JUVENILE FIBROMATOSIS*

A CASE REPORT SHOWING INVASION OF THE BONE

BY

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This paper reports a growth occurring at the angle of the mandible of a 6-year-old boy. The growth consisted of mature-looking fibroblasts with a generous collagenous matrix. The course of this patient up to the present is described as it portrays some of the problems encountered in children with fibrous growths. Despite recent articles on the nature of such growths there remain many instances where it cannot be decided on histological grounds alone whether they are true tumours or fibrous dysplasia. If they are tumours they cannot be separated with certainty into benign and malignant groups by their microscopic appearance. It cannot even be proven whether they are growths of mature fibroblasts, i.e. well-differentiated mesenchymomata, or growths of peripheral nerve cells, i.e. benign schwannomata (neurofibromata). The clinical course of such growths is not totally predictable. Hence the clinician is confronted with the problem of determining how best to manage them; steering a middle course between the Scylla of inadequate and belated removal and the Charybdis of unnecessary mutilation.

Case Report

W.H., a 6-year-old white boy, presented in 1957 with a painless mass at the angle of the right mandible. This mass was thought by his mother to have been present for about six months. She had noted little change in size. On examination the boy was normally developed with no other relevant physical findings and no relevant family or past history. He gave no history of trauma in this area. The mass was approximately 2 by 2-5 cm. and was situated at the angle of the right mandible below and a little anteriorly, deep rather than superficial. It was firm and slightly moveable in relation to the bone. It felt contiguous with, rather than continuous with, the bone. It was painless except for slight soreness on firm palpation. Radiographs showed a spicule of bone on the lower inner border of the mandible at about the site of the mass. No other abnormalities were seen. The chest was clear and blood and urine analyses were normal. On July 6, 1957, the mass was explored through a crease incision in the neck below the mandible. It proved to be ill-defined, whitish, firm fibrous tissue resembling a deep scar. A biopsy was taken and the frozen section reported as 'neurofibroma—benign'. The remaining portion of the mass, which extended posteriorly to the upper part of the sternomastoid muscle, deeply into the parotid gland and anteriorly into the submaxillary gland, was removed piecemeal. The bone spicule noted in the radiograph was found in the deep part of the mass and removed. Unfortunately the cervical branch of the facial nerve was injured and a weakness of the corner of the mouth ensued. Recovery was otherwise uneventful. The permanent sections were considered to confirm the diagnosis of benign neurofibroma.

Three months later there was no palpable mass in the area. Within a year a recurrence was noted. In August 1958, the mass was 2-5 by 3 cm. in the same location as before. It felt as before though it seemed more firmly fixed. Radiographs of the mandible showed a slight depression where the spicule had been removed and an area of rarefaction about 1 cm. in diameter near this site. A radiograph of the chest was clear.

In September 1958, a wide block dissection of the mass was performed. The previous scar was excised, although it was below the recurrence. The skin was reflected superficial to the platysma. The block of tissue containing the mass was dissected off the mylo-hyoid muscle from the hyoid bone upwards. The sternomastoid muscle was not involved and a lymph node in the jugular chain near the mass was removed for section. It was shown to be 'reactive'. After cutting across the inferior part of the parotid gland and freeing the submaxillary gland the mass was free except for its attachment to the mandible. As the angle of the mandible was cut across, after fracturing the outer table, a cavity was opened. This contained a grey homogeneous material. The cut across the mandible was altered to include bone beyond this cavity. The patient made a good recovery. The pathological report was again benign neurofibroma. The grey material within the mandible was histologically identical with that without. On examining the gross specimen it was seen that the mass within the mandible was connected to the larger mass without by a narrow neck of tumour tissue penetrating through an almost microscopic hole in the cortex.

Following this the mass recurred within six months.

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Fig. 1.—Patient showing second recurrence in February 1960.

At this time a dimpling was noted in the cheek, appearing like the scar of a penetrating injury. This did not at first seem to be connected to the recurring mass at the angle of the mandible. The recurrence grew slowly becoming more prominent and growing forward and upward until it became continuous with the dimpled area in the cheek. The patient was seen by Dr. A. P. Stout of New York in consultation. He considered that this case fitted into his classification as ‘juvenile fibromatosis’. He considered that the growth was likely to be self-limited and would stop growing if it could be left long enough. He considered that it was benign and that there was no danger of the cell pattern becoming more malignant or of metastases occurring. Over the next year the mass continued to grow by measurement and by February 1960, radiographs showed definite evidence of further involvement of the mandible (Figs. 2 and 3). After further consultation with Dr. Stout it was agreed that radical excision was now essential, whether the mass was considered benign or malignant or even simply fibromatosis. This decision was taken reluctantly as radical resection entailed removal of a large portion of the right mandible. A tube pedicle graft was raised from the right chest in stages and by June 1960, the right cheek and mandible with the mass were resected (Figs. 4 and 5).

Discussion

This case presents the problem of a child with a recurring mass which when excised and examined appeared to be little more than ‘scar tissue’. It consisted of mature-looking fibroblasts with elongated pointed nuclei, without evidence of mitoses normal or abnormal, and without palisading. Though some of the nuclei appeared ‘wavy’ there was inadequate evidence to classify this unequivocally as a schwannoma. There was no demonstrable nervous element in it. This mass had ill-
TABLE

Fibromatoses (tumour-like fibrous tissue proliferation):
Juvenile fibromatosis: (a) Cutaneous
(b) Deep
Plantar and palmar fibromatosis
Desmoid tumours
Progressive myositis ossificans
Congenital generalized fibromatosis
Sternomastoid tumour of infancy
Keloid

Certain bone lesions:
Fibrous dysplasia
Non-osteoid osteoma or non-osteogenic fibroma
Periosteal fibroma; periosteal desmoids

Dermatofibrosarcoma protuberans—fibrosarcoma well differentiated
(including periosteal and parosteal sarcoma)

Tumours of the peripheral nervous system:
Benign Malignant
Neurilemmoma Malignant schwannoma
Neurofibroma
Multiple neurofibromatosis

defined borders and appeared to be either infiltrating or invading surrounding tissues. By looking at the sections alone and without considering the course and the anatomy of the growth, it would be impossible to state whether it was scar tissue, a desmoid, a neurofibroma or even a well-differentiated fibrosarcoma. A brief classification of growths which may have such an appearance is given in the Table. Most of these are easily classified on their anatomical relations, distribution and clinical course. The present case does not fit any of the types of fibromatosis because of its unique penetration of the bone. Fibromatous dysplasia occurs in the medulla of bone and is what Jaffe (1958) called desmoplastic fibroma. This can only be distinguished from well-differentiated fibrosarcoma by the plumpness and roundness of the nuclei, but this remains within the bone. The periosteal desmoids that Kimmelstiel and Rapp (1951) reported were small lesions eroding the cortex and they did not recur after
excision. Nor can it be proved that the periosteal or parosteal fibrosarcoma has invaded the medullary cavity unless the cortex is first destroyed. This lesion could well have started as a periosteal lesion and the presence of a spicule of bone on the mandible when the lesion was first seen is suggestive.

The object of this presentation is not concern over the name that should be applied, but over the best management of such cases. By 'such cases' I refer to children showing areas of desmoplastic hyperplasia no matter in what part of the body they occur. From a study of the cases reported and from talking with Dr. Stout who has been interested in this type of lesion for some time the following conclusions seem justified: (1) The lesion, whether a true tumour or not, does not metastasize; (2) it tends to recur but the cell type remains the same with each recurrence (it does not become less well differentiated); (3) local excision, even piecemeal, may cure the lesion and, this being so, radical excision should not be undertaken at the first attempt, although if it is possible to take a wide margin of normal tissue without undue mutilation, this should be done; (4) recurrence should be treated with wide local excision without lymph node dissection; (5) even such an excision may not be successful and the lesion may kill by local invasion. If the lesion is in the neck, the problem is more precariously balanced because wide excision prejudices vital structures, but recurrences may, as in this case, require even more mutilating surgery. Stout (1954) reported six cases of juvenile fibromatosis occurring in the neck of children: one was excised piecemeal and did not recur; one was excised in the surgeon's office and did not recur; one was excised, recurrent, but then stopped growing without further treatment; two were excised, recurrent, but were cured by re-excision; one was excised, recurrent and it was impossible to re-excite; it killed by local invasion.

Radiotherapy seems to have had little effect on these lesions, but it should be borne in mind that fibrosarcomata have been reported in areas that have been radiated some time previously. More interesting are the possible effects of hormones. Much experimental work has been done by Lipschütz and Vargas (1941) and Lipschütz and Grismali (1944) who showed that oestrogens were fibromatogenic in guinea-pigs and that progesterone and certain adrenal cortical steroids were anti-fibromatogenic. A recurring abdominal desmoid in a 2½-year-old girl has been treated with cortisone
by Panos and Poth (1959) with apparent success. Bullock (1955) mentioned using testosterone in a 21-year-old woman with a recurring extra-abdominal desmoid starting in the trapezius which involved many structures, including the carotid sheath. He thought that it induced some control of the growth. The idea of controlling desmoplastic hyperplasia with steroids deserves more study.

**Summary**

A case of fibrous tissue growth occurring at the angle of the mandible in a 6-year-old boy and recurring after excision is reported. This case was unusual in that the mandible was not only eroded at the second recurrence but that the medullary cavity was invaded at the time of the first recurrence. The nature and management of desmoplastic growths are briefly discussed.

**References**


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