Dural mesenchymal chondrosarcoma

Case report

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A dural mesenchymal chondrosarcoma is reported in a 19-year-old man. This tumor had features of angioblastic meningioma (meningeal hemangiopericytoma), by light microscopy. However, ultrastructurally this tumor could be distinguished from angioblastic meningioma, and its similarity to extracranial mesenchymal chondrosarcoma was confirmed.

KEY WORDS • mesenchymal chondrosarcoma • chondrosarcoma • angioblastic meningioma • meningeal hemangiopericytoma • brain tumor

Mesenchymal chondrosarcoma is a rare malignant tumor. It was first described arising from bone by Lichtenstein and Bernstein in 1959,14 and was later reported as a primary tumor in extraosseous tissues.9 The tumor shows proliferation of primitive mesenchymal cells with islands of cartilaginous differentiation. Dahlin and Henderson9 were the first to note that in some areas the tumor had features of hemangiopericytoma. This resemblance was later stressed by others,6,9,23 and some tumors originally diagnosed as hemangiopericytoma with cartilaginous differentiation have been reclassified as mesenchymal chondrosarcomas.7,9,21 One such tumor was reported intracranially.7

We recently studied by light and electron microscopy a mesenchymal chondrosarcoma arising from the tentorium. The tumor was compared to previously reported mesenchymal chondrosarcomas, angioblastic meningiomas (meningeal hemangiopericytoma),11 and extracranial hemangiopericytomas; this comparison will be the subject of our report.

Case Report

This 19-year-old man had a history of increasing headaches for 6 months, difficulty in concentrating, and scholastic deterioration. He had transient episodes of diminution of vision, particularly in his right eye. Angiography, at another hospital, showed a lesion on the superior surface of the tentorium, which was interpreted as an arteriovenous malformation.

Examination. Physical examination during his present admission was unremarkable except for a mild right homonymous hemianopia. Routine laboratory examination was non-contributory.

Operations. Through a left parieto-occipital craniotomy, the lesion was approached as an arteriovenous malformation because large arterialized veins were seen on the surface of the occipital lobe. After a partial resection of the occipital lobe, a firm tumor attached to
the surface of the tentorium was encountered. Due to the length of the procedure, a partial lobectomy and devascularization of the tumor was done. In a procedure 1 week later, the highly vascular tumor was removed.

FIG. 1. Photomicrograph of a highly vascularized area of tumor resembling hemangiopericytoma. H & E, ×100.

FIG. 2. Photomicrograph of transition zone between cartilage and undifferentiated portion of tumor. H & E, ×100.

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The postoperative course was unremarkable. Work-up has since failed to reveal extracranial tumor. Multiple pieces of tumor, with an aggregate mass of 41 gm, were sent for pathological study. The tumor had a bosselated surface, and was firm. There was no dural or cerebral tissue attached. It had a gritty sensation and displayed yellowish foci.

**Microscopic Finding.** Sections were studied with hematoxylin-eosin, Masson's trichrome, Bodian, Alcian blue, Wilder's reticulin, and phosphotungstic-acid hematoxylin. For the most part the tumor was very cellular. The cells were spindle-shaped, stellate, or ovoid, with pleomorphic nuclei. Occasional mitotic figures were present. The cells were often arranged in clusters. Sinusoidal channels lined by prominent endothelial cells permeated the tumor. Reticulin outlined the vessels, and sometimes surrounded small groups of cells. The highly vascularized areas resembled hemangiopericytoma (Fig. 1).

Islands of cartilage in various stages of differentiation were present in other areas. The chondrocytes were usually plump, and had pleomorphic nuclei and prominent nucleoli. Occasionally, more than one chondrocyte were seen in a single lacuna (Fig. 2). Transition zones between cartilage, and the undifferentiated portion of the tumor showed increase in ground substance, and apparent transformation of the stromal cells into chondroblasts. Rare foci of metaplastic ossification were scattered throughout the tumor.

**Electron Microscopic Finding.** Formaldehyde-fixed tissue was treated with 5% phosphate buffered glutaraldehyde and postfixed in osmium tetroxide. It was dehydrated in graded alcohol, transferred to propylene oxide, and finally embedded in Epon. Sections 1 μ thick were stained with toluidine blue and selected areas examined.

In the more cellular regions the tumor consisted of groups of round-to-ovoid cells with large nuclei. The nuclei were spherical, and occupied most of the cell. Chromatin was irregularly clumped centrally and some was dispersed around the periphery of the nuclei. Nucleoli were frequently seen. Scattered mitochondria, showing swelling, were present in each cell. The endoplasmic reticulum was slightly dilated. Numerous free ribosomes, and occasional dense bodies were noted. Golgi apparatus, pinocytic vesicles, and base-
ment membranes were not seen. Maculae adherentes (desmosomes) connected contiguous tumor cells. Collagen was only present between clusters of cells and around vascular channels. The endothelial cells were plump, and showed microvilli (Fig. 3).

In areas where the tumor differentiated into cartilage, individual cells were seen in abundant intercellular matrix. They tended to be stellate with numerous stout processes. Nuclei were round with dispersed chromatin. Abundant rough endoplasmic reticulum with focal dilatation was seen. Rarely, tight clusters of filamentous material were present in the cytoplasm. These filaments measured approximately 40 Å. The intercellular matrix was composed of finely granular amorphous material, and contained filamentous material measuring 100 to 200 Å in diameter, with a periodicity of 750 to 850 Å. Gradations between these two cell types were noted.

Discussion

Reference to the literature revealed that primary mesenchymal chondrosarcomas of the central nervous system are rare. In a review of 30 extraskeletal mesenchymal chondrosarcomas, five cases occurred intracranially. In two of these cases the skull bones were involved as well. Since osseous mesenchymal chondrosarcoma can arise in the skull bones, the brain and meninges in these two cases may have been invaded by a tumor primary in bone. An additional mesenchymal chondrosarcoma, arising from the spinal dura, has been reported by Rubinstein.

The presence of highly vascular areas, along with islands of cartilage, in these tumors prompted us to review intracranial cartilage containing tumors including angioblastic meningiomas. The case of

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Fisher, et al.,\textsuperscript{7} which was originally diagnosed as meningeal hemangiopericytoma, and is now considered to be a mesenchymal chondrosarcoma was compared to the single chondroblastic meningioma reported by Cushing and Eisenhardt.\textsuperscript{2} The meningeal hemangiopericytoma described by Kruse (Case 8)\textsuperscript{12} also showed islands of cartilage, and closely resembled the tumor reported by Fisher, et al. It is noteworthy that this was the only tumor in Kruse's series to metastasize. We also found two cases of primary intracranial chondrosarcomas with extracranial metastases\textsuperscript{5,25} which displayed features of mesenchymal chondrosarcoma. In addition, Lynch and Uriburu\textsuperscript{15} reported an "intracranial cartilage containing meningeal tumor" with "many similarities to the so-called 'mesenchymal chondrosarcoma'." A common feature in all these tumors was the presence of a highly vascularized, poorly differentiated stroma, which in areas mimicked hemangiopericytoma. Such areas are also seen in many sarcomas, and probably represent a pattern common to undifferentiated mesenchymal cells. However, the similarity between hemangiopericytoma, and mesenchymal chondrosarcoma does not extend to the ultrastructural level.

Ultrastructurally, pericytes are characterized as having a basement membrane, numerous pinocytic vesicles, and intracytoplasmic filaments. These features are also found in extracranial hemangiopericytomas, with some variations, probably dependent on the maturity of the cells making up the tumor.\textsuperscript{1,10,13,16} They were also reported in cases of angioblastic meningioma (meningeal hemangiopericytoma).\textsuperscript{11,17-19} In the present case, intracytoplasmic filaments and desmosomes were found, but pinocytic vesicles and basement membrane material were absent. In this respect the tumor resembled previously reported cases of extracranial mesenchymal chondrosarcomas.\textsuperscript{8,25}

Even though the number of intracranial mesenchymal chondrosarcomas is small, there is an indication that these tumors have a higher mortality and morbidity than angioblastic meningiomas.\textsuperscript{9} In addition, it is interesting to note that there are only two reported cases of primary intracranial chondrosarcomas with extracranial metastases, and both resembled mesenchymal chondrosarcomas. Therefore, the differentiation between mesenchymal chondrosarcomas, and other intracranial cartilage-containing tumors is of prognostic significance, and this may be helped by electron microscopy.

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References

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