Neurosurgical desmoid tumors

Presentation of four cases with a review of the differential diagnoses

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Four neurosurgical tumors of desmoid appearance are presented, along with a brief review of the differential diagnoses of intracranial or spinal fibromatous or desmoid lesions. Two of the tumors were identified as extremely collagenized meningiomas by their typical fine structure. The classification of the other two tumors remains uncertain, but they were thought to belong to the family of desmoid tumors.

KEY WORDS • desmoid tumor • fibroma • meningioma • collagen • brain tumor

The various types of fibromas or desmoid tumors arising in the soft tissues of the body differ from meningiomas in their microscopic structure and their biological behavior. Only rarely is the neuropathologist faced with a cranial or spinal lesion that has greater similarity to a fibroma than to a meningioma. Four cases with neurosurgical tumors of a desmoid character are presented along with a review of differential diagnoses for such rare lesions.

Case Reports

Case 1

This 11-year-old girl had limped on her left foot for 18 months before admission. Her left leg was 1 cm shorter than the right. Spasticity and hypesthesia of her left leg had worsened during the 9 months before admission, leading to paraparesis. Myelography showed an incomplete block at T2-5. On laminectomy, an intramedullary mass was found beneath the pia mater and slightly to the left side underneath the dorsal surface of the distended cord. The tissue was whitish, avascular, and extremely firm; its cut surface resembled an intervertebral disc, but it was even harder and more dense. The resected mass was the size of a hazelnut; only a 3-mm thick segment of compressed cord remained after its extirpation.

All specimens of tumor tissue had the same structure on microscopic examination; there was a dense, hyaline, collagenous tissue with very few nuclei and with widely spaced capillary blood vessels (Fig. 1). Cells and collagen fibers were oriented at random, without forming streams or concentric whirls. No abnormally thick collagen fibrils were seen. The shape of the sparse, scattered nuclei varied, including rounded, elongated, bent, hooked, or irregular forms. They had little, fine, dusty chromatin, except for pyknotic forms. No mitoses were seen. Scattered small perivascular nests of myxoid cells were embedded in a ground substance that stained with Alcian blue. The surface of the tumor nodule interdigitated extensively with islands of proliferated fibrillary glial tissue, which included scattered Rosenthal fibers. Glial islands were always sharply delineated from the collagenous tissue. In the zone where the islands intermingled with the tumor the latter showed increased cell density with many large, vesicular fibroblast-like nuclei between the strands of collagen. No material suitable for electron microscopic examination was available. A tentative diagnosis of an “intraspinal desmoid neoplasm” was made.

Case 2

This 67-year-old woman had suffered from headaches for 3 years and had a first epileptic seizure 1 year before admission. Radiology disclosed a hyperostosis frontalis interna and a paramedian erosion of the frontal table on the left side, with reduced density and indistinct bone spicules. Angiograms showed a large avascular left frontal mass with veins
FIG. 1. Case 1. Much compact collagen is seen between widely scattered nuclei of variable shapes. H & E, X 400.

A nodular tumor, $5 \times 5 \times 5$ cm in size, was found on craniotomy; it centered on the coronal suture with infiltration of the dura mater and erosion of the frontal bone. The tumor was extremely firm — like cartilage, and could be cut only with difficulty. There was no bleeding from the cut surface and the tissue was greenish-white, like jade, and quite unlike any meningioma the surgeon (Prof. Yaşargil) had seen. The inner face of the tumor adhered to the frontal lobe; in the cortex there was evidence of pressure atrophy or ischemic necrosis.

Microscopic examination showed dense homogeneous masses of collagen rather similar to that in Case 1 (Fig. 2). The collagen fibrils were tightly interwoven and did not appear to be exceptionally thick. Much of the tissue showed extremely few and widely scattered nuclei and some fields were nearly acellular and avascular. There were also zones of medium cell density in which the evenly spaced nuclei were oriented with their axes parallel to the prevailing direction of the collagen fibers. Most nuclei were elongated, and of slightly irregular configuration with a moderate amount of fine, dusty chromatin. No mitoses were seen. Some parts of the tumor had patches of greater cell density, often surrounding the vessels. In these parts there was less collagen and the loosely woven stellar cells had a myxomatous appearance. These cell nests stained preferentially with Alcian blue, but not with periodic acid-Schiff (PAS). No glial tissue was found. There were extremely few mineral deposits, some concentric, but it was not clear whether they were entrapped by or were part of the lesion. The tumor was provisionally classified as a non-malignant fibroma.

The electron microscope showed scattered cells, most of them single, between bundles of collagen fibrils that were loosely woven through much larger amounts of a structureless, electron-lucent ground substance (Fig. 2). Most cells were slightly elongated or irregular, and their cytoplasmic bodies had sparse, plump, irregular, tongue-like or lamellar extensions. There was no basal lamina. Few intercellular junctions of indistinguishable type were seen. The prevalent cytoplasmic organelles were granular endoplasmic reticulum, discrete in some cells, and in profusion in others. Cytoplasmic filaments were sparse or absent. Few other organelles were seen. Nuclei were oval with smooth outlines and a relatively sparse heterochromatin at the nuclear membrane. Some nuclei with folded membranes were also seen. Many cells showed retrogressive changes with numerous fat droplets in their rounded cytoplasmic bodies.

During the next 7 months the patient's hemiparesis improved slowly. Then her condition began to deteriorate again and she had to be readmitted after a further 3 months. A large frontoparietal recurrence of the tumor was found by computerized tomography (CT). Repeat craniotomy showed that the recurrent tumor had infiltrated the brain tissue and the sagittal sinus; also, there were four large and seven small tumor foci in the dura mater. The tissue obtained at the second craniotomy, 11 months after the first operation, included large portions of tumor with the same hypocellularity as was seen in the first biopsy. In addition, there were portions with increased cell density and reduced collagen content (Fig. 3). The amount of intercellular Alcian blue-staining material was also greatly increased. The nuclei were now much more variable in size, shape, and chromatin content, with some very large scattered nuclei and many hyperchromatic forms. Spindle-shaped cells predominated, but there were also stellar or irregular shapes. Still, few mitotic figures were found. The cellular, dedifferentiated portions of the tumor blended smoothly with the hypocellular collagenous portions. A diagnosis of malignant change in a "dural fibroma" with a myxoid component was made. The fine structure of the tissue was still characterized by a dissemination of single cells within a loosely woven, collagenous ground substance. However, the tumor cells generally had greater electron density.
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FIG. 2. Case 2. Tumor specimens obtained at the first operation. Left: Photomicrograph showing that the tumor consists entirely of dense collagen and sparse nuclei, widely scattered or in small groups. H & E, × 250. Center: Electron micrograph showing the tumor cells are mostly single. Many have a prominent rough endoplasmic reticulum. × 18,100. Right: Electron micrograph of a tumor cell, intimately associated with collagen fibrils. All tumor cells lack a basal lamina. × 18,100.

cytoplasmic bodies were smaller, and most nuclei had deeply folded membranes (Fig. 3). The cytoplasm had a uniform, fine granular structure with relatively few mitochondria. There were many fat droplets. Much less rough endoplasmic reticulum was seen than in the first biopsy. Some cells had short pseudopodia or elongated interdigitating lamellar extension, but junctions were few and of an undefined type.

Case 3

This 9-year-old girl had epileptic seizures 8 months before admission and had complained of headaches since. An orange-sized, extremely hard, and poorly vascularized tumor was found on craniotomy; it was attached to the right sphenoid wing and compressed the right frontal lobe from below. Only a subtotal resection was possible.

Microscopic examination showed large masses of dense collagenous tissue (Fig. 4). The collagen fibrils were arranged into sharply delineated interwoven fascicles which criss-crossed each other, a feature different from the compact, homogeneous structure seen in Cases 1 and 2. Disseminated between the collagen fibrils were uniformly spaced, elongated nuclei with a dense chromatin. No mitoses were seen. Some parts of the tissue showed small cellular islands between the fascicles of collagen, resembling a loose mesenchyme of spindle-shaped or irregular cells. They did not have a patently meningioma-like tissue pattern. The islands did not stain with Alcian blue. A few psammoma bodies could be seen. On first inspection, this tumor was thought to be a fibroma similar to those in Cases 1 and 2, but the diagnosis was revised on electron microscopic examination.

The electron microscope disclosed large areas of tightly packed cells having large round to oval nuclei with heterochromatin arranged peripherally (Fig. 4). The cells had large, electron-lucent cytoplasmic bodies, including mitochondria, lysosomes, a profusion of thin filaments, and some rough endoplasmic reticulum. The density of the latter varied considerably from one cell to another. Most cells had abundant lamellar processes that frequently interdigitated, and were joined sporadically by gap junctions or by desmosomes (Fig. 5). Often the cells were packed so tightly that only narrow intercellular clefts
remained between them. Elsewhere, there were bundles of collagen and occasional elastic fibers between cell processes attached to each other in the manner of arachnoid trabeculae. Based on its typical fine structure, this mass of collagen tissue was identified as a meningioma.

**Case 4**

This fibroblastic falx meningioma from a 40-year-old woman presented no diagnostic difficulties. It is mentioned here because roughly two-thirds of the tumor was composed of dense collagen tissue, and because the fine structure of this tumor was indistinguishable from that of the tumor in Case 3.

**Discussion**

**Differential Diagnoses**

A brief review of differential diagnostic considerations may be useful in considering these and similar cases.

**Non-Neoplastic “Colloid” Scar Tissue.** Chronic inflammation or severely destructive traumatic or surgical lesions may cause intracerebral collagenous scars, which replace destroyed parenchyma; their periphery fuses with the adjacent tissue by way of a mixed gliomesodermal scar. Clinical manifestations, if any, are from the defect of cerebral tissue rather than from an expanding compressing lesion. An intracortical vascular mass of connective tissue described by Willson, et al., was thought to be akin to syphilitic colloid degeneration of the brain.

**Excessive Collagenous Transformation of Meningiomas.** Excessive amounts of dense tendon-like collagen may become the predominant component of a meningioma. These tumors are usually recognizable on sight by the residual islands of meningioma tissue or by the psammoma bodies. Identification may be difficult, however, if nearly all of the meningioma is replaced by collagen. Ghatak described such a spinal mass which was thought to be a collagenized meningioma.
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Fig. 4. Case 3. Specimens obtained at tumor resection. Left: Photomicrograph showing that this tumor was formed entirely of interwoven bundles of collagen fibrils. H & E, × 400. Right: Electron micrograph showing its fine structure is characteristic of a meningioma. It has tightly apposed cells with many filaments in their interdigitating processes. × 18,100.

Fibrosarcoma. Primary fibrosarcomas or other types of sarcomas of the central nervous system are patently malignant neoplasms, which present as pinkish, soft tumors. They do not exhibit a desmoid appearance, and their malignancy is evident from high cell density, pleomorphism, atypical cells, and mitoses.

Intracerebral Fibromas. Only a few presumably benign intracerebral fibromas have been recorded. All were found within the cerebral tissue and formed firm, sharply delineated nodes composed of well differentiated fibroblasts with variable amounts of reticulum fibers and collagen. Tightly packed collagen was seen in the case reported by Koos, et al., and hyaline-like areas of dense compaction with few nuclei were seen by Baker and Adams. A homogeneous desmoid appearance has not been observed in intracerebral fibromas.

Compound Intracranial Fibromatous Lesions. None of the few reported compound intracerebral, intracranial, or spinal fibromatous neoplasms, such as myxofibromas, xanthofibromas, or chondromyxoid fibromas of the vertebral column, had a uniform desmoid appearance.

Monostotic Fibrous Dysplasia. Monostotic fibrous dysplasia of the calvaria may present a trap to the unsuspecting neuropathologist who must distinguish this tumor from bone invasion by a fibroblastic meningioma. The lesion is most common before puberty. None of Henry's 50 cases involved the calvaria, but Reed found one among 16 lesions in the frontal bone, and Schlumberger reported five of 69 in the calvaria. However, Reed classified both non-ossifying and ossifying fibromas as variants of fibrous dysplasia.

Monostotic fibrous dysplasia has a characteristic “woven” texture of the bone spicules, and these are not rimmed with osteoblasts. The fibrous marrow tissue shows a loose mat of delicate fibers interlacing at random. Fibrous dysplasia does not form the circumscribed tumor islands seen when a meningioma invades the marrow spaces. It never assumes a desmoid
character and merits comparison with the present cases only in terms of the bone invasion seen in Case 2.

**Ossifying Fibroma.** Ossifying fibromas may distend the frontal bone, but they rarely form meningioma-like intracranial masses. Ossifying fibromas are distinguished from invasive meningiomas by the fibromatous texture of their marrow tissue; they differ from monostotic fibrous dysplasia in the lamellar structure of the bone spicules which are lined with rims of osteoblasts. Ossifying fibromas do not exhibit homogeneous desmoid features.

**Periosteal Desmoid Tumor.** Periosteal desmoid tumors form nodular masses with a striking predilection for the femur, proximal to its median condyle. A characteristic scooped-out bone defect results. The periosteal desmoid tumor forms a rare clinicopathological entity defined by localization and radiological features. Conceivably a periosteal desmoid tumor could arise in the calvaria, but no record of such a case has come to our attention.

**Desmoplastic Fibroma of Bone.** Desmoplastic fibroma of the bone is an uncommon lesion that occurs almost exclusively in patients below the age of 30 years, and in a variety of bones, particularly in the metaphysis of the long bones. Desmoplastic fibroma occurs in the vertebrae, but only in one of 50 cases was it found in the skull. Lately, there is an increasing number of reports on desmoplastic fibromas of the jaws, mainly the mandible, and conceivably such lesions may be found more frequently in the cranium. Dandy mentioned a circumscribed, entirely intraosseous fibroma of the calvaria, although no microscopic description was given.

Desmoplastic fibromas show an intermingling of well differentiated fibromatous tissue with collagenous zones having a desmoid appearance. This pattern is reminiscent of the tumors in our Cases 1 and 2. However, desmoplastic fibromas are typically intraosseous and have a trabeculated texture on radiological examination. Recurrence of a desmoplastic fibroma in the form of a fibrosarcoma has been observed.

**Soft-Tissue Desmoid Tumor.** Classification of soft-tissue desmoid tumors is determined by topography.
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rather than by the histological features, which are the same wherever soft tissue desmoid tumors occur. Hence, abdominal desmoid tumors, extra-abdominal, or musculoaponeurotic fibromatosis, plantar fibromatosis, dermatofibromas, or generalized juvenile fibromatoses are by definition beyond the scope of comparison with our cases. Such fibromatoses may present as single lesions of the head and neck. Conley, et al., and Masson and Soule reviewed 74 cases, seven of them in the scalp, but no intracranial extension was indicated. An infant with congenital generalized fibromatosis had one of 31 tumors located in the cranial dura. Cranial fibromatoses are slowly growing lesions, typically affixed to the scalp, muscle, and bone. They tend to invade along fascial planes and may destroy bone. Their recurrence rate is as high as 52%.

Desmoid Tumor Originating in Scar Tissue. Some soft-tissue desmoid tumors arise from old scars. Extrapinal desmoid tumors within laminectomy scars were described by Gonatas in his Case 3, and by Wyler and Harris. This latter tumor had blended into the low cervical muscles and fascia; it recurred once after partial excision. An extradural fibromatosis developed within a frontal craniotomy scar of a child who had had a glioma irradiated with 4500 rads.

Diagnosis of Present Cases

When comparing our cases with the differential diagnoses, one encounters the least difficulty in identifying the tumors in Cases 3 and 4. The lesion in Case 4 was a fibroblastic meningioma which had formed excessively large discrete masses of collagen. In Case 3, virtually no meningiomatosus tissue was left. This lesion was thought to be a desmoid tumor on first sight, but its fine structure was indistinguishable from that of a meningioma. It was also similar to the collagenous portions of the tumor in Case 4. Our diagnosis of Cases 3 and 4 agrees with that given by Ghatak for a spinal collagenous mass. We did not find the excessively large collagen fibers described by Ghatak, but we have seen such fibers on occasion in other meningiomas. When reassessing Case 3 it was felt that the patterning of the collagen fascicles still mimicked the structure of the cell streams in a meningioma. One of us has seen a number of small, completely collagenized, dural nodules, found incidentally at autopsy, which all had the same whirly pattern of collagen fibers. These were thought to be meningiomas that became collagenized before attaining their full growth potential.

The tumors in Cases 1 and 2 cannot be dismissed as meningiomas with the same ease. The tissue structure of these tumors was quite unlike that of a meningioma. The random disposition of fibroma-like and smaller myxomatous zones within a compact, hyaline collagen was reminiscent of a desmoid fibroma of bone. None of the cellular islands was unequivocally meningiomatics. The short interval before the tumor recurred in Case 2 was quite unexpected and was thought to be rather unusual for a highly collagenized meningioma. The behavior of this tumor reminds one of the recurrence seen in some extracranial desmoid tumors after subtotal resection or after radiotherapy. An additional confusing aspect was that the tumor in Case 1 was intramedullary, whereas in Case 2 it was extracerebral, infiltrating the frontal bone. The fine structure of the tumor in Case 2 was reminiscent of that described for intracerebral fibromas or desmoplastic fibromas. We felt justified, therefore, in classifying Cases 1 and 2 as belonging to the desmoid family rather than as meningiomas. One may argue that such a distinction is trivial since both kinds of tumors have the fibroblast as a host cell. Yet, desmoid tumors originating elsewhere in the body differ from meningiomas in so many ways that identification of an intracranial desmoid lesion could have prognostic implications.

Acknowledgment

The authors are indebted to Prof. Yaşargil for permission to review the patients' records.

References


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