Giant-Cell Fibrosarcoma of Brain

Case Report

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The purpose of this report is to describe an example of this unusual controversial type of tumor in which the component cells as a whole are abnormally large and include a multinucleated form of monstrous size.

Almost all of the reported tumors in the literature are fast-growing with recurrence occurring a few months after the surgical resection. Radium and radiotherapy were used extensively for some tumors and did not seem to help at all. A great majority of these tumors occurred in the cerebral hemispheres, particularly in the frontal lobes. Their occurrence was rather infrequent in the posterior fossa. In general, macroscopically, the neoplasms that extended to the surface of the hemisphere tended to be well-demarcated from the surrounding brain. In 1 case, the tumor was bilateral in the internal capsule without cellular connection, and in another case there was arachnoidal seeding of the neoplastic cells. Only in 1 exceptional patient was there a resectable tumor in the cerebellum, and it was removed completely. The patient was alive 17 years after the onset of the disease.

History and Synonyms

The first reported case was in 1914 by Schmincke, who used the term "ganglioneuroma amyelineum." Later a few other authors, describing histologically identical tumors, used the terms of "gangliogioma," "ganglioglioneuroma," "spongioblastoma," "spongioneuroblastoma" and "rethelial sarcoma."

In 1956 Züich interpreted tumors of this type as arising from the cerebral blood vessels and called them "monstrocellular sarcoma." Among 2,250 brain tumors, he classified 17 as "monstrocellular sarcoma," with an incidence of 0.8 per cent. Three of these 17 growths metastasized to the lung and heart.

In 1959 Russell and Rubinstein presented 4 cases in which they regarded the tumor as a variety of glioblastoma multiforme. They pointed out the histological similarity between this type of tumor and the giant-cell tumors of tuberous sclerosis which were described by Roussy and Oberling in 1931, called "astrocytoma sous-épendymaire à grosses cellules fusiformes." The authors suggested that the tumors under discussion may be a malignant form of the latter and the presence of reticulin was not sufficient evidence for its interpretation as a sarcoma.

Hitzelberger et al. in 1961 reviewed the available literature and presented 27 cases of their own. Among these 27 cases there were histopathological variations of these tumors from those in which most cells were bizarre giant cells to those in which giant cells were relatively infrequent. The largest number of tumors was in an intermediate group that contained many unusual giant cells, but also had many areas of typical fibrosarcoma.

They demonstrated a network of reticulin that formed the borders of many fibrous cells and giant cells, but was also seen to arise intracellularly in both these types of cells. They used the term "giant cell fibrosarcoma of the brain."

Case History

A 20-year-old, alert, well-oriented housewife and mother of two children was admitted to Pahlavi Hospital on June 26, 1963, because of a headache and a progressive loss of vision.

During several months before entry she experienced a headache increasing in intensity, accompanied by nausea and vomiting. Two months prior to admission the headache became very severe and she began to notice that she could not see well.

Examination. She was a well-developed and moderately well-nourished young woman in no acute distress. There was right facial paresis with bilateral paresis of the 6th cranial nerve. The vision in both eyes was markedly diminished and she had severe bilateral papilledema. Temperature was 37°C, pulse rate was 80 and respirations were 20 per min.

Fig. 1. Ventriculography showing dilatation of ventricular system.

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Fig. 2. Sagittal section through tumor and brain showing compression and infiltration of medulla oblongata by tumor.

Angiography disclosed stretching of the anterior cerebral artery, indicating a dilatation of the ventricular system.

Ventriculography showed dilatation of the 3rd and lateral ventricles and the aqueduct of Sylvius (Fig. 1).

Preoperative diagnosis: probable tumor of 4th ventricle.

Operation. The procedure was begun in an upright position. After incision of the scalp and removal of the posterior arch of the atlas, the patient suffered a cardiac arrest. An extensive open-cardiac massage was performed and the heart started to beat again.

Course. The patient remained comatose and died on July 16, 1963, three days after the operation.

Autopsy. Gross Findings. The brain weighed 1400 gm. There was a well-demarcated cone-shaped tumor, measuring 6.5×4×4 cm., compressing the inferior part of the cerebellum and very loosely attached to it.

Fig. 3. Extreme degree of anaplasia with many bizarre giant cells. Hematoxylin and eosin stain, X100.

Fig. 4. Areas with many multinucleated giant cells. Hematoxylin and eosin stain, X480.
tumor was also compressing the roof of the 4th ventricle and dorsal part of the medulla oblongata. The attachment of the tumor to the medulla oblongata was rather firm and it could not be separated from it manually. The cut surface of the tumor was grayish-pink, moderately firm and exhibited many small cystic spaces (Fig. 2).

**Microscopic Study.** Multiple sections from the tumor and surrounding brain were removed and the sections were stained with hematoxylin and eosin, Gomori's method of impregnation for reticulin, Mallory's phosphotungstic acid hematoxylin and Masson's trichrome stain.

The histological sections showed areas with an extreme degree of anaplasia. There were huge bizarre giant cells having amorphous or pink granular cytoplasm and bilobed, branching or multiple nuclei (Figs. 3 and 4). Some of the large cells were vacuolated and others incorporated smaller cells in their cytoplasm (Fig. 5). Inclusion-like nuclear vacuoles were also present. Large nucleoli were seen singly or in groups. We also noted cytoplasmic connections of these cells to the adventitia of the blood vessels (Fig. 6). There were other areas where the giant cells were much less numerous. The tumor cells were spindle-shaped, having a typical fibrosarcomatous appearance. Transitional cells between the fibroid cells and bizarre giant cells were also noted. Masson's trichrome staining showed collagenous fibers around the blood vessels and in the substance of the tumor (Fig. 7). Gomori's stain for reticulin exhibited a network of reticulin around the blood vessels and in areas without vascular association forming the borders of many spindle-shaped cells and isolated or grouped giant cells. There was microscopic infiltration of the surrounding brain tissue by the tumor cells (Fig. 8).
In many areas the tumor cells were present around the blood vessels. Focal necrosis and calcification were also present.

Discussion

As we have seen, there have been controversial opinions about the histogenesis of giant-cell fibrosarcoma in the literature. In our case, grossly, the tumor was very well-demarcated and resembled a meningioma. However, microscopically, there is structural variation in areas in which most of the cells are bizarre, giant and multinucleated, but in many places the cells are spindle-shaped and indistinguishable from a typical fibrosarcoma. Lymphocyte-like cells, described in the literature and seen in many cases, are very few in our case. In the adjacent brain tissue there is infiltration of the tumor cells, but in many areas the tumor cells are present only around the blood vessels. As suggested by Hitselberger et al.,¹ this seems to be an explanation of the method of propagation of these tumors as well as offering evidence of their sarcomatous nature.

The observation in our case is very much in accordance with the opinion of Zülch, and Hitselberger et al. The presence of reticulin forming the borders of numerous neoplastic cells, vacuoles, inclusion bodies and smaller cells in the bizarre giant cells, unique staining characteristics and cellular structure of these cells, close association of

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Fig. 7. Collagen and fibers of reticulin around the blood vessels, isolated cells and groups of cells. Masson’s trichrome stain, ×100.

Fig. 8. Tumor cells infiltrating brain tissue in area of medulla oblongata. Tumor cells are present also around blood vessels. Hematoxylin and cosin stain, ×100.
the adventitia of the blood vessels and perivascular propagation in the surrounding brain make us believe in the sarcomatous origin of this malignant neoplasm.

**Summary**

A case of giant-cell fibrosarcoma of the brain is described. The tumor was in the posterior fossa, well-demarcated, simulating grossly a meningioma, compressing the inferior part of the cerebellum and superficially infiltrating the medulla oblongata. The tumor had histological characteristics of a fibrosarcoma with areas having bizarre giant cells with an extreme degree of anaplasia. A network of reticulin was present in the tumor. The controversial literature is briefly reviewed and discussed.

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**References**