Primary leiomyosarcoma of the dura mater

Case report

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A rare case of leiomyosarcoma arising from the dura mater is presented. A definite histological diagnosis was based on the electron microscopic features. Extensive removal of the tumor and postoperative radiation therapy resulted in complete remission. The etiology and the origin of this tumor are discussed.

**KEY WORDS** • leiomyosarcoma • dura • brain neoplasm

Although leiomyosarcoma may develop from any site where smooth-muscle cells exist, the most common locations are the uterus, retroperitoneum, mesentery, and mediastinum. The head and neck region is one of the rarest sites and only a few cases of leiomyosarcoma arising from the paranasal sinus and hypopharynx have been reported. To our knowledge, leiomyosarcoma originating in the skull or the dura mater has not been reported previously. This report concerns a case of leiomyosarcoma developing from the blood vessels of the dura mater and extending extracranially through the squamous suture to form an encapsulated mass in the temporal muscle.

**Case Report**

This 73-year-old man first noted a right temporal mass about 1 year before admission, and presented to our hospital because of recent rapid growth of the mass.

**Examination.** The results of the neurological examination were normal. An immobile, elastic-hard mass about 3 cm in diameter was palpated subcutaneously in the right temporal region. There was neither pain nor tenderness. All blood laboratory findings were within normal limits.

Computerized tomography (CT) scans disclosed a space-occupying calcified lesion lying in the right temporal muscle. The mass was strikingly enhanced with contrast medium. The underlying skull and intracranial cavity appeared normal (Fig. 1). Angiography revealed a hypovascular mass, which was not fed by the superficial temporal artery or by the middle meningeal artery. A bone scintigram with technetium-99m-HMDP was normal except for uptake in the right temporal region. A gastrointestinal tract examination, chest x-ray film, whole-body CT scan, gallium scintigram, and abdominal ultrasonogram did not reveal any other primary or metastatic lesion.

**Operation.** At surgery, the scalp was normal and the surface of the muscle fascia showed no evidence of tumor invasion. The mass was removed from the temporal bone complete with the adjacent temporal muscle. The periosteum connected with the tumor capsule was found to enter the squamous suture (Fig. 2). The periosteum was excised. Because tumor tissue was noticed at the resected margin of the periosteum, the temporal bone was removed. The inner and outer surfaces of the excised bone flap were normal in appearance, but marked tumor invasion was recognized in the diploe. The tumor was densely and extensively adherent to the surface of the dura mater. This part of the dura mater was excised except for the segment over the sigmoid sinus that the tumor had invaded. The inner surface of the dura mater and the surface of the cerebrum were normal macroscopically. The operation was completed with duraplasty and cranioplasty.
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**Pathological Examination.** Macroscopic examination of the resected specimen revealed that the tumor was circumscribed by a thick capsule. The tumor capsule was continuous with the periosteum and entered the squamous suture with the tumor tissue.

Hematoxylin and eosin (H & E) staining of the tumor specimen obtained from the temporal muscle demonstrated long spindle-shaped cells with clear nucleoli densely arranged in a streaming pattern. Many mitoses and a certain degree of calcification were observed, but neither osteoblasts nor chondroblasts were present in the specimen (Fig. 3 upper left). Silver impregnation staining showed abundant collagen and reticulin fibers in the intercellular space and rather fewer in the perivascular region, suggesting marked periluminal tumor cell proliferation. These findings implied that this tumor was of blood vessel origin (Fig. 3 lower right). The specimens from the areas of dural invasion (Fig. 3 lower left) and bone invasion (Fig. 3 upper right) disclosed the same findings.

Electron microscopic examination disclosed myofilaments with dense bodies (Fig. 4 left), pinocytic vesicles, and basement membrane material around the cytoplasmic membrane (Fig. 4 right). All of these features are characteristic of smooth-muscle cells. A diagnosis of leiomyosarcoma was therefore made.

**Postoperative Course.** Postoperative radiation therapy was administered with a total dose of 60 Gy to the temporal region. The patient was discharged with no residual tumor visible on CT examination.

**Discussion**

Sarcomas arise from tissues of mesodermal origin and account for 0.1% to 0.6% of all primary intracranial tumors. Although leiomyosarcoma can develop from any site where smooth-muscle cells exist, it usually originates in the uterus or the retroperitoneum. In this case, precise macroscopic and microscopic examination was required to establish the site of tumor origin, since the sarcoma lay in the dura mater, the skull, and the temporal muscle. A diagnosis of intramuscular tumor was made before surgery, but this was abandoned postoperatively because the tumor was circumscribed by a thick capsule macroscopically and revealed no invasion of the muscle microscopically.

If leiomyosarcoma develops from the cranium or intracranial contents it is possible that it originates in the smooth-muscle cells of blood vessels. Matsutani, et al.,7 reported that the hemangiopericytes of intracranial blood vessels are of smooth-muscle cell origin. It is thus possible that these cells could be the origin of leiomyosarcoma. In this case, silver impregnation staining disclosed rather fewer fibers around the blood vessels and marked periluminal tumor proliferation. These findings strongly supported the possibility that the tumor was of blood vessel origin, and that it had arisen from blood vessels of the dura mater or those of the skull, including the periosteum. There was no definite evidence to confirm whether the tumor had developed from the dura mater or the skull. However, it was strongly suggested that the tumor was of dura mater origin, since it firmly adhered to and invaded the dura mater extensively without destruction of bone. On the basis of this suggestion, the following hypothesis is
FIG. 3. Photomicrographs of the resected specimen. Upper Left: Section from the extracranial mass showing long spindle-shaped cells arranged in a streaming pattern. H & E, × 100. Upper Right: Section from the area of diploic invasion. The tumor cells are clearly separated from the bone tissue. H & E, × 40. Lower Left: Section from the area of dural invasion showing tumor cells diffusely and inseparably invading the dura mater. H & E, × 35. Lower Right: Section from the same specimen as shown in upper left. There are rather fewer fibers in the perivascular region. Silver impregnation stain, × 125.

provided to account for the growth and extension of the tumor. The tumor originated in a blood vessel in the outer surface of the dura mater and did not grow in the dura mater (where connective tissue is dense) but extended into the histologically loose space between the dura mater and the inner surface of the skull; the tumor then entered the squamous suture which is also a histologically loose space and advanced into the less-resistant extracranial space, forming a round mass.

Fibrosarcomas arising from the skull have previously been reported, but few authors have presented electron microscopic findings. It seems difficult to differentiate leiomyosarcoma from fibrosarcoma histologically by light microscopic examination alone. It should thus be emphasized that electron microscopic examination is essential in the diagnosis of sarcoma. Extensive surgical extirpation and radiochemotherapy is the treatment of choice for leiomyosarcoma of the head and neck, as with primary leiomyosarcomas of other organs. It is stressed that early diagnosis and treatment is important, since distant metastases result in a poor prognosis.

References
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Fig. 4. Electron micrographs of the resected specimen. × 26,000. Left: Myofilaments with dense bodies (arrows) are apparent. Right: Pinocytic vesicles (left arrow) and basement membrane material (right arrow) are clearly demonstrated.


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