MALIGNANT LYMPHATIC TUMOURS OF THE ORBIT

A REVIEW OF EIGHT CASES*

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WHEN a case presented at the Manchester Royal Eye Hospital which was diagnosed as a malignant lymphatic tumour of the orbit, a search was made in the records for similar patients. Follow-up of this trail led naturally to the Christie Hospital and Holt Radium Institute, where the records contained further cases referred there direct.

Altogether eight cases reported as malignant orbital lymphatic tumours have been collected. On five of these a second pathologist's report is available from the block or slide from which the original report was made.

Case Reports

Case 1, a man aged 47, first attended the Manchester Royal Eye Hospital on June 29, 1955, under the care of Dr. S. B. Smith, complaining of proptosis and diplopia.

Examination.—There was right-sided proptosis due to a retrobulbar mass. There was no extra-orbital glandular enlargement.

A biopsy was taken from this orbital tumour, and the following pathologists' reports were received:

(i) "Lymphosarcoma. Firm yellowish-grey tissue. Specimen is composed of closely packed lymphocytic cells with considerable infiltration of the rectus muscle. In places there are groups of "blast" cells, occurring as separate entities—with large and mostly hyperchromatic nuclei, amongst the latter mitoses are plentiful."

(ii) (From the same block). "From the notes? peri-orbital tissue. A small round nodule which looks like a lymph node but contains striped muscle. This may be an infiltrated eye muscle, in which case unilateral ophthalmoplegia or early thyrotoxicosis must first be excluded I think. The cells are the shape and size of lymphocytes, and I have not found any mitoses."

Treatment.—The case was accepted as lymphosarcoma at the Christie Hospital. A tracer dose of radioactive iodine showed poor take up, and a radical course of x-ray treatment was given.

Follow-up.—The patient was last seen on September 11, 1957, when he was noted to be well with no recurrent lymph nodes.

Case 2, a man aged 62, first attended the Manchester Royal Eye Hospital on May 25, 1957, under the care of Mr. P. L. Blaxter, wishing to have his eyes tested.

Examination.—Annular swellings of both conjunctiva were seen, which were diagnosed clinically as conjunctival lymphomata. There were enlarged nodes in both axilla, both

* Received for publication June 16, 1958.

groins, and in both cervical gland fields. A biopsy was taken from the conjunctival swelling in the right lower fornix, and the following pathologists' reports were received:

(i) "A 1.25 cm. strip of yellowish tissue with a mainly smooth surface. There are two main nodules of tumour tissue comprised of densely-packed lymphocytic elements of small and medium size. The borders of the nodules are fairly sharply defined. Mitoses are only moderately numerous.

I think this lymphoma must be regarded as malignant though the grade of malignancy is low. Radiosensitivity will of course be high."

(ii) (From the same slide). "Small blobs of tissue infiltrated by small lymphocytes, and although a few dividing forms are seen I think this is more like an infiltration than a tumour. Is there any question of thyrotoxicosis here? Compare cases . . . sent here as sarcoma, . . . which may fall into the 'pseudo-tumours of the orbit' group."

Treatment.—The case was accepted at the Christie Hospital, and an initial treatment of 6 days' x-ray therapy was given.

The patient developed signs of intestinal obstruction, thought to be due to abdominal involvement in the general neoplastic process, and a further course of 7 days' deep x-ray therapy to the abdomen was given.

The patient died on August 28, 1957, and *post-mortem* examination revealed an intussusception due to a hyperplastic Peyer's patch, with widespread involvement of the abdominal glands in the neoplastic process.

Case 3, a man aged 58, was referred to the Christie Hospital on September 18, 1947, from Preston Royal Infirmary, where he was under the care of Mr. Sumner. He was complaining of a swelling arising from the subconjunctival tissues of the right inner canthus.

Examination.—There was no significant extra-orbital glandular enlargement. There were bilateral small axillary nodes but these were regarded as normal.

Biopsy was performed from the orbital mass, and the following pathologists' reports were received:

(i) "Reticulum cell sarcoma."

(ii) (From the same slide). "A small gland-like mass of tissue which appears to be well encapsulated. It consists of a mass of lymphocytes among which some dividing cells are present. Lympho-sarcoma I think."

Treatment.—Radical x-ray treatment was given.

Follow-up.—The patient was last seen on November 2, 1956, when he was reported to be well, with no lymph-node enlargement.

Case 4, a man aged 66, first attended the Manchester Royal Eye Hospital on January 25, 1952, under the care of Mr. A. Stewart Scott. He gave a history of a small lump behind the left inner canthus that had gradually grown over the last 2 years, and was now presenting as a case of proptosis.

Examination.—There was no evidence of any extra-orbital spread or glandular enlargement.

A biopsy was taken from the tumour, and the following pathologists' reports were received:

(i) "An irregular piece of grey-white tissue of a rather soft consistency. Section shows a uniform picture of a diffuse lymphoid tissue with closely packed cells. The appearances are compatible with the benign lymphomata which occur in this region, but in some of the blood vessels there is a relative excess of lymphocytes and it may be that this localized growth is part of a chronic lymphatic leukaemia."

(ii) "A piece of tissue infiltrated by lymphocytes. One small tuberculated cluster of cells seen (histiocytes). Perhaps Wassermann reaction is indicated (this was negative)."

Treatment.—The patient was accepted at the Christie Hospital and was given a radical course of x-ray therapy to the orbit.

Follow-up.—He was last seen on August 21, 1957, when there were no enlarged lymph nodes. There was slight residual proptosis, and the visual acuity in the left eye was reduced to perception of light because of cataract.

Case 5, a married woman aged 63, was referred to the Christie Hospital on May 23, 1950, after being seen by Sir Geoffrey Jefferson at Manchester Royal Infirmary.

Examination.—There was a large tumour involving the upper part of the orbit, which had spread forwards and upwards onto the forehead. A piece had been removed for biopsy. Apart from this extension there was no extra-orbital spread or glandular enlargement.

The following pathologists' reports were received:

(i) "Lymphosarcoma."

(ii) "Slide shows densely packed small cells which may be either mesenchymal or epithelial. There are also a few cells containing what I think are melanin deposits. Could it be a retinoblastoma (*i.e.* neuroblastoma)? It does not look like a leukaemic deposit."

Treatment.—The patient was accepted for radical x-ray treatment at the Christie Hospital, and after this the tumour regressed rapidly.

Follow-up.—When seen on March 7, 1951, she was reported as being well, with no enlarged lymph nodes. Unhappily however, she subsequently developed a carcinoma of the colon, which caused intestinal obstruction. After an operation for this she made a good recovery, but died on April 30, 1952. *Post-mortem* examination showed a recurrence of the carcinoma of the colon, but no evidence of lymphosarcoma.

Case 6, a married woman aged 76, first attended the Manchester Royal Eye Hospital on April 24, 1956, under the care of Mr. O. M. Duthie, complaining of diplopia.

Examination.—The left eye was proptosed and displaced laterally. There was a palpable tumour at the inner canthus, and bilateral enlarged cervical lymph nodes, one of which was removed for biopsy. The following pathologist's report was received:

"The normal architecture of the node has been lost, and instead a uniform sheet of lymphocytes is seen. The appearances are compatible with either lymphosarcoma or chronic lymphatic leukaemia."

Treatment.—The patient was accepted at the Christie Hospital as a case of "proved lymphosarcoma".

Follow-up.—There was a good response to irradiation and the diplopia improved, but subsequently her condition deteriorated and she died on November 24, 1956.

A *post-mortem* report stated that there was still disease present in the treated area (the orbit and the neck), and that new foci had arisen outside it.

Case 7, a man aged 64, first attended the Manchester Royal Eye Hospital on September 11, 1953, under the care of Mr. A. Stewart Scott, complaining of proptosis and diplopia.

Examination.—There was an orbital tumour displacing the globe downwards and forwards, and a mobile enlarged pre-auricular gland. A biopsy was taken from the orbital tumour, and the following pathologist's report was received:

"Lymphosarcoma. An irregular piece of fibro-fatty tissue 1.5 cm. long lying in the orbital tissue, and large and small areas of closely packed small lymphocytes. There is marked

infiltration of and replacement of the orbital fat by tumour cells. Mitoses are only moderately numerous.

The picture indicates a malignant type of lymphatic neoplasia, though of low grade, and the prognosis may be relatively good—at least for some time."

Treatment.—The patient was accepted at the Christie Hospital as an "apparently localized lymphosarcoma—and essential to treat radically", and was accordingly given a radical course of x-ray therapy.

Follow-up.—When he was seen on March 27, 1957, the visual acuity in the left eye was poor because of irradiation cataract, but otherwise the patient was well, with no recurrent lymph nodes.

Case 8, a married woman aged 38, was first seen in May, 1948, by Sir Geoffrey Jefferson at Manchester Royal Infirmary. She then gave a history of swelling of the right upper and lower lids, which had been present for 2 years.

Treatment.—On June 26, 1948, she underwent an operation for the removal of this tumour, which was found to originate in the orbit and to surround the globe. A large part of the tumour and the globe were removed, and the following pathologist's report was received:

"Typical lymphosarcoma."

Progress.—She was seen at the Christie Hospital on August 26, 1948, with some residual tumour still present. An indefinite swelling was present in the right upper deep cervical region.

She was given a radical course of x-ray therapy to the orbit and cervical region after which the tumour disappeared.

Follow-up.—This has been difficult, as the patient is now living in Jamaica, but the last letter from her doctor received in August, 1956, stated that she was well.

Discussion

As Scott (1958) points out, one's attitude towards lymphatic tumours may rest at any position between two extremes. The view may be taken that they are variable in appearance and behaviour, and capable of different histological structure and variable pathological activity during the course of the disease. Classification is easy in these circumstances: all tumours are lymphomata, of different degrees of malignancy.

On the other hand each histological picture may be regarded as characteristic of a disease entity, necessitating a detailed classification such as that of Robb-Smith (1938).

It is not intended to discuss this problem further, but it undoubtedly makes it difficult to compare cases when the only medium is the printed word.

Various classifications have been proposed to help the situation, such as those of Feinstein and Krause (1952), Forrest (1949), and Robb-Smith (1938), all of which differ from each other considerably. Even so, terms are frequently employed which are not in common use, such as lymphocytoma (van Wien, 1948).

Furthermore, even when common terms are employed, cases are reported which might be differently labelled by other pathologists. For example, a

case of lymphosarcoma of the orbit was reported by Friedman, Borrelli, and Geleris (1955) as having been cured by β rays. It is impossible to be certain that this tumour was identical with those in the seven cases of lymphosarcoma reported by Forrest (1949), four of which were dead in 2 years even after radical x-ray therapy.

Indeed the statement that the diagnosis may rest on clinical behaviour of the growth (Duke-Elder, 1952) is probably the most satisfactory rule to follow, and is certainly borne out by the small series reported above.

If these cases are considered in terms of extra-orbital spread alone, it is seen that at the time of presentation there was evidence of glandular involvement outside the orbit, or direct spread of the tumour outside the anatomical confines of the orbit, in four of the eight cases.

Of these four, three have died within 2 years of being first seen. One patient may be considered to have died from intercurrent disease (Case 5), but excluding this, there still remains a mortality of 50 per cent. within 6 months of presentation.

Of the patients without extra-orbital spread, all are alive and well after an interval varying from 2 to 9 years.

Summary

(1) Eight cases reported as malignant lymphatic tumours of the orbit are described.

(2) The difficulty of comparing cases on an histological basis is noted and the series is analysed in terms of extra-orbital spread.

It is a pleasure to acknowledge the generous facilities granted to me at the Christie Hospital for examination of their case records, and to thank all the consultants at the Manchester Royal Eye Hospital and elsewhere for permission to publish cases under their care. Especial thanks are due to Mr. P. L. Blaxter, without whose initial encouragement the inquiry would not have been undertaken.

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