We evaluated all reports (clinical, pathological, death certificates) on these patients and excluded 3 because of obvious or possible primary tumour outside the CNS. The histological slides of the series were not reviewed. We accepted the case of a 9-year-old boy who died one year after diagnosis without autopsy. He was thoroughly investigated and no primary tumour was found outside the CNS.

The 6 patients with primary meningeal melanoma (Table 1) were reported from 4 different university hospitals. The incidence rate of this rare tumour was 0.005 new cases per 100000 per year (with the population in 1970 as basis). The series included 4 men and 2 women. The median age at time of diagnosis was 29 years. Four tumours were located in the intracranial and 2 in the intraspinal region (Table 2). Case 1 from our hospital will be presented more extensively:

*Case report.* A 25-year-old woman was admitted to hospital in March 1982 because of left-sided deafness, headache, nausea, vomiting and impaired vision of 3 months' duration. Physical examination revealed bilateral papillary oedema, facial nerve palsy and impaired vision. Cerebral computer tomography showed a tumour in the left cerebellopontine angle and a tentative diagnosis of acoustic neurinoma was made. A left suboccipital craniotomy was performed. The operation was not radical due to considerable tumour extension medially in the cerebellopontine angle. Two months later the patient was reoperated and the rest of the tumour was removed. The tumour was initially classified as a variant of acoustic neurinoma, but later reclassified as a primary meningeal melanoma (evaluated in collaboration with Prof. L. J. Rubinstein, University of Virginia School of Medicine, Virginia, USA.)

In November 1985 the patient had increasing ataxia and nausea. Cerebral CT confirmed recurrence of tumour and further tumour was removed in December 1985. Magnetic resonance imaging in June 1986 showed lesions suggestive of neoplastic tissue in the pons and the left cerebellar hemisphere. She then received radiation therapy from July to September 1986. During the last 16 months there has been no clinical or radiological sign of tumour progression. Her main complaint is a right-sided hemiparesis and ataxia. She also has a permanent left-sided facial nerve palsy in spite of a hypoglossal-facial nerve anastomosis in February 1983.

*Discussion.* The terminology and classification of neoplastic lesions originating in melanocytic cells of the meninges are not consistent in the literature. We follow the WHO classification (2) which makes a distinction between a tumour that arises in a focal site in the meninges and meningeal melanomatosis, the latter being a melanoma diffusely spreading in the subarachnoid space.

Primary meningeal melanomas follow a variable clinical course which can range from tumours with favourable prognosis to highly malignant tumours. Limas & Tio (3) therefore proposed to call the former 'meningeal melanocytoma'. We share the view (4) that the so-called 'melanocytoma' cannot be considered entirely benign. Its histological resemblance to uveal melanoma may

**Table 1**

*Summary of cases of primary meningeal melanoma reported to the Norwegian Cancer Registry 1955–1984*

| Case | Age | Sex | Location of tumour | Period of symptoms | Initial symptoms | Survival | Autopsy |
|---|---|---|---|---|---|---|---|
| 1 | 25 | F | Left posterior fossa | 3 months | Headache, deafness | 6 years[a] | |
| 2 | 62 | M | Thoracic medulla | 8 years | Increasing paresis | 4 years | + |
| 3 | 31 | M | Intracranial meninges | 4 months | Headache, nausea | – | +[b] |
| 4 | 27 | F | Right hemisphere | 2 weeks | Hemiparesis | 1 day | +[c] |
| 5 | 33 | M | Cervical to lumbar medulla | 4 years | Paresthesiae left foot | 8 years[a] | |
| 6 | 9 | M | Right occipital lobe | 1 week | Headache | 1 year | – |

[a] Alive at end of follow-up.
[b] Diagnosed by autopsy.
[c] Pregnant (7 months).

**Table 2**

*Primary malignant melanoma of the CNS in Norway 1955–1984*

| | All histologically verified tumours of the CNS | Primary malignant melanomas (percentage of total) |
|---|---|---|
| Intracranial | 6858 | 4 (0.06) |
| Intraspinal | 396 | 2 (0.51) |
| Total | 7254 | 6 (0.08) |

indicate a similar biology in that they both can recur after many years. The tumours with favourable prognosis are most often localized to the spinal canal and the posterior fossa (3).

Meningeal melanomatosis is often associated with large pigmented naevi of the skin (1, 5, 6). The base of the brain is most often involved and if there is involvement of the basal cisternas, internal hydrocephalus may result (1). Meningeal melanomatosis, like melanomas, represents a spectrum from clinically benign to highly malignant. There are a few reports of tumours exclusively involving the dura mater (7).

Primary meningeal melanoma was first reported by Virchow in 1859 (8). Since then approximately 220 cases of primary pigmented tumours of the CNS have been reported (9). Schoenberg et al. (10) have given a survey of the distribution of primary intracranial neoplasms by histological type in 8 different series, but only one of the series (that of Zimmermann) included 2 primary melanomas among 888 patients. The present series is, to our knowledge, the first to report an incidence rate based on cases in a defined population. We observed an interesting, although not significant difference between the relative frequence in the intraspinal and intracranial region (Table 2). No tumour was reported in the meninges of the medulla oblongata where most melanocytes normally are found.

The presenting symptoms resemble those caused by other neoplastic processes in the subarachnoidal space. The peak incidence appears to be in the 3rd to the 5th decade (9, 11). The tumours may be visualized on CT-scans and by MRI and, in cases of spinal meningeal melanomatosis, on myelograms. However, CT-scans may fail to show the tumour (12, 13), and in cases of spinal meningeal melanomatosis the myelogram may mimic arachnoiditis (14). Brain or meningeal biopsy is the only definite diagnostic procedure. The tumours with favourable prognosis are most often composed of many spindle cells arranged in fascicles with fine melanine granules in the cytoplasm. The highly malignant tumours show marked cellular pleomorphism, mitosis, necrosis and hemorrhages (3). As in melanomas of the skin, many tumour cells may be amelanotic. Cytology of CSF can show cells containing melanin and electronmicroscopy can be used to separate melanoma cells from melanophages (9, 12). As the great majority of pigmented tumours in the central nervous system are metastases from malignant melanoma, a careful examination of the patient, including the eyes, is necessary. As a main rule, metastasis outside the CNS from a primary meningeal melanoma is not accepted. However, one well documented cases has been reported (15).

The series from the Cancer Registry demonstrate the variable course of the disease and a forecast of prognosis based on histological examination alone is uncertain. Both meningeal melanoma and malignant melanoma of skin are neoplastic growths from melanocytic cells of a common origin. In the skin, changes in the antigenic profile of melanocytic cells during tumour development and progression are documented (16). Further studies on primary meningeal melanomas with special emphasis on antigenic factors may help to identify tumours with favourable prognosis.

*Key words:* Primary meningeal melanoma, incidence, case report.

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