Fibrosarcoma of the scalp

Case report


Departments of Surgery, Radiology, and Radiotherapy, Lagos University Teaching Hospital, Lagos, Nigeria

A parieto-occipital scalp tumor, noted in a Nigerian boy during the first month of his life, gradually increased in size, and after two attempts at partial removal there was a rapid increase in its size. When he was 17 years old, it measured $21 \times 17 \times 16$ cm. There was no underlying skull defect. Complete excision of the tumor was achieved, and on histological examination it was found to be a fibrosarcoma. The patient was given a course of radiotherapy following wound healing. He remained well for 5½ months but then had manifestations of small local recurrence and distant metastases that responded only minimally to another course of radiotherapy combined with cancer chemotherapy.

Key Words • fibrosarcoma • scalp • congenital fibroma • malignant transition

In the world literature there are only a few examples of primary mesenchymal tumors of the scalp, such as leiomyoma, leiomyosarcoma, dermatofibroma, dermatofibrosarcoma, malignant angioendothelioma, plexiform neurofibroma, and fibrosarcoma. The rarest of all is fibrosarcoma, and to this extremely limited list we add a case of our own. Because of its rarity, exact statistics, its mode of evolution, and a comparative account of its histological and biological characteristics are generally lacking in reported cases, although some of these aspects have been highlighted.

Case Report

This 17-year-old Nigerian boy was admitted to Lagos University Teaching Hospital (LUTH) on January 20, 1977, because of a large fungating mass over the posterior half of the vertex. This mass had been present since birth, although it was quite small when he was an infant. There had been a slow and steady increase in the size of the mass, but it had grown more rapidly after an attempt to excise it 2 years before the present admission. It further increased in size following another attempt to remove it 6 months prior to admission. A few days before admission, the mass bled profusely, and this episode precipitated his admission to LUTH.

Examination. The patient was a healthy-looking boy with no anemia, lymphadenopathy, hepatomegaly, or splenomegaly. The general examination was unrevealing with no stigmata of neurofibromatosis. He had an
extensive scalp lesion that measured 21 × 17 × 15 cm. It occupied the posterior half of the vertex, extending bilaterally in the parietal and occipital regions (Fig. 1). Its superficial surface had ulcerated. It was extremely soft, vascular, and friable at its summit, but was hard and firm at its base. Engorgement of scalp vessels at its base indicated its high vascularity. The edges of the ulcer at the summit were not raised and rolled out but were rather inverted.

Blood biochemistry and hematological examination revealed no abnormality. Plain skull films clearly showed that the underlying bone was not involved. Internal and external carotid angiography revealed a highly vascular lesion of the scalp, which derived its blood supply almost exclusively from the external carotid circulation. There was no communication between the external and internal carotid circulation across the calvaria at the site of the lesion. The external carotid circulation was clearly hyperdynamic. Vertebral angiography was not carried out.

**Operation.** Two days after angiography, the patient underwent surgery. Early ligation of feeding blood channels proved extremely helpful in minimizing blood loss during the operation. The mass was found to be extremely soft, friable, and vascular, and appeared to have involved some of the surrounding skin near the ulcerated area at the summit of the lesion. The superficial part was sucked out. The deeper part, which was hard, relatively less vascular, whitish in color, and bosselated at its periphery, was easily separated from the surrounding skin, although no distinct capsule could be discerned. The base of the tumor was free of pericranium but appeared to have involved the galea aponeurotica. All the involved tissue was removed, and on examination with the naked eye the neoplasm appeared to be totally removed. The scalp was closed quite easily in a single layer. The patient was transfused with 2 pints of blood to compensate for the blood loss.

**Pathological Study.** Gross examination of the excised specimen showed a grayish-white tumor tissue in several parts, together measuring 4 × 13 × 8 cm, and weighing 390 gm. The cut surface showed gelatinous changes. Microscopically, it consisted of moderately dense proliferation of predominantly mature fibroblasts that were separated by reticulin and collagen fibers arranged in an interlacing fashion. The tumor cells were sufficiently differentiated, although mitotic activity was rather intense and showed some variation in different parts of the specimen. Only one giant cell could be seen. Histologically it was unequivocally a fibrosarcoma (Fig. 2).

**Postoperative Course.** Recovery was uneventful except for local infection which required premature removal of stitches and drainage of pus underneath the scalp. Bacteriological examination of the pus yielded multiple organisms along with *Staphylococcus aureus*, which was sensitive to Ampiclox (ampicillin and cloxacillin). A course of this antibiotic for 7 days and drainage of pus promoted healing except in the tiny central part of the wound. There appeared to be excessive granulation near the edges thought to be due to local sepsis. However, the wound healed well after 6 weeks. Three weeks after the operation, the patient was given a course of radiotherapy. Six weeks postoperatively, he was discharged home. Four months after discharge, on
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Fig. 2. Photomicrograph of the histological section showing fibrosarcoma characterized by frequent mitotic figures. H & E, X 750.

follow-up examination, he was well, and there was no sign of local or distant recurrence.

Second Admission. There was a dramatic decline in his clinical condition 5½ months after the operation. He was readmitted to LUTH with a 1-week history of persistent productive cough, occasional hemoptysis, pain in his chest, dyspnea on exertion, generalized aches and pains in the body, anorexia, and weight loss. On examination he was found to be pale and anemic, rather anxious and apprehensive. His liver was moderately enlarged; chest expansion was poor at both bases and there was a suggestion of bilateral pleural effusion. He noted tenderness over the mid-dorsal spine. There was a small nodule, 1.25 X 0.5 X 0.5 cm in size, in the center of the operative scar. He had no neurological deficit.

Plain skull films were normal, but the chest films showed multiple pulmonary opacities and bilateral pleural effusion. Mid-dorsal spine x-ray films were indicative of osteolytic lesions in the T-5 and T-6 vertebral bodies. Hemoglobin was 7.6 gm%, but improved on blood transfusion. A pleural tap yielded serosanguinous fluid that did not contain neoplastic cells. Sternal marrow examination revealed megaloblastic anemia and non-specific plasmacytosis, megakaryocytosis, and eosinophilia.

He was treated with repeated pleural taps, blood transfusion, and another course of radiotherapy combined with cancer chemotherapy (vincristine sulphate, cyclophosphamide, and prednisolone in combination). He improved slightly and was discharged home on September 8, 1977. The nature of
the nodule in the operative scar was not con-

firmed by histological examination. Since his
discharge he has been lost to follow-up

review.

Discussion

Considering the fact that there is a
generous disposition of collagen and fibrous
tissue in various layers of scalp, the rarity of
fibrosarcoma in this situation is rather sur-

prising.9,17

In 1854, Virchow reported the first case of
leiomyoma cutis,5 and subsequently only a
few examples of leiomyosarcoma, der-
matofibroma, dermatofibrosarcoma, malig-
nant angioendothelioma, pseudosarcoma
(atypical fibroxanthoma), congenital ele-
phantiasis nervorum (plexiform neurofi-
broma), and fibrosarcoma were added to the

literature.4,6,10

Histological as well as macroscopical fea-
tures distinguish pure fibrosarcoma from
other mesenchymal tumors. In general,
leiomyoma, leiomyosarcoma, dermato-
fibroma, and dermatofibrosarcoma of the
scalp do not attain massive dimensions as do
plexiform neurofibroma and fibrosarcoma.18
The one exception is a case of leiomyoma
with dimensions of 9 × 6 × 5 cm reported by
Stout.14

Fibrosarcoma is a very vascular tumor, but
never as vascular as malignant angioen-
dothelioma. The latter may be extensive but
is relatively flat and not as protuberant as
fibrosarcoma; it is accompanied by extensive
discoloration of the overlying skin surface
(purple, black, and port-wine).

Plexiform neurofibroma arises from the
sheath element of one of the nerves of the
scalp, and can attain an enormous size with
the passage of time.11 The striking feature is
an underlying skull defect, regarded as a
manifestation of inherent mesodermal
dysplasia rather than erosion.7,8 Skull defect
is not a feature of fibrosarcoma.

Histologically all these mesenchymal
tumors are different from fibrosarcoma. Der-
matofibrosarcoma resembles fibrosarcoma;
however, the former arises from the corium.
The occurrence of spindle cells associated
with collagen production at the periphery of
the tumor, and more cellular, rounded or
histiocytic elements in the central portions
with cellular distribution in a storiform or
cart-wheel pattern, according to Taylor and
Helwig, is a characteristic feature of der-
matofibrosarcoma.15

Pure fibrosarcoma, on the other hand, is
rich in fusiform fibroblasts showing varying
degrees of anaplasia in a collagenous matrix,
the relative quantity of which determines the
texture and consistency of the tumor.
Histiocytic elements and deposition of
hemosiderin are lacking.

The etiological role of different factors or
agents is difficult to ascertain in an individual
case. In general, radiation from X-ray studies
or radioactive materials appears to have a
role in evoking such tumors as fibrosarcomas,
osteosarcomas, and leukemias.16 There is
abundant experimental evidence to prove this
hypothesis. Another likely etiological factor
is malignant transformation in a pre-existing
neurofibroma of the scalp as shown in a case
reported by Romieu, et al.12 Similar transfor-
mation of a fibroma is not ruled out in our
case.

Trauma, chronic irritation, and chronic in-
flammation play some role in individual
cases, although in general their role has been
somewhat discounted by workers like Willis.
Fibrosarcoma is rare following head injury.
Although reparative and chronic inflam-
matory changes are everyday occurrences,
mesenchymal neoplasia is rare.17 However, in
certain cases such as ours it is difficult to dis-
entangle the role of trauma and chronic in-
flammation.

The long-standing history and congenital
nature of the lesion in our patient preclude the
etiological role of any carcinogenic agent
such as exposure to irradiation. It appears
that the lesion was a simple fibroma which
steadily got bigger with the passage of time,
but perhaps attained neoplastic attributes
when subjected to operative trauma 2 years
previously. This hypothesis seems plausible,
as a rapid increase in size of the lesion dated
back to that episode.

Fibrosarcomas of the scalp grow slowly
and tend to recur only if tumor removal has
been incomplete. Except in very advanced
cases, they rarely metastasize. They are also
relatively radiosensitive. Thus, in early
stages, if the operative removal is complete as
judged by the naked eye, and is followed by a
full course of radiotherapy, a complete cure is
possible. However, when the tumor is far ad-
vanced in malignancy, as is evidenced by
rapid and massive expansion in a short space
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of time and frequent mitoses on histological section, radical surgery combined with radiotherapy will be more palliative than curative. It will only delay the inexorable and inevitable advance of the disease process for some months, as is exemplified by our case. However, this palliation, even though only for a few months, is much appreciated by the patient because of freedom from the weight of the mass lesion, fungation, maceration, sepsis, and frequent local hemorrhages.

In the early phases of evolution of the lesion, an incomplete excision not only fails to guard against a recurrence but may provoke a malignant change in a benign lesion, as illustrated in the case reported by Romieu, et al.\(^\text{12}\) In that case there were several local recurrences of local neurofibroma following incomplete excision, with eventual development of fibrosarcoma of Darier-Ferrand. Later, a wide but incomplete excision, local skin grafting, and extensive radiotherapy could not ensure against the final fatal recurrence. One should aim for early detection and total removal; radiotherapy should be used as an adjuvant.

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References


Address reprint requests to: Abdul B. Chaudhari, M.B., B.S., F.R.C.S., Department of Surgery, Lagos University Teaching Hospital, Private Mail Bag 12003, Lagos, Nigeria.