EPITHELOID SARCOMA OF THE VULVA

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ABSTRACT

A rare case of epitheloid sarcoma of the vulva is described. This is a soft-tissue malignancy arising from tenosynovial tissue. The patient presented with a painless lump of the vulva of a month's duration. An excision biopsy was performed followed by a wide local excision after the actual diagnosis was confirmed.

Post-operatively, her recovery was uneventful and she was seen in the Cancer clinic at regular intervals. Three years following surgery, she was well with no evidence of any recurrence. The suggested mode of treatment ranged from a wide local excision to radical vulvectomy with groin node dissection.

Key Words: epitheloid sarcoma

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CASE REPORT

A 21-year old woman first noticed a painless lump in the left vulval region a month before seeking medical attention on 30th October 1983. There was no tenderness, pruritus, contact bleeding or increase in size of the nodule. She did not experience any irregular vaginal bleeding, discharge or change in bowel or urinary habits. There was no history of any anorexia, weight loss, fever, cough or haemoptysis.

On examination, the patient appeared well. There was no pallor. Examination of the heart and lungs revealed no abnormality. Her breasts were normal. There were no palpable supraclavicular lymph nodes. Examination of the abdomen also revealed no abnormality. The liver, kidneys and spleen were not palpable. On inspection of the vulva there was a nodule in the left labium majus. The nodule was one centimetre in diameter, round, mobile, firm and non-tender. The edges were well-defined. Notably absent were any punctum, abnormal vessels, surface fissuring, haemorrhage or surrounding inflammation. Clinically, the nodule appeared to be located in the subcutaneous tissue with no tethering to the over-lying skin or underlying tissues. The clitoris and rest of the the vulva were normal. The inguinal lymph nodes on either side were not palpable. A gentle one-finger vaginal examination revealed the uterus to be normal-sized, axial and mobile. There were no adnexal masses felt. Rectal examination revealed no abnormality. An excision biopsy of the nodule was performed on 18.11.83 under local anaesthesia. A skin crease incision was made and a total excision of the nodule carried out with a wide, clear skin margin. She was well post-operatively.

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Microscopic examination revealed fibroadipose tissue with well-defined nodular areas composed of polygonal cells with eosinophilic cytoplasm, mild to moderate nuclear pleomorphism and occasional mitosis. There was no cellular keratinization present. The surrounding fibrous tissue showed a few focal areas of spindle-shaped tumour cells. The diagnosis was epitheloid sarcoma of the vulva (Figs 1 and 2). This was confirmed by a world renowned pathologist from the Indianna University Hospital, on recommendation by our local pathologist.

Fig 1: Epitheloid Sarcoma showing irregular nodular masses of cells with central necrosis or fibrosis (H + E X 100)

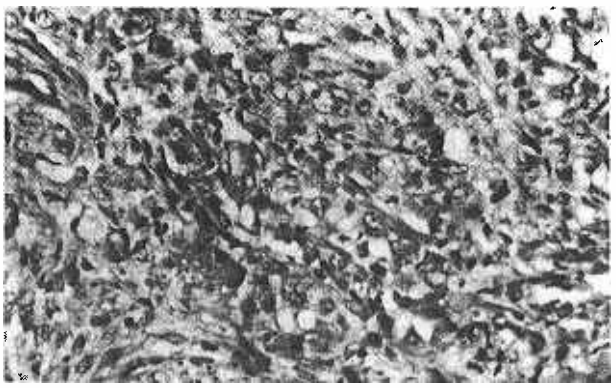

Fig 2: Eosinophilic polygonal and spindled tumour cells at the periphery of the modules (H = E X 400)

Following a wide excision of the vulva, the patient was seen at the Gynaecological Cancer Clinic at two-weekly intervals for two months and monthly intervals thereafter. Three years after surgery, she appeared well with no evidence of any recurrence.

DISCUSSION

Epitheloid sarcoma, a rare neoplasm, was first recognised in 1970 as a distinct tumour of the subcutaneous tissues. The actual histogenesis is still an area of debate but the consensus of opinion is that the tumour arises from synovioblastic mesenchyme (1) Gabbiana presented evidence in support of a synovial origin (2).

Primary soft tissue sarcomas of the vulva are rare neoplasms comprising approximately one per cent of all malignant neoplasms of the area (3). Most vulva sarcomas occur in patients between 30 and 50 years of age (mean 38 years) (4).

The tumour occurs in the subcutis and deeper tissues of the vulva. When located in the subcutis, it usually presents as a firm nodule that may be solitary or multiple, has a callus-like consistency, and is often described as a 'firm lump' that is slow-growing and painless (5). Nodules situated in the dermis appear elevated above the skin surface and frequently become ulcerated. Deep seated lesions are usually firmly attached to fascial structures, are less well-defined and often manifest as areas of induration. Pain or tenderness is never a prominent symptom with the exception of occasional tumours that encroach on large nerves. Gross inspection usually reveals the presence of one or more nodules measuring 0.5 to 5 centimetres in greatest diameter. The cut surface has a glistening, grey-white appearance with focal yellow or brown areas caused by focal necrosis and haemorrhage.

The principal microscopic features are the distinct nodular arrangement of the tumour cells, their tendency to undergo central degeneration and necrosis, and their epitheloid appearance and eosinophilia. Necrosis of tumour nodules is a common finding. The constituent cellular elements range from large ovoid or polygonal cells with deeply eosinophilic cytoplasm to plump spindle cells. As a rule, cellular pleomorphism is minimal and multi-nucleated giant cells are absent or scarce. Calcification and bone formation occasionally occur.

The frequency with which the tumour is mistaken for a benign process is chiefly the result of its deceptively harmless appearance during the initial stage of the disease. Superficial tumours of small size with a nodular or multi-nodular pattern are likely to be mistaken for an inflammatory process particularly an infectious granuloma. This was the case in our patient and it is regretted that no photographic record of the original tumour was taken. An epitheloid sarcoma arising in the labium majus might be diagnosed as a Bartholin cyst. Other benign conditions likely to be confused with epitheloid sarcomas are fibromas, lipomas, dermoid cysts and viral warts. The tumour has a potential to recur and metastasize and this occurs most commonly to the regional lymph nodes and the lung. Multiple recurrences are a characteristic feature. Recurrences gradually develop within the first year after diagnosis but late recurrences (in one case 25 years later) may occur. Intravascular growth and lymph node involvement are ominous features (5). A summary of the clinical course of the 5 reported cases is presented below:

CASE 1

The patient was a 55-year old white lady. The primary lesion was in the right labium majus. A local excision was done, followed by a recurrence in the inferior vulva at 8 months. This was treated by a radical vulvectomy with removal of bilateral groin lymph nodes. Further recurrences appeared two months later in the pouch of Douglas and the recto-vaginal septum followed by subcutaneous nodules in the lower abdominal wall and upper thigh.

The patient expired 15 months after the initial diagnosis from widespread lung metastases (6).

CASE 2

The patient was a 30-year old white lady. The primary tumour occurred in the right labium majus and this was treated with a wide local excision. Recurrences appeared in the clitoris at 6 weeks and the pubis ramus at 4 months. The patient died eight months later with lung metastases (7).

CASE 3

The patient was a 27-year old white lady. The primary lesion arose inferior to the clitoris. This was treated with wide local excision. No recurrences developed and the patient was alive and well nine years later (8).

CASE 4

The patient was a 31-year old white lady. The primary lesion occured in the left anterolateral edge of the vulva. Wide local excision was carried out but recurrences developed in the left labium majus and left groin at 6 years, left vulva at 9 years and right vulva and left thigh at 10 years. The patient expired 11 years later with lung and liver metastases (8).

CASE 5

The patient was a 31-year old white lady. The primary lesion occured in the left labium majus. Radical vulvectomy with removal bilateral groin lymph nodes was performed. Subcutaneous vulval nodules appeared at 17 months and the patient died of her disease at 21 months.

CASE 6

This is our patient, a 21 year old Chinese girl. The primary lesion occurred in the left labium majus. A wide local excision was done and the margins of resection were free of tumour. The patient is alive and well a year after the diagnosis.

The majority of patients with epitheloid sarcomas have sought medical attention because of a slowly growing painless mass. Pain is more prone to occur with the more deeply situated lesions (9).

The superficial lesions have a tendency to cause ulceration of the overlying epidermis.

The best line of treatment for epitheloid sarcoma is still a subject of controversy. In their review, Seemayer et al concluded that the treatment of choice for epitheloid sarcoma would be the least radical operative procedure necessary to remove the tumour completely (10).

Gallup (11) recommends a radical vulvectomy and bilateral groin lymphadenectomy, particularly if the inguinal nodes are clinically suspicious. On the other hand, Piver's case(8) was treated with wide local excision as was the treatment of our patient. Both authors stress the importance of early recognition of epitheloid sarcoma of the vulva with adequate treatment.

Radiation therapy has not been consistently helpful

and the use of chemotherapy needs to be evaluated more thoroughly.

Our patient has been clinically free of the disease for three years. Although most recurrences occur within six months, the prognosis is still guarded owing to the possibility of late recurrences.

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