A Rare Case of Adenoid Cystic Carcinoma Presenting as a Vulval Mass

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ABSTRACT
A 51 years old lady presented to the gynaecology OPD with complaints of a slowly increasing mass in the vulva for the last 6 months.
A simple-partial vulvectomy was performed and on histopathologic examination revealed predominant cribriform pattern with prominent perineurial invasion. On the basis of these findings, a diagnosis of adenoid cystic carcinoma of the vulva was given.
Key words: Adenoid cystic carcinoma, vulva.

INTRODUCTION
A 51 year old lady presented to the gynecological - OPD with complaints of a slowly increasing mass in the vulva for the last 6 months. There were no other complaints and the inguinal lymph nodes were not palpable. There was history of menopause four years back.
A simple partial vulvectomy was performed and the specimen was sent to the pathology department.
Gross examination revealed a 4x3cm nodule on the sub-cutaneous region with a slight infiltrative margin. Cut surface showed a homogenous grayish-white tissue with foci of haemorrhage.
Histopathology revealed tumour cells infiltration in the dermis. The tumour cells were arranged predominantly in cribriform pattern and in solid sheets, small cell nests and tubules. These are composed of small basaloid cells having scant amount of eosinophilic cytoplasm and round to ovoid basophilic nuclei exhibiting mild nuclear atypia. These cells are separated by thick strands of fibrous tissue. In between the tumour cells are present small lumina and cyst-like spaces containing homogenous basophilic material. Marked perineurial invasion was observed at places. Nuclear atypia and atypical mitotic figures were not seen in the serial sections examined.
The overlying epidermis and the resection surgical margins were free from tumour cells infiltration.
A diagnosis of adenoid cystic carcinoma of the vulva was made.

DISCUSSION
Adenoid cystic carcinoma is the rarest type of eccrine sweat gland carcinoma first described in 1975. ¹
Adenoid cystic cancer is a rare type of cancer that can exist in many different body sites. It most often occurs in the areas of the head and neck, in particular the salivary glands but has also been reported in the breast, lacrimal gland of the eye, lung, brain, Bartholin gland, trachea and the paranasal sinuses. It is sometimes referred to as adenocyst, malignant cylindroma, adenocystic, adenoidcystic carcinoma. ²
This is generally a slow-growing and a well-differentiated tumour and the patients may survive for years with metastases. Metastasis to the lungs has been reported but is very rare. Frequent nodal metastasis are also observed and the lymph nodes are palpable in a majority of cases.
In a 1999 study of a cohort of 160 adenoid cystic carcinoma patients, disease specific survival was 89% at 5 years but only 40% at 15 years, reflecting deaths from late-occurring metastatic disease. ¹
Adenoid cystic carcinoma of the vulva is a very rare neoplasm occurring mostly in the Bartholin’s gland of the vulva and presenting with the typical cribriform pattern. True and Pseudo-cysts are observed in within
the tumour islands. The adenoid and cystic spaces contain pale-staining mucin that in some cases is hyaluronidase sensitive. The diagnostic important feature observed on histopathologic examination is the prominence of perineurial invasion.  

Beppu Michiko reported a case of adenoid cystic carcinoma of the Bartholin’s gland in a 68-year old women presenting with history of a slow-growing mass in the right vulva since 4 years.  

Fauzer S. Abrao et al. reported two patients with adenoid cystic carcinoma of the Bartholin’s gland. Both patients had large vulvar masses with a short clinical history, and several local tumor recurrences within the first 2\textsuperscript{1/2} years after radical vulvectomy. The characteristic cribriform pattern and perineural involvement in addition to vascular invasion were present in the pathological material. No metastases were found in the inguino-femoral lymph nodes removed. Both patients are alive, without evidence of local recurrence but with lung metastases.  

The tumour is immunoreactive for carcinoembryonic antigen, amylase and S-100 protein.  

Primary treatment for this cancer, regardless of body site, is surgical removal with clean margins. This surgery can prove challenging in the head and neck region due to this tumour’s tendency to spread along nerve tracts. Adjuvant or palliative radiotherapy is commonly given following surgery. In some cases surgery is not an reasonable option, however radiation used alone may provide an effective treatment. Chemotherapy is used for metastatic disease. Chemotherapy is considered on a case by case basis, as there is limited trial data on the positive effects of chemotherapy. Clinical studies are ongoing, however.  

CONCLUSION

Although, adenoid cystic carcinoma is common in other sites of the body, the most common being the salivary glands; Adenoid cystic carcinoma can also occur in the vulva arising from the eccrine sweat glands.

This is a slow growing and well-differentiated tumour with rare metastasis.

Surgical removal with clean resected margins is the primary treatment of choice.

REFERENCES