Myxofibroxanthoma of the Fourth Ventricle
—Case Report—

Mau Nan CHEN, Shozo NAKAZAWA, Toshiro SHIMURA and Kouzo YAJIMA

Department of Neurosurgery, Nippon Medical School, Tokyo

Abstract

Intracranial fibrous xanthomas are extremely rare. Only 10 cases have so far been reported. A fourth ventricular tumor in an elderly female, identified histopathologically as a myxofibroxanthoma, is described here. After total excision of the tumor, she recovered uneventfully. The histological features and diagnosis of intracranial fibrous xanthomas are discussed.

Key words: myxofibroxanthoma, fourth ventricle, computed tomography scan

Introduction

Fibrous xanthomas of the skin and soft tissues are not uncommon, but such lesions very rarely occur in the central nervous system. To our knowledge, only 10 cases have been reported. Recently, we treated a patient with a fibrous xanthoma arising in the choroid plexus of the fourth ventricle. The tumor also had a myxoid component. It was first discovered on a computed tomography (CT) scan, and the pathological diagnosis was made after surgical extirpation.

Case Report

A 64-year-old female was admitted to our hospital on August 13, 1980 with a 5-month history of dizziness, nausea, and vomiting. Shortly before admission she had developed difficulty in standing and in urinating. About 10 years previously, she was found to have hypertension. Her family history was not contributory.

On admission, the patient was mildly disoriented as to the date and was slightly drowsy. Standing and urinating were difficult. Neurological examination revealed bilateral papilledema and horizontal nystagmus of the right eye, but visual acuity and the visual field were normal. There was slight motor weakness in both the upper and lower extremities, and the finger-to-nose test showed mild bilateral dyssynergia, which was worse on the right than on the left side. The deep tendon reflexes were normal in the arms and the left leg, but were mildly exaggerated in the right leg. A plain CT scan disclosed disappearance of the fourth ventricle and a mild hydrocephalus (Fig. 1 left). However, after injection of a contrast medium, a diffuse, high-density area was discernible in the midline posterior fossa (Fig. 1 right). Vertebral angiography showed neither remarkable changes nor tumor staining. An electroencephalogram showed slightly abnormal patterns. The background rhythm consisted of slow α or θ waves and there was neither laterality nor focal spiking.

On October 8, 1980, following a shunt procedure, the patient underwent a suboccipital craniectomy in the prone position. On the floor of the fourth ventricle, connected to the choroid plexus, we found a solid, vascular, well demarcated tumor. The mass was dark red and oval-shaped, about the size of a ping pong ball (4 × 3 × 3 cm). We totally removed the tumor, after ligating the feeding arteries. Her postoperative course was uneventful. The headache, nausea, vomiting, and horizontal nystagmus disappeared and the cerebellar signs improved. However, walking remained difficult and she was transferred to another hospital for rehabilitation.

Histopathological evaluation revealed a mesenchymal tumor composed of stellate and spindle-
shaped cells, which tended to form a loose, mucin-containing network (Fig. 2). There was no PAS-positive substance in the tumor cells. The tumor was moderately vascular, containing mainly capillaries and a few venous structures. A large number of xanthoma cells with pyknotic nuclei and large, polygonal, foamy cell bodies were also present (Fig. 3). Between the cells, reticulin fibers were abundant (Fig. 4). There were no mitotic cells in the tumor.

Plasma cells and lymphocytes were present throughout the lesion. A glial fibrillar acidic protein immunostain was negative.

**Discussion**

Fibrous xanthomas most often arise in the skin and soft tissues. A few occurrences in other organs have been reported, and there are indications of a recent increase in the incidence of skeletal fibrous xanthomas. There have been only 10 previously reported cases of fibrous xanthoma of the brain and the leptomeninges. The salient features of these cases are summarized in Table 1.

This report, we believe, is the eleventh documented case of intracranial fibroxanthoma and the second reported case showing myxoid changes. In 1973, Kepes et al. documented four intracranial lesions, three of which were fibrous xanthomas of the temporal lobe. Two were cystic and one was solid, and all three patients were boys. The fourth case was a solid xanthosarcoma of the frontal lobe in a 26-year-old female. Two years later, Caillo et al. described two fibrous xanthomas. One patient was a 27-year-old female with a subcortical cystic tumor in the left parietal lobe. The other was a 45-year-old male with an intraventricular solid tumor attached to the...
right trigone. In 1976, Gonzalez-Vitale et al. described a radiation-induced malignant fibrous histiocytoma in the area of the sella turcica, discovered 11 years after irradiation of a pituitary chromophobe adenoma. Three years later, Lam and Colah reported a 24-year-old male with an intradural atypical fibrous histiocytoma arising from the floor of the left anterior fossa. The mass, which did not invade the brain, had many areas of myxoid changes, which accounted for approximately 40% of the entire lesion. Martin et al. reported a 7-year-old girl with a solid fibrous xanthoma in a right parieto-occipital region. In 1981, Abiko and Orita described a 6-year-old boy with a bilateral tumor of the temporal lobes. The left-side mass was resected and found to be a solid fibrous xanthoma; the authors conjectured that the tumor originated in the dura of the base of the middle fossa. Theirs is believed to be the first case of fibrous xanthoma diagnosed on the basis of CT findings. The mass was slightly hyperdense on a plain CT scan and was markedly enhanced, especially in the medial portion, after injection of a contrast medium. In our case, the mass was slightly hypodense on the plain CT scan and was homogeneously and markedly enhanced after injection of a contrast medium.

The fibrous xanthoma in our patient contained the following components: fibroblast cells arranged in a pinwheel or storiform pattern; foamy cells; Tonton type giant cells; and reticulin fibers surrounding the individual tumor cells. Glial fibers were absent and Cajal's astrocyte impregnation test was negative. These are the characteristics that Kepes proposed for the histological definition of fibrous xanthomas or xanthosarcomas of the brain, as distinct from tumors of the soft tissues.

In the case we have described, the appearance of the lesion was similar to the descriptions given in most of the reported cases. However, in addition to the basic histological pattern of interlacing bundles of spindle cells, there was prominent plasma and lymphocyte infiltration, with mucin stored between the tumor cells. We feel that "myxofibroxanthoma" is a better designation for this lesion, which was similar to that described by Lam and Colah. Their patient's tumor was completely intradural, without involvement of the leptomeninges or the brain parenchyma. Our patient's tumor originated in the floor of the fourth ventricle and was connected to the choroid plexus. Hence, we think the tumor arose from the mesenchymal stem cells of the choroid plexus.

Only one of the reported fibrous xanthomas recurred, 14 months after the initial attempt to remove the lesion. The 12-year-old male patient, however, was free of symptoms 3 years after the second resection of the lesion. The relationship between the histopathology and the biological behavior and prognosis of intracranial fibroxanthomas is not well understood. To elucidate this relationship and adequately treat these tumors, we need long-term follow-up of
a large number of patients.

Acknowledgment

The authors wish to express their gratitude to Professor Y. Oyake, Department of Neuropathology, Brain Research Institute, Niigata University, Niigata, for reviewing the microscopic preparations.

References

2) Albert PS, Kane LH: Malignant fibrous xanthoma of retroperitoneum. Urology 1: 460-461, 1973

Address reprint requests to: M.N. Chen, M.D., Department of Neurosurgery, Kosei General Hospital, 5-25-15 Yayoi-cho, Nakano-ku, Tokyo 164, Japan.