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Original article

Neuroglial intramedullary tumors: The collaboration between neurosurgeons and neuropathologists

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ABSTRACT

Intramedullary tumors constitute approximately 5% of spinal tumors and about 80% are of neuroglial origin. We reviewed our series of adult patients with spinal neuroglial intramedullary tumors operated on between 1984 and 2011 at the neurosurgical department of Bicêtre hospital. The histopathological records for 196 patients were retrospectively analyzed. The majority of tumors were ependymomas (68%) and astrocytomas (27.5%). The importance of a proper and detailed neuropathological diagnosis is the key to define patient management. The available literature data about the genetic profiles of these rare tumors are summarized and reviewed.

1. Introduction

Primary spinal cord tumors represent 2–4% of all CNS tumors and they are classified as extradural, intradural extramedullary and intramedullary. Primary intramedullary tumors represent about 8–10% of the spinal cord tumors. The majorities are classified as WHO grade I or grade II tumors and consist of tumors of neuroglial origin in more than 80% of cases [1–3]. In the adult population, ependymomas are identified in 60–70% of cases and astrocytomas in 30–40% of cases [2–4]. Spinal hemangioblastoma is the other relatively frequent tumor and accounts for about 3–8% of intramedullary tumors [5]. Other rare lesions may also be identified, like gangliocytomas, oligodendroglioma and neurocytomas. Metastases account for only 1–2% of intramedullary spinal tumors.

The aim of this review was to describe our experience in the pathological analysis of intramedullary tumors of neuroglial origin and to underline the importance of the collaboration between neurosurgeons and pathologists to achieve the best clinical management for the patient. We would like to integrate our personal experience with the most recent literature data to offer a view on the fields not sufficiently explored and to help clinicians in opening new ways towards the best management of these tumors.

2. Methods

A retrospective analysis was performed to review the pathological reports of all the patients with intramedullary tumors operated on at the Neurosurgical department of the Bicêtre hospital, Assistance publique des Hôpitaux de Paris, between January 1984 and December 2011. The inclusion criteria were the pathological confirmation of a tumor of neuroglial origin and the adult age (> 18 years).

Cases of spinal hemangioblastomas as well as myxopapillary ependymomas of the filum terminalis were not considered in our analysis. Pediatric cases were also excluded.

The material available for the pathologic analysis was often limited. Some very small pieces were recovered after the use of the CUSA® Excel Ultrasonic Tissue Ablation System.

For a preliminary analysis, cytology smears were obtained and the fragments were embedded in paraffin and stained with hematoxilin-eosin (H&E) to perform a morphologic analysis. Masson's trichrome stain was also routinely used.

Immunohistochemical studies were routinely performed for glial fibrillary acidic protein (GFAP), neurofilament, synaptophysin, NeuN, Ki67/MIB1, p53, CD68, Olig2 and IDH1. In some difficult cases specific immunostaining were used for EMA, cytokeratin,

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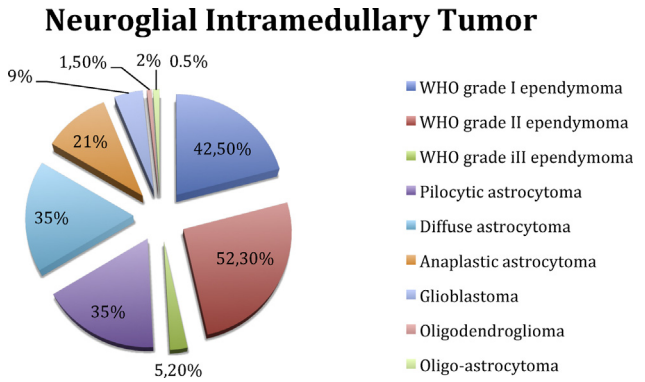

Fig. 1. Graphic summarizing the pathological records of our series of intramedullary tumors of neuroglial origin.

CD3, CD20, CD34. Additional pieces were frozen at −80 °C or fixed in zinc formalin to perform further molecular studies.

3. Results

During the study period, 196 cases of intramedullary tumors of neuroglial origin were identified in our series of spinal tumors.

The series consisted of 134 cases of ependymomas (68.4%), of which 57 cases were WHO grade I ependymomas (42.5%), 70 cases WHO grade II (52.3%) and 7 cases were anaplastic ependymomas (WHO grade III, 5.2%). In 54 cases, an astrocytoma was diagnosed (27.6%): in 19 cases, a pilocytic astrocytoma (35%), in 19 cases a diffuse astrocytoma (35%), in 11 cases an anaplastic astrocytoma (21%) and in 5 cases, a glioblastoma (9%). In 3 cases, an oligodendroglioma was diagnosed (1.5%) and in 4 cases an oligoastrocytoma (2%). One case of spinal gangliocytoma was also described (0.5%) (Fig. 1).

4. Discussion

Intramedullary tumours are not easy to access and in many cases, a complete resection is not possible. The resection is almost never an en-bloc resection and usually only small pieces are sent to the pathologist. The majority of the tumor is often removed by the Cavitron Ultrasonic Surgical Aspirator (CUSA) (Cavitron Lasersonic Corp., Stamford, CT). The tumor may be extremely heterogeneous and therefore, it is important to be able to analyze as much as tissue as possible. A close collaboration should be established between the neurosurgeon and the pathologist: the neurosurgeon should provide a detailed clinical history, past medical records of the patient and describe the radiological features of the lesion to the pathologist in order to achieve the correct diagnosis and the pathologist should provide as much information as possible to guide the follow-up clinical management of the patient.

In some rare cases, the tissue samples are too small to permit a histopathological conclusion. It is also important to rule out any pseudotumoral lesion, like infectious or inflammatory lesions.

The management of spinal intramedullary tumors of neuroglial origin is mainly based on the experience oncologists have on the cranial counterpart. To establish specific protocols is a delicate question because of the limited number of cases and of the limited knowledge about the genetic profile and the molecular pathways involved in the pathogenesis of these tumors.

4.1. Ependymomas

Ependymomas are the most frequent intramedullary tumors. They were previously thought to derive from the ependymal lining of the spinal canal, but they are now thought to arise from radial glial stem or progenitor cells [6]. They are most often well-circumscribed lesions unencapsulated, not infiltrating the adjacent spinal tissue and a complete resection is possible in the majority of cases. Ependymomas are classified as subependymomas or myxopapillary (WHO grade I), classic ependymoma (WHO grade II), or anaplastic (WHO grade III). WHO grade II ependymomas are the most frequently encountered tumors (70 cases/134 in our series, 52.3%). They are moderately cellular tumors, constituted by a well-defined proliferation of ependymal cells, elongated and disposed in perivascular pseudo-rosettes (anuclear zones surrounding blood vessels and composed by a collar of cytoplasmic processes of tumoral cells) or rarely organized in ependymal rosettes (cuboid or columnar cells surrounding a cavity). Perivascular pseudorosettes constitute the histological hallmark of ependymomas. The cellular nucleus is round or oval with a fine chromatin and rare atypical features. In some cases, cystic cavities are present, lined by ciliated ependymal cells. Several vessels are observed, with a hyaline wall and a pseudoangiomatous aspect. Some calcifications may be also present. The necrosis is exceptional and the mitotic index is weak (Fig. 2).

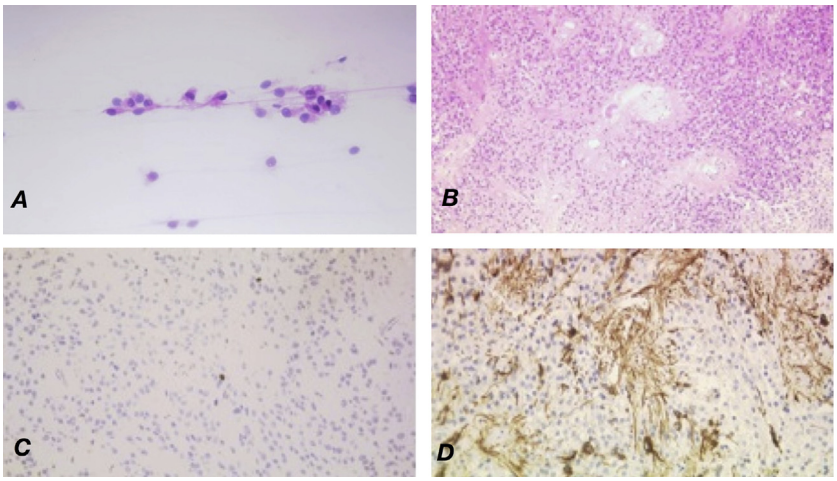

Fig. 2. Smear showing a WHO grade II ependymoma with elongated cells with round or oval nuclei with a fine chromatin. Rare atypia are observed (Fig. 2A). Paraffin inclusion of a WHO grade II ependymoma (Fig. 2B): ependymomas are moderately cellular tumors with tumoral cells disposed in perivascular pseudo-rosettes or rarely in ependymal rosettes. In some cases, cystic cavities are present. The vascularization is usually rich and vessels have a hyaline wall. The mitotic index is weak as visualized with the Ki-67 immunostaining (Fig. 2C). A diffuse positivity is evident at immunohistochemical staining for GFAP, stronger in cells forming pseudorosettes. (Fig. 2D).

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