Fibromatosis presenting as a cranial mass lesion

Case report

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The authors report a case of fibromatosis presenting as a mass lesion following previous craniotomy and radiation therapy. The clinical and pathological characteristics of this rare condition are discussed.

KEY WORDS: fibromatosis, radiation therapy, craniotomy, fibrosarcoma

Fibromatosis is an unusual pathological growth of mesenchymal tissue that occupies an intermediate position between normal reparative scar tissue and malignant fibrosarcoma. It takes the form of a tumor with vigorous cellularity and a mild degree of anaplasia but with only rare mitoses. We present here a case of a young boy who developed such a lesion after bifrontal craniotomy and subsequent irradiation. His lesion was located on the forehead beneath the coronal incision and grew to the size of a tennis ball in a 6-month period. Although fibromatosis has been known to occur in cicatrical tissue after irradiation, this has not been reported before in a neurosurgical context.

Case Report

This 7-year-old boy was admitted to the Neurological Institute of New York in August, 1974, because of an enlarging mass beneath an old craniotomy scar.

History. He was first seen at another hospital in September, 1972, because of somnolence, unsteadiness, and vomiting. He had been the product of a normal pregnancy and delivery and had no family history of neurological disease. He was slow in achieving the normal childhood milestones, walking at the age of 2 1/2 years and never learning more than a few simple words. Evaluation revealed a left frontal lobe tumor which involved the corpus callosum and third ventricle. He underwent a bifrontal craniotomy and subsequently received 4500 rads over a 6-week period to the tumor site. The histopathology of this tumor was that of a subependymal giant-cell astrocytoma.

In January, 1973, he was readmitted because of drainage from the craniotomy wound. Dehiscence soon followed and the infected bone flap and plastic burr hole covers were removed. He was then transferred to a chronic care facility because of his severe psychomotor retardation. In February, 1974, a...
firm swelling at the operative site was noted and its rapid growth prompted admission to the Neurological Institute.

Examination. The patient did not talk but grimaced and rocked himself. Scattered small papules and hypopigmented lesions were noted about the nose and malar areas. He walked with a spastic gait and frontal lobe reflexes were present. There was a 6 × 6 × 5-cm mass in the forehead at the midline, directly beneath the coronal incision (Fig. 1). This lesion was firm, nontender, and did not pulsate.

The initial impression was that this boy suffered from tuberous sclerosis and that the frontal lesion was either a postoperative encephalocele, abscess, or tumor (recurrent or radiation-induced). Dermatological pathology of the biopsied skin lesions was consistent with tuberous sclerosis. An arteriogram revealed the frontal mass to be extra-axial and relatively avascular. The superior sagittal sinus was displaced downward but was patent, and the mass was supplied by the external carotid circulation and a small branch of the left ophthalmic artery.

Operation. Surgical exploration disclosed an irregular, solid, gritty tumor attached to but not penetrating the dura. Pathological examination revealed that this lesion was fibromatosis (Fig. 2).

Discussion

Fibromatosis is a rare mass lesion which has puzzled pathologists. Stout, who had vast experience with soft tissue tumors, considered fibromatosis as neoplastic tissue which continues to proliferate beyond the point of simple reparation and thereby becomes an infiltrating fibroblastic tumor. The tumor is made up of well-differentiated fibroblasts and does not contain the degenerated, glassy appearing collagenous tissue which characterizes the keloid. Differentiation of this tumor from fibrosarcoma is more difficult; generally fibromatosis has more reticulin and collagen fibers and these are more symmetrically arranged. The fibroblasts are more uniform in size and shape and are more regularly spaced. Mitoses are evident in fibrosarcomas but rarely present in fibromatosis. The clinical behavior of fibromatosis is that of a low-grade tumor manifesting persistent growth and local invasion with a very high incidence of recurrence but no tendency to metastasize.

There are a few reports of fibromatosis that involve the scalp, skull, and brain, but none is similar to the present case. Conley, et al., reported 40 cases of fibromatosis of the head and neck, of which five involved the muscular compartments in the temporal and occipital
regions. In the remaining cases the neck, face, sinuses, and oral cavity were the sites affected. None of the scalp lesions had undergone prior operative or radiation therapy. One patient, who had a lucent skull defect, had an antecedent history of trauma to the region 9 years earlier.²

Congenital generalized fibromatosis, a specific entity, may occur in a multitude of sites. One skull lesion has been documented and at surgery the tumor was adherent to the outer layer of the dura but did not penetrate it. In this patient complete spontaneous regression of all the remaining skeletal lesions occurred by the age of 18 months.³

Baker and Adams⁴ reported a case of “fibroblastoma” of the brain in a 10-year-old girl. The tumor was in the frontal lobe and consisted of numerous fine and coarse strands of intertwining collagenous fibers which sometimes fused into dense, homogeneous wavy bundles of tissue that contained only an occasional cell nucleus and appeared quite hyaline. The description of this lesion is not that of fibromatosis, although it may be related.

Several authors state that fibromatosis is most commonly associated with prior radiation therapy.⁴,⁵ The oncogenic effects of irradiation are well known. Radiation has been used to induce tumors experimentally and numerous cases of epidermoid carcinoma and osteogenic sarcoma following therapeutic radiation have been reported. The occurrence of fibromatosis and fibrosarcoma after radiation is quite rare, however, and may be related to excessive dosage.⁶ Of the 206 cases of fibrosarcoma reported by Stout,⁷ only four had undergone prior radiotherapy. Sarcomas after irradiation for intracranial tumors have been summarized by Waltz and Brownell.⁸ These tumors followed massive doses of radiation, occasionally given in multiple courses, and appeared from 3 to 24 years after radiotherapy.

It is possible that fibromatosis after radiation therapy may, with time, develop into fibrosarcoma. In this case the appearance of fibromatosis occurred much sooner after irradiation than the reported cases of fibrosarcoma. These two lesions seem to be related both clinically and pathologically, and it seems plausible that fibromatosis could differentiate into the more aggressive fibrosarcoma.

Fibromatosis should be considered among the differential diagnoses when a rapidly growing mass is encountered in neurosurgical practice after irradiation to the head. Complete excision is essential to prevent the possible tendency toward more aggressive invasion.

The possible relation of this fibrous tumor to tuberous sclerosis in this patient must be considered; however, no propensity for such growths has been documented in tuberous sclerosis⁹ and the combination of a cicatrix and radiation therapy seems to account for the appearance of fibromatosis in this case.

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References


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