Multiple intraspinal low-grade astrocytomas mixed with lipoma (astrolipoma)

Case report

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Multiple intraspinal low-grade astrocytomas without neurofibromatosis stigmata and low-grade astrocytoma with intermingled areas of adipose tissue have not been reported previously. The authors present the case of a 48-year-old woman with a 7-month history of paraparesis. When she underwent surgery, multiple intraspinal mixed tumors made up of astrocytes mingled with adipose cells were found and excised. In this report, the authors refer to this tumor as an “astrolipoma” and discuss its characteristics.

Key Words • astrocytoma • spinal cord • intramedullary tumor • lipoma • magnetic resonance imaging

Spin al cord astrocytomas are relatively rare lesions with indolent clinical evolution and long patient survival times. The many series related to this subject that have been published do not mention the occurrence of low-grade astrocytoma with simultaneous multiple locations. Two other cases of multiple astrocytoma locations have been mentioned, but they were associated with von Recklinghausen’s disease. We report multiple intraspinal low-grade astrocytomas that showed intermingled areas of lipoma. We have not been able to find any other case in the English literature that presents both of these characteristics: multiplicity and mixed histological appearance.

Case Report

This 48-year-old woman presented in March 1992 with a 7-month history of dorsolumbar pain and progressive weakness of both lower limbs with gait disturbances.

Examination. Neurological examination disclosed a suspended hypesthesia from T8–L2, a paraparesis (3/5 to 4/5) that was worse on the right side with brisk patellar reflexes, absent Achilles reflexes, and bilateral Babinski’s sign. There was also some impairment of sensation in the anterior aspect of both legs, the soles, and the ends of the toes. There were no von Recklinghausen’s disease stigmata. Gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA)–enhanced magnetic resonance (MR) imaging of the spine demonstrated, in the first site, a nodular intramedullary mass between T-5 and T-8 with defined margins in the upper half and blurred limits in the lower half. In the second site, enhanced MR imaging revealed another mass occupying the entire intradural space between L-4 and S-2 (Fig. 1).

First Operation. An L3–S3 laminectomy was performed. On dural incision, part of the tumor mass protruded like a mushroom. Intraoperative biopsy showed the specimen to be a low-grade astrocytoma. The mass was made up of multiple encapsulated tumors, each one attached to a different lumbar or sacral root. The tumors were totally removed with the use of an operating microscope and an ultrasonic aspirator. A lateral/bilateral arthrodesis from L3–S1 was performed.

First Postoperative Course. In the postoperative period the paraparesis worsened to the level of 2/5 to 3/5 and sphincter disturbances appeared.
Second Operation. Three weeks later the patient underwent a second operation and a T5–9 laminectomy was performed. An exophytic intramedullary tumor (T5–6) was found on the right posterolateral aspect of the spinal cord. This tumor was 3 cm long, 2.5 cm deep, and 1 cm wide. The mass was excised until the interface between the tumor and surrounding spinal cord became unclear. On the left of the posterolateral aspect of the spinal cord and in contact with it lay two more independent tumors at levels T6–7 and T7–8. These two 1.5-cm diameter tumors were also excised.

Second Postoperative Course. In the immediate postoperative period the patient was completely paraplegic. She was transferred to rehabilitation and 6 months later she began radiotherapy. Two years after the operation she can walk without any help and presents with only a monoparesis of the right lower limb (4/5) with right Babinski’s sign; she requires bladder catheterization once a day. The enhanced spinal MR image has not shown any remaining tumor.

Histological Examination. Histological examination of the dorsal intramedullary tumor (second operation), represented by several fragments, showed a low-grade astrocytoma mixed with randomly scattered adipose cells either alone or in small clusters (Fig. 2A). The astrocytic tumor had a low cellular density of unipolar or bipolar pilocytic cells as well as round or stellate astrocytes with strong phosphotungstic acid–hematoxylin staining and positive glial fibrillary acidic protein (GFAP) immunoreactivity. Microcystic appearance and occasional astroblastic (gliovascular) pseudorosettes, but not ependymal rosettes, were present (Fig. 2B). Additionally, areas of lobular adipose tissue and GFAP-positive glial cells were seen (Fig. 2C). Immunohistochemical reactions for actine were negative. The histological appearance of the lumbosacral specimen from the first operation also showed a mixed tumor composed of similar astrocytic tumors, intermingled with adipose tissue, usually represented by isolated or a few grouped lipomatous cells (Fig. 3), although lobular adipose tissue areas were not infrequent (Fig. 4).

Discussion

This case has two peculiarities: its histological appearance and its multiple location.
Multiple intraspinal astrolipomas

Histological Appearance

Interestingly, the tumor held both glial and adipose tissue. The glial aspect of the tumor was the largest component. Its unequivocal tumoral nature allowed us to exclude a hamartomatous origin. The adipose tissue was represented by isolated cells, grouped cells, or by lobular adipose tissue. It is difficult to discern whether both types of cells (astrocytic and adipose) grew simultaneously in the same mass, or whether the adipose cells were secondary to an aberrant metaplasia in the glial tissue. There were no data that would objectively suggest a metaplastic meningial origin. We think that these lesions are different from the complex form of hamartoma, described by Russell and Rubinstein,11 which was constituted by parts of lipoma together with islands of fibillary neuroglia, neurites, and occasional ganglion cells. The angiolipomatous nature of the lesions was also excluded (actine-negative immunostain). Thus, although the pathogenesis of this entity is uncertain, the term astrolipoma can be applied to this type of lesion, because the two cellular components are quite obvious.

Multiple Locations

There are three possible explanations for the multiple locations of this tumor: multifocality, glial meningeal heterotopia, and/or leptomeningeal spread.

Multifocality. The tumor might have had multiple primary foci, and in this sense, it could be a multicentric astrocytoma. After excluding cases with concomitant disease such as neurofibromatosis, tuberous sclerosis, or multiple sclerosis, Barnard and Geddes3 found 18 cases (7.5%) of multicentric tumor among the 241 cerebral gliomas they examined postmortem. Only six cases of the 18 were classified as astrocytomas, which represents, in turn, 2.5% of the series. This reflects the rarity of multicentric astrocytoma in a cerebral location. This tumor would be much more infrequent if we consider the spinal location, because spinal cord tumors are less than 10% as frequent as intracranial tumors. However, the multifocality hypothesis has some shortcomings: except for the T-5 intramedullary tumor, all of the remaining tumors in our patient were located within the subarachnoid space. Although Reimer and Onofrio9 in their spinal cord astrocytoma series noted a case with a tumor extending between L-3 and the sacrum, there is no explanation in our case for the presence of the other tumors in the subarachnoid space, unless the two following possibilities can also be considered.

Glial Meningeal Heterotopias. Tumors located in the subarachnoid space might actually be meningeal heterotopias. In fact, some decades ago Kerntohan, et al.,11 which was constituted by parts of lipoma together with islands of fibillary neuroglia, neurites, and occasional ganglion cells. The angiolipomatous nature of the lesions was also excluded (actine-negative immunostain). Thus, although the pathogenesis of this entity is uncertain, the term astrolipoma can be applied to this type of lesion, because the two cellular components are quite obvious.

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brain barrier in low-grade astrocytomas. Therefore, the passage of contrast material through the barrier would be slow and progressive, and thus, the image should become clearer over time. Consequently, it is possible that if we had performed a later scan in our case, tumor margins would have been more precise.

In conclusion, we have presented the case of a low-grade spinal astrocytoma mixed with areas of lipoma (astrolipoma) presenting with simultaneous multiple locations at the dorsal and lumbosacral regions. This manifestation of tumor growth should be taken into consideration in the future. Radical surgical removal should be attempted so as to permanently cure the disease.

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