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REGIONAL NEUROFIBROMATA

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NERVE tumours may be found anywhere along the course of a nerve or its tributaries. These may be single or multiple, and may also be superficial or deep.

The tumours are simple and encapsulated and similar in structure to acoustic nerve tumours. The exact origin is still disputed, hence the multiplicity of terminology. Verocay (1910), Ewing (1953) and R. S. Murley (1948) refer to them as Neurinoma, whereas, Somers (1952) and Stout (1935) call similar types of tumours, Neurillema. Guggenheim (1953), Ehrlich and Martin consider that the cells of Schwann play an important part in the structure of the tumour and refer to it as a Schwannoma. If necessary, the superficial tumours can be easily dealt with, but when more deeply placed, particularly in the skull cavities such as the internal auditory meatus or intraorbital region, it becomes a more serious surgical problem.

Case Report

A woman aged 40 came to see me in May, 1951. She complained of blockage of right ear for several months, which had gradually become worse with now, as well, a feeling of pressure in the right ear and right side of her head. Nine years previously, a tumour had been removed from the right side of her neck, and again six years ago, from the right eye and again the right side of her neck.

She now had a swelling in the right temporal region, which was smooth, firm, not attached to skin, and which did not move completely freely over the bone, but was tender with swellings in the scalp behind the right ear which were not attached to skin or bone. One of these was pushing the posterior

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meatal wall forward, almost completely obliterating the lumen. The mouth and nose were normal. The left ear was normal with good hearing. The right external canal was narrowed by the bulging forward of the posterior meatal wall with a marked reduction of hearing on that side. The left Rinne was normal but the right Rinne was negative with the Weber lateralized to the right. She therefore, had a right conductive deafness. Two scars were noticed over the right sternomastoid with an oval swelling 1 in. by ¾ in. under lower scar. This was tender and not fixed to deep structures. The right eye was pushed forward, downward and medially. Eye movements were slow, hesitant but full. Eyesight normal. No papillœdema or nystagmus.

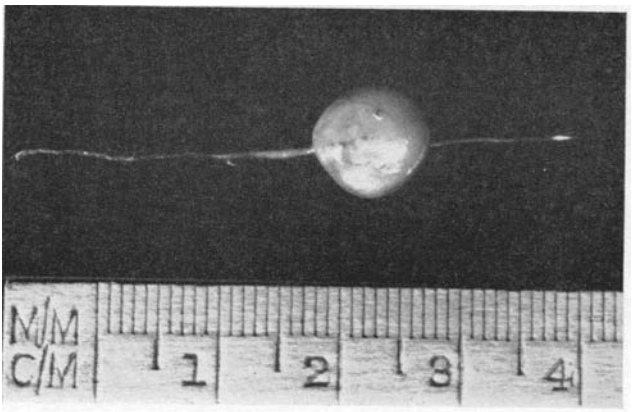


FIG. 1.

A tumour displacing nerve. For proper removal, portion of the nerve had to be sacrificed.

Blood Investigations

1.5.51.

Hb	90%	W.B.C. per c.mm.....	8,300
R.B.C. per c.mm.	4,830,000		
CI	0·95		

Differential White Cell Count

Polymorphs	61·5%	Lymphos.	32%
Monocytes	3·5%	Eosinos.	2%
Basos.	1%		

X-ray of Skull Request

1.5.51

Swelling right temporal region. ? fibroma lipoma ? erosion subjacent bone.

Radiologist's Report

9.5.51

No bony lesion detected.

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Operation

Incisions were made over the swellings, back of right ear, right side of face and scalp. Tumours were found in the deep fascia in the middle of nerves, they were freed from surrounding tissue until the nerves could be seen entering and leaving the mass in each case. The nerves were so widely displaced by the tumours, that it was impossible to free them and consequently had to be severed on either side of each mass. Fig. 1. Hearing now normal.

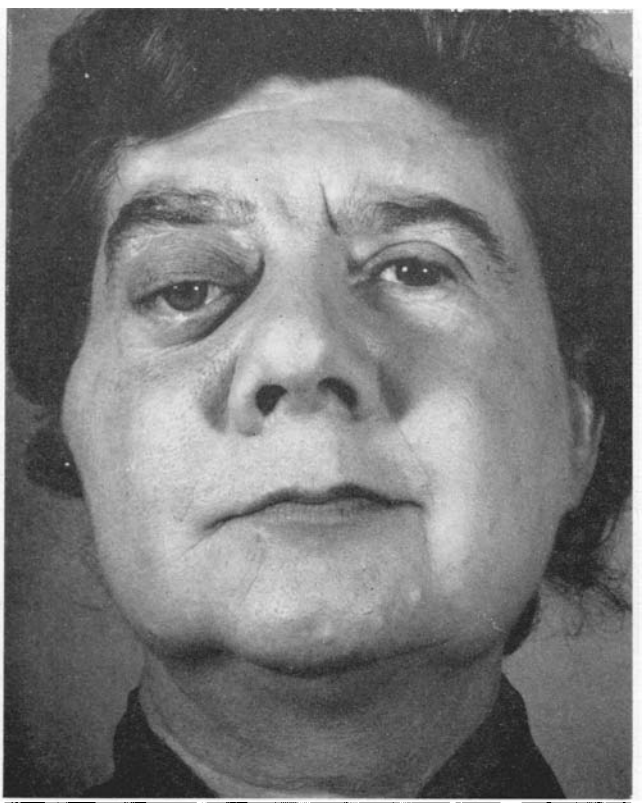


FIG. 2.
Protrusion of the right eye and swelling of the right cheek.

Pathological Report. Dr. Lucas

9.5.51

Specimen: Biopsy of swellings in neck, face and scalp. The nerve tumours are neurofibromas circumscribed and arising from the nerve sheaths. They are cellular in the centre which is composed of nerve sheath cells and are surrounded by a capsule of fibrous tissue in which there are a few fat cells. The reticulin pattern is typical of neurofibromas.

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She was seen again at intervals, but by 1956, swellings in neck and face had enlarged and become more painful. The right eye was protruding very much more, so that she found it difficult to cover it with the upper lid completely. (Fig. 2). The eye movements were very limited. The vision in this eye, however, was normal and we presumed that an intra-orbital tumour was displacing the eye and limiting its movements (Fig. 2). At this juncture, she was seen by Mr. A. P. Cross, Ophthalmologist to R.N. E.N.T. Hospital, who reported as follows:

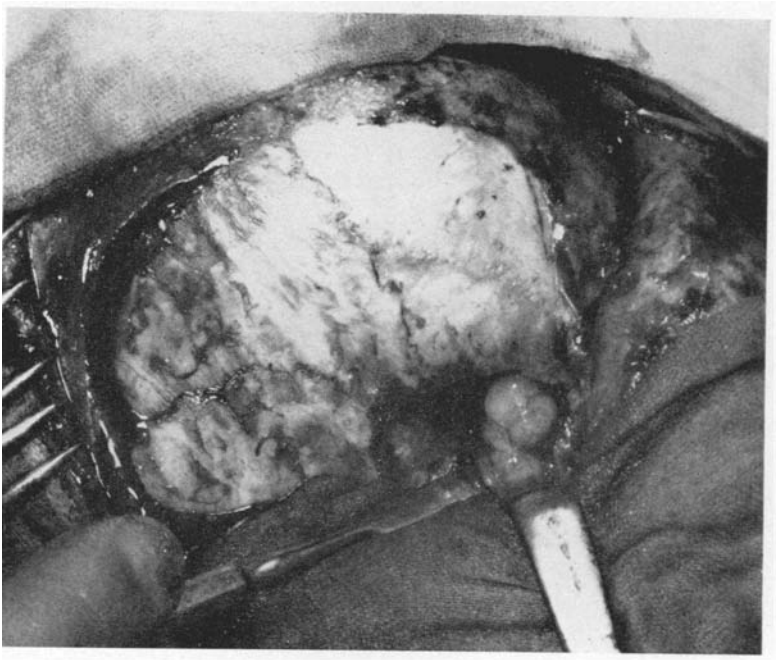


FIG. 3.
Orbital tumour in the process of being removed.

"Mrs. ——— came to see me this morning and since she has satisfactory vision in her right eye, I think it is desirable that she should have an exploration of the right orbit. In my opinion the fibroma is likely to be in the upper part and I would recommend that the exploration is carried out by a neurosurgeon through the roof of the orbit."

Mr. Radley Smith, Neurosurgeon to R.N. E.N.T. Hospital agreed to explore the orbital space.

X-ray

2.11.56
Orbit. ? any bony abnormality in right orbit.

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Report

The optic foramina are normal and the orbits are normal.
No abnormality seen in the sinuses or skull. N.A.D. in chest.

Blood

4.1.57

Hb.....	12·6 mg. (81%)	MCD	7·1%
R.B.C. per c.mm.....	4·95 m.	W.B.C. per c.mm.	4,500

Red Cells mainly normocytic and normochromic.

Differential White Cell Count

Polymorphs	52%	Lymphos.	43%
Monocytes	4%	Eosinos.	0
Basos.	1%		

No significant abnormality.
Blood Group O Rh. positive.

8.1.57

Cross-Matching of Blood

O Rh. positive.

11.1.57

Decompression of right orbit. Mr. Radley Smith.

About 4½ in. long curved incision starting about ½ in. below the hairline opposite the mid point of superciliary arch, to the root of zygoma just up to the level of tragus. Temporal flap was reflected well up to the level of zygomatic arch. A neurofibroma encountered in flap was removed. Temporal fascia, temporal muscle and the periosteum incised by an incision parallel and ½ in. below the original incision and were reflected downwards.

Burr hole made into the greater wing of the sphenoid just at the lowest part of temporal fossa. Bone around this hole was nibbled till the dura of middle fossa and anterior fossa and the orbital periosteum formed three bulges. Orbital periosteum incised. Lateral rectus, which was seen stretched over the tumour, was retracted down with other normal orbital contents. The tumour was taken out in bits in the beginning but the capsule was encountered and the tumour then came out easily in one large mass. (Fig. 3.) Periosteum and the temporal fascia were stitched back and a rubber drainage tube was put in. Wound closed by subcutaneous and skin stitches.

Over a period of several months the eye gradually settled to near normal position and now moves freely and the lid can easily be closed. In November, more tender swellings were removed from the right side of the face, neck and scalp. A few small ones were found attached to the side of the nerve trunk and could be easily removed without sacrificing the nerve (Fig. 4).

A more recent pathological report by Dr. Friedmann was as follows:

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Microscopical Findings (Fig. 5.)

Sections stained with hæmatoxylin and eosin show interlacing bundles of collagen fibres and Schwann cells, forming a dense network. There is a good deal of reticulin, as demonstrated in silver-impregnated sections. There is also a small number of nerve fibres in the tissue. The nuclei of the Schwann cells show some tendency to palisading but this is not as orderly as in typical schwannomas. The histological diagnosis lies between neurofibroma and



FIG. 4.
A tumour attached to nerve.

schwannoma or neurinoma. On balance the overall picture favours a diagnosis of neurofibroma.

According to some writers the neurofibromas are not clearly distinguishable from the schwannomas. Typical schwannomas are found more frequently as solitary growths, but according to Russell and Rubenstein (1959) schwannomas also form an important aspect of Von Recklinghausen's neurofibromatosis. The development of schwannomas in Von Recklinghausen's Disease and the overlapping of the histological appearances constitute one of the more characteristic, but puzzling features of this condition (Russell and Rubenstein, 1959).

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Discussion

Neurofibromata are seen in various parts of the body in different situations. Virchow first divided tumours into true and false neuromata. The former is rare and met with in connection with the autonomic nervous system. The false neuromata are more common and are growths of the connective tissue of sheaths of nerve. The exact origin of these false neuromata is not clear but cells in the sheath of Schwann play an important part in the formation.

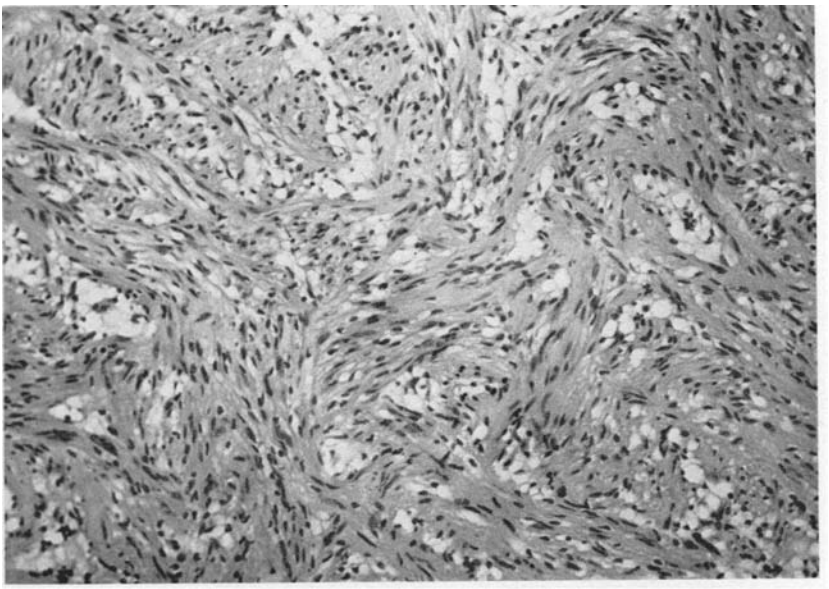


FIG. 5.

H. & E. Mag. × 170 section showing interlacing bundles of Collagen Fibres and Schwann Cells forming a dense network.

These tumours may be superficial or arise from deeper structures, the former being covered by skin whereas the latter may appear at different depths. Those situated in the subcutaneous tissue may be single or multiple, painless, moveable tumours found associated with subcutaneous and deeper, larger nerve trunks. In some cases, the whole nerve trunk becomes involved as a plexiform neuroma is formed.

The exact origin is not yet established, but Friedmann and Eisenberg (1935) point out that neuroglia is embryologically nerve tissue that is derived from ectoderm, and in the central nervous system is known as glioma. Thus, pure fibromas, just as pure neuromas and pure gliomas of nerve fibre are rare, as most of the tumours are composed of both connective tissue and of cells of Schwann.

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It is thought by some observers, that the solitary peripheral neurofibromata belong to a different disease process from neurofibromatosis. In support of this, it is stated that, in the solitary peripheral neurofibromata, myelinated fibres are found predominantly in the periphery of the tumour, whilst in neurofibromatosis, they run through the substance

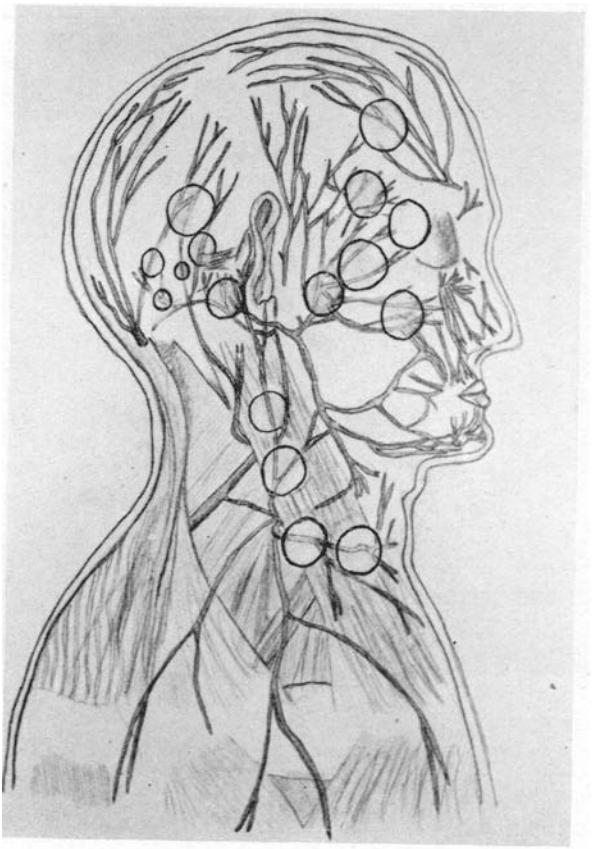


FIG. 6.
Areas on the right side of the head and neck where tumours have so far been removed.

of the tumour. Others however, consider that there is no definite histological basis for the separation of these two conditions. Greenfield (1938) with an experience of many cases, proven by autopsy to be neurofibromatosis, has, especially in the nodular form of neurofibromatosis, observed many swellings on the peripheral nerves which were similar histologically to "solitary peripheral neurofibromata". In such nodules, obviously part of neurofibromatosis, the myelinated fibres were found chiefly at

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the periphery of the tumour. In his opinion, there is no essential difference between the structure of the conditions here described as acoustic neurofibroma, solitary peripheral neurofibroma and neurofibromatosis.

An extensive search in the literature during the last decade, failed to find another case with multiple neurofibromata confined to a region of the body, such as the right side of the head and neck (Fig. 6). Further, no reasonable explanation can be given why these nerves in this region alone should be involved. There are many of these tumours on this side varying in size, but as they become larger, they become tender and painful, and in the face give rise to disfigurement.

Another appeared over the mastoid and as it enlarged it displaced the post-meatal wall forward causing a middle-ear deafness. It becomes more serious, however, when these simple tumours arise in the nerves traversing the skull, such as the internal auditory meatus or the orbit, as in these cases, its removal can only be undertaken through a major surgical approach, though once they are uncovered, removal is very simple.

Comments

There is some confusion in terminology of these tumours. The tumours are simple and benign and can easily be removed. The only difficulties arise however, when they appear in the orbital cavity or the internal acoustic meatus, when by virtue of its inaccessibility and its situation in a non-expansile cavity, it can give rise to serious consequences, unless an early diagnosis is made, and the tumour is removed. In the early stages, the small tumour could be plucked off the nerve, but as they increased in size, the nerve became involved and had to be sacrificed in order to remove the tumour.

Summary

A case of multiple regional neurofibromata confined only to the right side of the head and neck. The most serious consequence so far, was the development of one of these tumours inside the orbit which was removed by our neurosurgeon with a very good result.

I wish to thank Dr. I. Friedmann for his care in studying the pathology of this case and Mr. D. Connolly of the Photographic Unit at the Royal National Throat, Nose and Ear Hospital for the photographs and photomicrograph.

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