Case Report

Intracranial Dural-Based Perineurioma: A Case Report

Gregory J. Imbarrato1, Bernd W. Scheithauer2, Jason K. Waddell1 and Emilio M. Nardone1*

1Division of Neurosurgery, Central Illinois Neuro Health Sciences, USA
2Pathology and Laboratory Medicine, Mayo Clinic, USA

Abstract

Perineuriomas are rare tumors arising in either a soft tissue or an intraneural form. We report the first case of intracranial dural-based perineurioma. Following histological confirmation of diagnosis, a rigorous literature review was undertaken for intracranial dural-based perineuriomas. Finding no previous such reports a case review and literature review of perineuriomas as a whole, and specifically intracranial perineuriomas, was undertaken. Mimicking a meningioma, a large left frontal dural-based tumor occurred in a 63-year-old woman experiencing personality change. The diagnosis on this gross totally resected tumor was based on its histologic features (spindle cells with long narrow processes), the finding of EMA, GLUT-1, and claudin-1 immunoreactivity, and the ultrastructural presence of surface micropinocytotic vesicles, partial basal lamina, and of simple intercellular junctions. Soft tissue perineurioma is rarely encountered in the intracranial space. Radiographically, the present tumor mimicked a meningioma with its well defined extra-axial location. The gross totally resected tumor was examined histologically, as well as by immunocytochemistry and electron microscopy.

INTRODUCTION

Perineuriomas are uncommon peripheral nerve sheath tumors arising from or differentiated toward perineurial cells. In peripheral nerve, the latter form the perineurium, enveloping individual nerve fascicles [1-3]. Alone, or in association with other nerve sheath elements, perineurial or perineurial-like cells contribute to the formation of various nerve sheath lesions, both reactive and neoplastic. The former include traumatic, plantar, and palisaded encapsulated neuromas. The latter includes mainly neurofibroma, a benign nerve sheath tumor. Tumors composed entirely of perineurial cells are uncommon and occur in two anatomic and histologic forms - one, a soft tissue tumor unassociated with nerve and far more often benign than malignant [4], the other, an intraneural, highly distinctive lesion characterized by pseudo-onion bulb formation [4].

Soft tissue perineurioma was first described in 1978 by Lazarus and Trombetta [5]. Since their initial description, approximately 50 additional examples have been reported [6-8]. The only reported case to date of a perineurioma arising in the intracranial space and affecting the central nervous system involved choroid plexus stroma [9]. In contrast, our tumor was extra-axial and dura-based. The cellular source of these rare intracranial tumors remains unclear. No parenchymal invasion or association with nerve was described. Perineuriomas of the central nervous system remain to be clinicopathologically characterized. Given the considerable histologic diversity of soft tissue perineurioma, they must be distinguished from other tumors in the differential, including meningioma and solitary fibrous tumor. Immunohistochemical and even ultrastructural studies are required [10]. No evidence-based guidelines exist for the management of perineuriomas. Resection may well be the mainstay of treatment. At present, it is unknown whether they will or will not respond to adjuvant therapies.

CASE PRESENTATION

A 63-year-old black female was admitted for neurosurgical evaluation following three weeks of progressive speech disturbance preceded by altered personality. The patient’s family stated her usual demeanor was ordinarily that of an unhappy person, whereas during the previous month, she had become docile and pleasantly tolerant of others. Quite literally, she sought medical attention for having become “too happy”. Although a lifelong smoker, she had no significant medical history.

On neurologic examination, the patient was pleasant. She exhibited expressive aphasias and showed no focal neurologic deficits. A Hoffman sign was evident on the right. A magnetic resonance imaging (MRI) scan revealed a large, left frontal, extra-axial tumor (Figures 1a, 1b) with significant mass effects upon the frontal and temporal lobes. The imaging characteristics were consistent with those of a meningioma [11]. The patient underwent a left frontal craniotomy and a gross total resection.
of the lesion and overlying dura was achieved. No parenchymal invasion or nerve association was seen. The postoperative course was uncomplicated and an MRI scan showed no residual tumor. On one-month and six-month follow-up evaluations, the aphasia had resolved, but her pleasant personality was unchanged.

Pathological Findings

Measuring 6 x 5 x 3 cm, the red-pink tumor was round in contour and somewhat cylindrical. Histologically, a tentative diagnosis of “mesenchymal tumor, perhaps hemangiopericytoma of the meninges” was made. The specimen was referred to Mayo Clinic in consultation. Further examination included immunohistochemical testing and electron microscopy. The tumor was monomorphic and featured elongate tumor cells, narrow process of which often encompassed collagen aggregates (Figure 2). No significant cytologic atypia was noted. Neither mitoses nor necrosis was present. No meningothelial cytology or architectural features of meningioma were seen. Alcian blue staining revealed interstitial mucus. The tumoral immunophenotype (streptavidin-biotin peroxidase complex method) included reactivity for epithelial membrane antigen (EMA) (Dako, Carpinteria, CA; clone E22 1:50), glucose transporter 1 (GLUT-1) (Dako, rabbit polyclonal, 1:200), claudin-1 (Invitrogen, Carlsbad, CA; rabbit polyclonal, 1:50), and collagen IV (Dako, clone CIV22, 1:25) (Figures 3, 4). Progesterone receptor (Dako, clone 636, 1:100), S-100 protein (Dako, polyclonal, 1:600), and neurofilament protein stains (NF) (Dako, BRD 2F11, 1:800) were immunonegative. Electron microscopy revealed elongated, remarkably narrow processes featuring membrane-associated micropinocytotic vesicles, rudimentary intercellular junctions, and discontinuous basal lamina (Figures 5a-5c). Collectively, these morphologic findings confirmed the diagnosis of perineurioma.
DISCUSSION

Comprised largely of perineurial cells, perineurium is a distinctive sheath of neuroectodermal cells that surround nerve fascicles [2, 3]. Tumors composed entirely of such cells are termed perineuriomas and occur in two anatomic and histologically distinct forms. One is a soft tissue tumor unassociated with nerve, and the other an intraneural lesion featuring pseudo-onion bulb formation. Both variants are rare within the intradural craniospinal space [4,9]. First reported in 1978 by Lazarus and Trombetta [5], the soft tissue subtype varies considerably in histologic pattern. Multiple soft tissue subtypes, including conventional [10], reticular [1], and sclerosing [12] types are described. Only occasional examples are malignant [10, 13].

Soft tissue perineuriomas usually present in adulthood and more commonly affect females [1]. Discrete and non-infiltrative, they are widely distributed throughout the musculoskeletal system; most affect the extremities and trunk. Sclerosing perineuriomas typically involve the hands of males [1, 12].

To our knowledge, only one other report describes a soft tissue perineurioma involving the central nervous system (CNS) [9]. The patient was a 65-year-old female who presented with obstructive hydrocephalus. Her lesion was intraventricular, arising from the choroid plexus at the level of the foramen of Monro. The preliminary clinical diagnosis was that of colloid cyst. Subsequently, the solid lesion was found to be a soft tissue perineurioma based upon its immunohistochemical and ultrastructural features [9].

Our lesion, a soft tissue perineurioma, exhibited the clinical and radiographic appearance of a benign, extra-axial, dura-based tumor, most consistent with meningioma. Were it not for thorough immunohistochemical and ultrastructural study, it would likely have been classified as a metaplastic meningioma or hypocellular hemangiopericytoma.

We suspect the diverse histology of soft tissue perineurioma contributes to their low frequency among neuropathologic specimens, perhaps being mistaken for World Health Organization (WHO) grade I meningioma. The benign, indolent course of most soft tissue perineuriomas furthers the confusion. An erroneous diagnosis is of little clinical significance. Not so if it is a misdiagnosis of hemangiopericytoma, a tumor which, when arising in the CNS, is considered malignant (low- or high-grade) by definition [14]. No evidence-based guidelines exist for management of central perineuriomas. The optimal approach to treatment of peripheral soft tissue perineuriomas is complete excision, in which case the prognosis is excellent. To date, no case has recurred or metastasized under these circumstances. Even in instances in which total excision may be difficult, careful observation is in order, as opposed to aggressive adjuvant therapy. At present, it is unknown whether peripheral soft tissue perineuriomas, even rare malignant examples, respond to such ancillary therapies.

Soft tissue perineurioma is rarely encountered in the intracranial space. Radiographically, the tumor mimicked a meningioma. After gross total resection the tumor was examined with histology, immunocytochemistry, and electron microscopy. The case presented represents the first described intracranial dural-based perineurioma.

ACKNOWLEDGEMENTS

This manuscript is dedicated to Dr. Bernd W. Scheithauer, who passed away unexpectedly on September 19, 2011. Without his expertise and contribution, this manuscript would not have been possible.

REFERENCES


