Lymphosarcoma of larynx

By T. S. Dogra (Manchester)

Lymphosarcoma of the larynx is rare. Kratz and Ritterhoff (1961) analysed 26 cases of sarcoma of larynx reviewed by Havens and Parkhill (1941); they found a further 45 cases from world literature and added three cases of their own. Of the total of 74 cases of sarcoma, they found only three cases of lymphosarcoma of the larynx. Sacco (1962) states that only eight cases have so far been recorded. The purpose of this paper is to record a further case of lymphosarcoma of the larynx, treated by primary surgery followed by radiotherapy with a ten-year survival.

Case report

L.J., female, aged 69 years, was seen at Wythenshawe Hospital on 17 June 1960, as an urgency with an eighteen months' history of increasing hoarseness following an operation for varicose veins. Recently she had started developing stridor at rest. She also complained of bilateral otorrhoea and deafness of long standing.

On examination she was found to be a fit woman for her age. Indirect laryngoscopy revealed a large smooth swelling in the left false cord which extended down to, but did not involve, the vocal cord. The left hemilarynx was fixed. The airway was just adequate. There were no palpable cervical nodes present. The spleen and liver were not palapable nor was there evidence of any other lymph node enlargement. Apart from a few rales at the bases of the lungs, the cardio-respiratory systems were within normal limits and she was normotensive. She had bilateral chronic suppurative otitis media.

Previous history

As stated she had had an operation for varicose veins prior to being seen in June 1960. In March 1959, she was investigated at another hospital because of persistent hoarseness since she had the operation. A small swelling above the left vocal cord was found. Under general anaesthesia direct examination of the larynx and biopsy was carried out on three occasions at different intervals within twelve months, the last one being on 20 March 1960. No malignancy was detected in any of the three biopsy specimens.

Investigations

Blood examination showed haemoglobin to be 13·2 gr.; W.B.C. 4,600 with polymorphs 47 per cent, lymphocytes 50 per cent; eosinophils 1 per cent; basophils 0 per cent, and monocytes 2 per cent.

Chest X-ray showed left ventricular enlargement with unfolding of aorta. There was no active pulmonary lesion nor any evidence of metastasis. X-rays

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of cervical spine and S.T.V. of neck showed no lesion apart from osteoarthritic changes with narrowing of disc spaces between C5, 6 and 7. Tomograms were not carried out.

Treatment

She was admitted to Wythenshawe Hospital the same day and under general anaesthesia a direct laryngoscopy was carried out. The operative findings were as follows:

'A large smooth myxomatous looking swelling was found to occupy the whole of the left false cord and extended down to, but did not involve, the vocal cord. Left side of the larynx was fixed. It was extremely difficult to get an adequate bite of the tumour as whatever type of forceps were used the growth tended to slip out. The interesting feature was the most peculiar feeling on the bite forceps reminiscent of the plasma-cytoma seen a few years previously. Four adequate bites were however obtained with difficulty and sent for histology.'

The sections were examined and reported upon by Dr. K. Lodge and later seen by Professor A. C. P. Campbell.

'Four fragments of tissue, the largest measuring $3 \times 2 \times 2$ mm. The specimen consists of a portion of tissue covered by squamous epithelium. The underlying tissue is very heavily infiltrated by small round cells of lymphocytic type with a few plasma cells and reticulum cells. The pattern is regular and mitotic figures are extremely scanty; no lymphoid follicles are seen. There is an inflammatory reaction in this tissue and it is just possible that the whole pathology is inflammation but the findings favour neoplasia and I think the most probable diagnosis is a low-grade lymphosarcoma.

'Additional sections confirm the neoplastic nature of the specimen which must be regarded as a lymphosarcoma of low-grade malignancy. I have shown these sections to Professor A. C. P. Campbell, who concurs with this opinion.'

On 23 June 1960 she was referred to Professor V. F. Lambert for his opinion and advice. His remarks were: 'Left half of the larynx is swollen and fixed. I cannot recognize the true cord on this side and there is ulceration in the false cord mass. Her airway is only just adequate. I cannot hazard or even guess at the pathology of this swelling.'

Laryngofissure and a frozen section was thought logical but after discussion with the pathologists it was decided to excise the lesion. On 1 July 1960, through classical laryngofissure approach the left laryngeal ventricle, including the tumour and most of the left true cord was excised (Figs. 1 and 2), and submitted for histopathology. Her post-operative recovery was uneventful.

The histology report (Dr. K. Lodge) (Figs. 3 and 4):

'A portion of tissue measuring $3 \times 2 \times 1$ cm. The cut surface shows soft white tissue forming a nodule which appears encapsulated on one surface, but on the other extends to the line of excision. The surface epithelium of the larynx is intact (Fig. 3). The tissue beneath the epithelium (Fig. 4) shows widespread infiltration by small regular round cells with scanty cytoplasm among which there are very occasional larger cells of reticulum type. Mitotic figures are present, and while the mitotic activity would be assessed as only moderate, it is considerably more than appeared in the previous biopsy sections.

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Tumour tissue extends to the line of excision on the deep surface of the specimen. The appearances are of a lymphosarcoma of the larynx.'

In view of the histological findings, the patient was referred to Christie Hospital and Holt Radium Institute for radio-therapy. Total tumour dose of 2,750 rads (M.V.) was given over a period of 20 days. The post-irradiation oedema was slow to resolve. A large lobulated swelling was noted overlying the left hemilarynx. Soft tissue views and tomograms showed a rounded mass lying

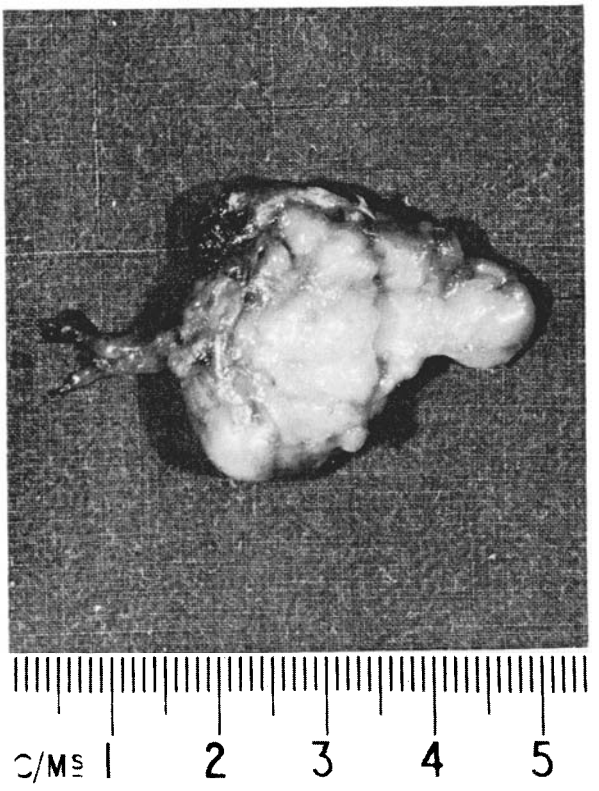


FIG. 1.
Photograph of excised tumour.

at a higher level on the fasle cord on the left with extension upwards on the lateral wall and downwards into the subglottic area. A direct laryngoscopy was carried out on 19 November 1960, under general anaesthesia. Scarring and remains of the left vocal cord were observed. There was oedema of the arytenoid and postarytenoid regions, but there was no macroscopic evidence of recurrence.

Regular follow-up has since been carried out. Blood and bone marrow studies were normal. Chest X-rays remained clear. No evidence of local recurrence or distant metastasis was noted. She was last seen in December 1970;

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she had remained well, her voice was reasonable, the left side of the larynx remained fixed as before but the right side was clean and fully mobile.

Discussion

Incidence: Sarcoma of the larynx is known to be rare. Lymphosarcoma, first described by Koschier and Stoerk, is even rarer (Ewing, 1940). Kahler (1928) saw only two cases of sarcoma of the larynx in his practice over a period of 27 years. St. Clair Thomson (1930) in a wide experience, saw a single case of sarcoma of the larynx. Figi (1933) found only four cases of sarcoma among

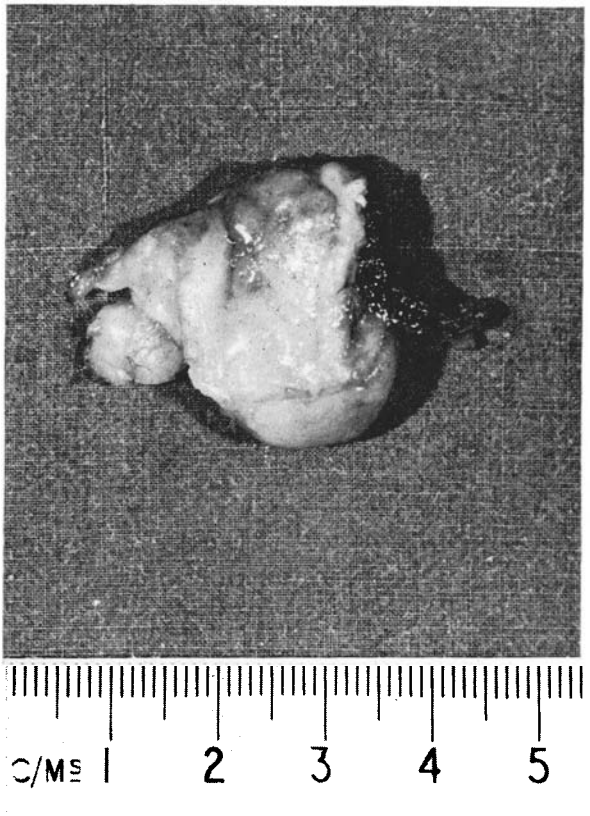


FIG. 2.
Photograph of excised tumour.

717 cases of laryngeal malignancy seen at the Mayo Clinic over a period of 23 years between 1910 and 1933. Havens and Parkhill (1941) reviewed pertinent English literature up to 1939 and found fifteen recorded cases of laryngeal sarcoma to which they added eleven cases treated at the Mayo Clinic—the latter included four cases reported by Figi. Their conclusion was that for every hundred cases of carcinoma of the larynx there was one case of sarcoma. None of these cases of sarcoma was a case of lymphosarcoma.

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Norris and Peale (1961) state that the incidence of sarcoma is probably less than 1 per cent of all malignant tumours of the larynx. There is a possibility of twelve different types of malignant mesenchymal tissue. They analysed nine cases of sarcoma observed at the Chevalier Jackson Clinic from 1941 to 1959 inclusive. There were four cases of fibrosarcoma, one each of neurogenic

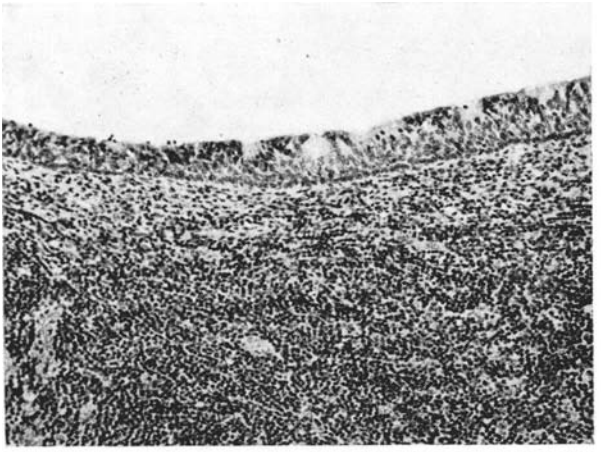


FIG. 3.
Photomicrograph × 120 magnification to show intact epithelium over tumour.

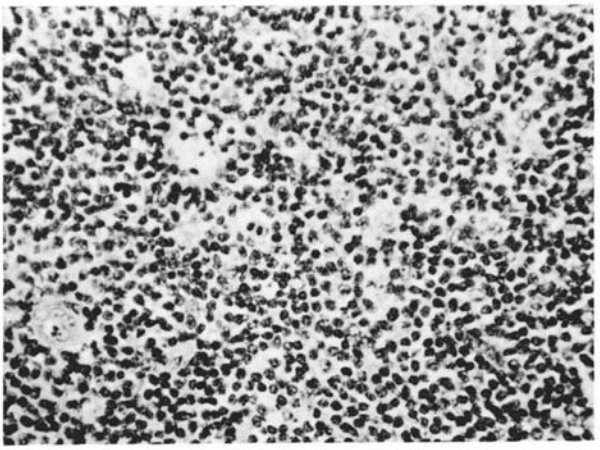


FIG. 4.
Photomicrograph × 180 magnification to show cells and a mitosis.

sarcoma, Hodgkins' sarcoma, reticulum-cell sarcoma and two of so-called carcinocarcinoma. They emphasize the difficulty in histological distinction between Hodgkins' sarcoma and lymphosarcoma.

Kratz and Ritterhoff (1961) reviewed 45 cases of laryngeal sarcoma recorded in world literature and three cases of their own. Only three cases of lympho-

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sarcoma were to be found. Sacco (1962) states that only eight cases of lymphosarcoma have so far been recorded. Reference to Index Medicus up to the end of 1970 showed no new case report.

Diagnosis may present various difficulties and this has been stressed by various authors (Clerf, 1946; Fergusson, 1943; and Lachmann, 1951). The case reported in this paper showed many of the points stressed by them. Three examinations of the swelling seen in 1959 and three biopsies at intervals over a period of twelve months failed to reveal the true nature of the lesion. The difficulty in obtaining an adequate bite of the forceps may result in failure to obtain a representative portion of the tumour with enough depth to it. The specimen may therefore only show an inflammatory reaction in its superficial part and adjoining deeper part while the true pathology may lie still deeper. Hence the necessity and desirability to obtain an adequate and fair-sized biopsy material with adequate depth to it. Zeiss Laryngoscopy may prove of great value. Histological diagnosis may present difficulty and excision biopsy may become necessary, as it did in this reported case.

Treatment: According to Figi (1933) 'Treatment of sarcoma of the larynx depends on its size, situation, and activity. Since many of these tumours are pedunculated and show less tendency to infiltrate the laryngeal structures and metastasise later than do carcinoma of the larynx, they may remain operable for a considerably longer period. Thyrotomy is especially useful in the treatment of sarcoma of the larynx . . .'

Lymphosarcoma is an exception to local removal of or laryngectomy. As a rule it is pedunculated and may be lobulated and show no ulceration. Its response to irradiation is excellent and this is the recommended choice of treatment (Clerf, 1946; and Lachmann, 1951). Experience, however, is limited because of the rarity of the condition. The three cases in Kratz and Ritterhoff's report were those documented (Mackenty, 1934; Clerf, 1946; and Lachmann, 1951).

Mackenty treated his case with laryngectomy with a twelve years' survival; Clerf treated his with irradiation with a ten years' cure. Lachmann's treatment was with hemilaryngectomy; the patient unfortunately succumbed to pulmonary embolism on the ninth postoperative day.

The case reported in this paper is alive and well ten years following primary surgery followed by radio-therapy.

Summary

A case of lymphosarcoma with a ten years' survival following combined surgery and radio-therapy is recorded. The desirability of and the difficulty in obtaining adequate biopsy material is noted. The difficulty in histological diagnosis is mentioned.

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Clinical records

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