A RARE TUMOUR AT RARE SITE - DERMATO FIBROSARCOMA PROTUBERANS OF CLITORIS

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Abstract: Vulval malignancies are relatively rare and accounts for around 5% of female genital malignancies. There has been a marginal increase in the incidence and mortality rate from vulval malignancy. The incidence has appreciably increased in the younger age group. The prognosis and conservation of vulval structures are best if this condition is diagnosed early, with a survival rate of over 90% in the absence of nodal involvement. Nearly 90% of vulval cancers are Squamous cell carcinomas and 2% are sarcomas. Basal cell carcinoma, Melanoma, Adenocarcinoma, Lymphoma, Merkel's cell carcinoma, Dermatofibrosarcoma protuberans, and Malignant schwannoma are some of the rare tumours of vulva. A teenage girl presented with enlargement of clitoris and was clinically diagnosed to have a fibromatous growth. Wide excision was done. After histopathological examination it was diagnosed as Dermatofibrosarcoma protuberans which is a very rare type of vulval malignancy. This case has been presented here for its rarity.

Keyword: Dermatofibrosarcoma protuberans, Immunohistochemistry

CASE REPORT
A 16 years old girl was admitted with history of swelling at external genitalia for 1 month, gradually increasing in size and not painful. She attained menarche at 14 years. No significant menstrual/family/past history. Examination of genitalia revealed a firm to hard irregular swelling of 6x4 cm at the site of clitoris, extending between the labia majora and minora. There was induration around the swelling without tenderness. Swelling was not adherent to deeper structures. No palpable lymph nodes. We proceeded with FNAC and the report was inconclusive with occasional spindle cells and the pathologist suggested excision biopsy.

Pic-1. Vulval growth

Despite not understanding the exact pathology, we decided to do a wide excision since we suspected a neoplastic growth. Under spinal anaesthesia, with patient in lithotomy position, an elliptical incision was made around the swelling and the fibrous mass was dissected out from its bed with wide margin. Dead space was obliterated in layers. Except for the slightly excessive blood loss due to high vascularity of the tumour, the intraoperative period was uneventful. She had an uneventful postoperative course.

Pic-2. Excised growth

The Histopathology report was “Bundles of uniform spindle shaped cells arranged in storiform pattern and cells having hyper chromatic nuclei, suggestive of Dermatofibrosarcoma Protuberans

Pic-3. Bunch of cells
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Pic-3. Histopathology

As per the oncologist opinion, patient was followed with radiotherapy. There was no recurrence in the past one year.

**DISCUSSION**

Vulval malignancies accounts for around 5% of all female genital malignancies. Hoffman introduced the term Dermatofibrosarcoma protuberans in 1925. Dermatofibrosarcoma protuberans (DFSP) is a low to intermediate grade soft tissue sarcoma arising from the dermal layer of the skin. It tends to grow in a more infiltrative manner. This tumor can assume irregular shapes and extend in a villous or finger-like manner.

**Incidence:**

DFSP accounts for less than 2% of all soft-tissue sarcomas (less than 0.1% of all cancers). It presents in all ages, including children, but the median age of patients with DFSP is 30–45 years. The incidence among blacks is almost double that among whites.

**Clinical Features:**

DFSP is characterized by specific chromosomal abnormalities involving the platelet derived growth factor B locus (PDGFB). It has a slow indolent course, with early tumours appearing as painless areas of cutaneous thickening. They may have pink, dark red or bluish discolouration, particularly at its periphery. At early stages patients usually report to the dermatology clinics with complaints of thickening of skin with or without discolouration. Over time, they develop into a larger nodular mass, and ultimately can develop into a large fungating lesion. Unlike tumors of the subcutaneous tissue, DSPF is adherent with its overlying skin. The most common site is trunk (47%), followed by lower extremity (20%), upper extremity (18%), and finally head and neck (14%). Though it is a malignant tumor, the incidence, the incidence of metastasis is 1-4% only. But typically it has an infiltrative growth pattern and local recurrence.

Core needle or open incision biopsy is required for diagnosis. DFSP has a characteristic histological appearance of monomorphic bland, spindle cells arranged in a storiform or "herringbone" pattern. Early tumors may demonstrate a "Grenz zone," which is a tumor-free region separating the tumor from the epidermis. Around 15% of cases exhibit features of high-grade sarcoma which is known as DFSP-FS. Immunohistochemical analysis helps to diagnose DFSP-FS. Positivity for CD34 is lost within the areas of sarcomatous change in cases of DFSP-FS.

**Management**

Recent guidelines recommend surgical excision with margins of 2 to 4 cms. Moh’s surgery yields excellent outcomes. The application of imatinib to DFSP has been tried with good results.

**Conclusion**

Though DFSP is an extremely rare type of vulval malignancy, it is better to think of rare types especially in younger individuals. The outcome is good with early recognition and proper surgical management.

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**REFERENCES**