A CASE OF LEIOMYOSARCOMA OF THE VULVA WITH SECONDARIES IN THE LUNGS AND BONES

by

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Sarcoma of the vulva is among the uncommon diseases of the genital tract. Very few cases have been reported in the literature but the few reported would seem to be of varying origin. At the King George Hospital, Visakhapatnam, there were 16,874 gynaecological inpatients during the last 10 years among whom there were 50 cases of carcinoma of the vulva and only one case of sarcoma, the present one. This case is reported for its rarity and some interesting features.

Case Notes

Mrs. G. A., aged 55 years (I. P. No. 1044), was first admitted in the surgical wards of this hospital on 21-1-1964 as a case of osteogenic sarcoma of the upper end of the left humerus. She came for a painful swelling in the left shoulder. As she also had a tumour mass in the vulva, she was referred to the gynaecological department.

Past History: Three years ago, she had a tumour on the right side of the vulva which was removed by her own doctor. No histo-pathological report was available on the growth. After a year, the tumour recurred and grew to the size of an orange in two years' time. Three months ago, she developed a very tender, fusiform swelling of the left shoulder region. She also had a swelling on the chest wall lateral to the right breast. Except for the shoulder swelling, the others were not painful. She came to the hospital only for the shoulder swelling (Fig. 1).

She attained menarche at the 15th year. She had been married for 35 years and had eight uneventful pregnancies and labours. Her last child was aged 20 years. She attained menopause five years ago. On examination, she was illnourished and in great pain. She was not anaemic, nor dyspnoeic. The cardiovascular, respiratory and gastro-intestinal systems were clinically normal.

There was a tender, fusiform swelling over the left shoulder 8″ x 6″ x 6″ in size involving the upper end of the humerus and restricting the movements at the shoulder joint. It was warm to touch, with large distended veins, and very vascular and pulsating. There was a swelling 2″ x 1″ x 1″ in size on the 8th rib just lateral to the right breast.

There was a lobulated, firm, swelling 4″ x 3″ x 3″ in size in the right groin just below the inguinal ligament. It extended below on to the right side of the vulva up to the posterior end of the labium majus. The swelling was not adherent to the skin, nor deeper structures. The skin was intact and there was a linear scar 2″ long on the vulval portion of the tumour. This freely mobile swelling which was not tender nor ulcerated, simulated a mass of lymph nodes, except for the large size and extension onto the vulva. Figures 1 and 2). Pelvic examination revealed normal external genita-

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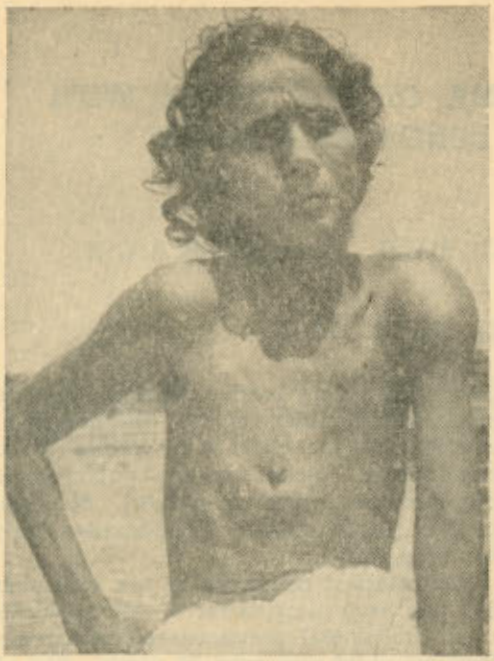

Fig. 1
Clinical photograph showing the swelling over the left shoulder and on the right 8th rib.

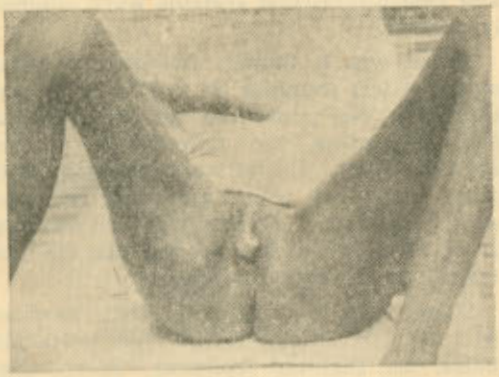

Fig. 2
Clinical photograph showing the vulval portion of the growth.

lia, an atrophic uterus and a healthy cervix. The adnexae were normal.

On 31-11964, a biopsy was taken from the mass. The report of the pathologist was (No. 494/64) "structure of a highly cellular fibromyoma." (Figure 3).

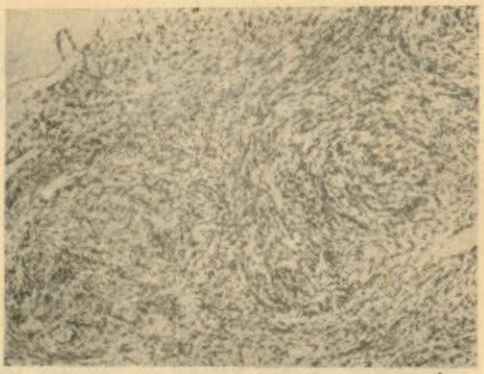

Fig. 3
Photomicrograph. No. 737/64 x 100 showing structure of the leiomyoma.

On 15-2-1964, under local anaesthesia, the whole mass was easily enucleated (Figure 4). The pathological report was as

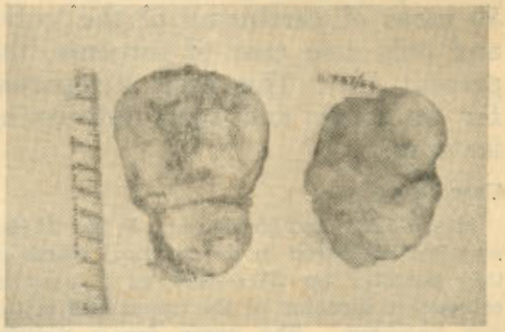

Fig. 4
Clinical photograph showing a cross-section of the two portions of the tumour in the groin and vulva.

follows: (No. 734/64) "There is spindle-cell pattern of the cells, arranged in facicles. There is marked anaplasia with mitotic figures. The nuclei are hyperchromatic. There are numerous gaint cells, areas of massive haemorrhage and necrosis consistent with a diagnosis of Leiomyosarcoma (Figure 5)." Biopsies were taken from the growth on the chest wall and the left shoulder. Both showed a picture of leiomyosarcoma (Figure 6 and 7).

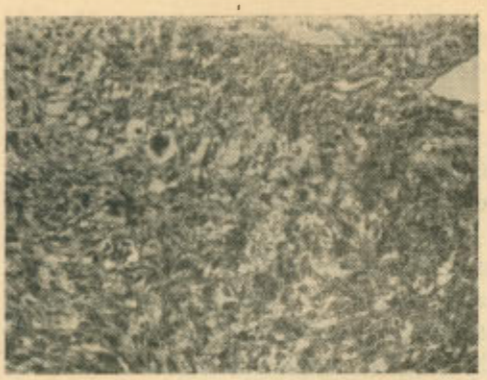

Fig. 5
Photomicrograph of vulval growth. X 100 showing structure of leiomyosarcoma.

Fig. 6
Photomicrograph of growth on rib showing structure of leimyosarcoma.

Fig. 7
Photomicrograph of growth on left shoulder X 200 showing structure of leiomyosarcoma.

Other investigations

Skiagram of chest: (Figure 8). **A. P. View:** secondaries in two areas in the lung. Secondary in the 8th rib right anterior end.

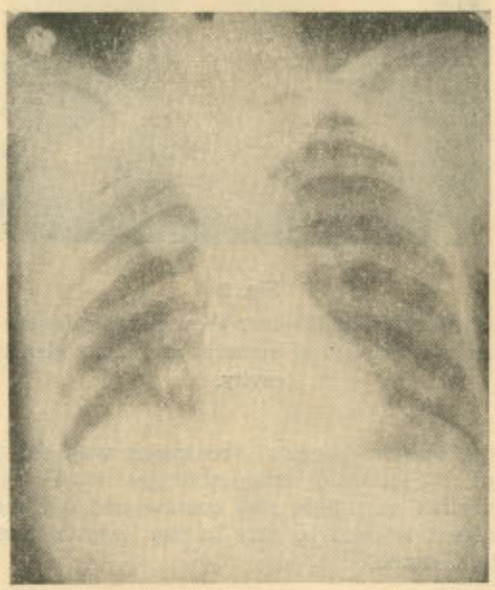

Fig. 8
Skiagram of chest—A. P. view—showing 2 secondary nodules on the right side in the lungs and an eroded area on 8th rib right anterior end.

Skiagram of spine: - no secondaries.

Skiagram of left shoulder: (Figure 9). Upper and left humerus and shoulder show soft tissue swelling with destruction of the bone in the upper end of the humerus. Glenoid cavity not involved. A portion of the head of the humerus is intact.

Primary osteogenic sarcoma. Secondary malignant growth.

Haemoglobin: 80% (Sahlis).

Urine: No albumin. No sugar. No pus cells.

W.B.C. (differential count) polymorphs 58%, lymphocytes 22% eosinophils 26%.

Blood urea: 23 mg%. **Blood group:** "A".

Urine—nil abnormal—**Stool;** no ova, no cysts.

Diagnosis. Primary leiomyosarcoma of the vulva with secondaries in the lungs, rib and left shoulder region.

Treatment. As the lesions were exten-

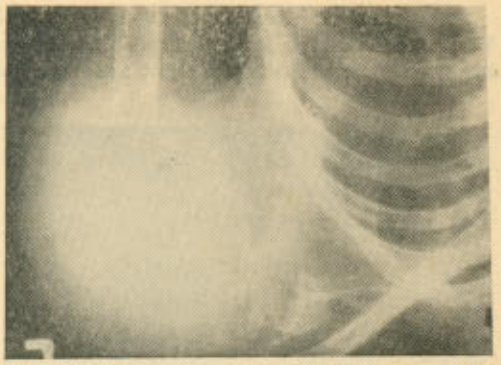

Fig. 9

Skiagram of left shoulder area showing destruction of upper end of humerus and intact glenoid cavity.

sive further surgical treatment was ruled out. Palliative chemotherapy with triethylins melamine was commenced but the patient refused to stay in the hospital and went home.

Comment

It would appear that the local recurrence was due to inadequate removal. The growth was rather slow at first and then produced distant blood-borne metastases. The large size of the secondary gave some difficulty in diagnosis of the primary site. Histological examination alone decided the site of the primary as the vulva.

Sarcoma of the vulva is a rare lesion. It may be of the pigmented or nonpigmented type. Blairbell (1907) made the earliest collection of 18 cases. Kehrer (1909) collected 77 cases up to his time. Hunt (1954) in her monograph says that sarcoma of the vulva is a rare disease. It arises from the skin and three types are differentiated. Two of these (a) non-pigmented sarcoma and (b) melanotic sacoma are reported in the vulva. The exact aetiology is not known and all ages from infancy to old age are affected, some even in pregnancy (Nolan 1957). A variety of histological types such as liposarcoma (Taussig, 1937), myxosarcoma, fibrosarcoma, fibromyxosarcoma, rhabdomyoblastic sarcoma, and reticular-celled sarcoma have been reported. Our case appears to be a leiomyosarcoma. This may arise from muscle cells around blood vessels or from the terminal part of the round ligament. Other rare benign tumours like granular myoblastoma or haemorrhagic pericytoma may also become malignant (DeSouza and Lash, 1959). The non-pigmented sarcoma arises as a small soft growth, which may be mistaken for a lipoma or fibroma or sebaceous cyst. Ulceration occurs later. If local removal is adequate in some cases the prognosis is said to be good. Melanotic sarcoma on the other hand is highly malignant and fatal. In this case the tumour appears to have remained localised for the first few years and later spread by the blood stream, making the prognosis very poor.

In reporting this case, we thank the Medical Superintendent for permission to use the case notes and Dr. D. J. Reddy, M.D. Director of the Department of Pathology for the histological reports.

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