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CASE REPORT

CASE REPORT- PLEXIFORM SCHWANNOMA OF VULVA WITH BRIEF REVIEW OF LITERATURE

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ABSTRACT

Schwannomas are benign, slow-growing tumors of peripheral nerve sheath and are rarely found in female genitalia. The most common locations for this tumor are head and neck, extremities, posterior mediastinum and retroperitonium Herein, we report a case of vulvar schwannoma in a 48 year old female, who presented with a large vulvar mass. Histopathologycal examination revealed a spindle cell neoplasm with arrangement of cells in short fascicles, whorls and a characteristic wavy nuclei in a nodular pattern. Few neoplastic spindle cells displayed pleomorphic nuclei with hyperchromasia with S-100 positivity. Inspite of its low incidence, plexiform schwannoma should be considered in the differential diagnosis of a cutaneous or subcutaneous mass in the vulva.

INTRODUCTION

Schwannoma is usually a benign tumor of peripheral nerve sheath. The most common locations for this tumor are head and neck, extremities, posterior mediastinum and retroperitonium [1]. Schwannoma arising from external female genitalia is rare. With our best knowledge, only few cases of vulvar schwannoma have been reported in the literature. Herein, we describe a case of vulvar plexiform schwannoma in a 48 year old female with review of literature.

Case presentation

A 48 year old female who was gravid 4 and para 2 presented with a mass in the right labia majora. The mass was increasing in size for last 1 year. It was associated with discomfort in performing daily activities because of its size for the last 4 months. Patient was postmenopausal with no co-morbidities. On general examination, vital signs (body temperature, respiratory rate, heart rate and blood pressure) were within normal limits. Local examination revealed a subcutaneous mass measuring 18x16x15cm arising from right labia majora. The skin over the mass was excoriated with ulceration. The mass was immobile and non-tender. Rest of the examination of external genitalia was normal. There was no history of neurofibromatosis. The routine investigations, complete blood count (CBC), biochemistry and tumor markers were within normal limits. Imaging study of abdomen and pelvis by MRI was done for further evaluation, which revealed a large mass arising from the right labia and measuring 20x16x13.5cm. There was presence of tortuous vessels giving appearance of bag of worms. Contrast enhancement was seen within the tumor and tortuous structures. Focal areas of hyperintensity was noted in the central region suspicious of blood which blooms on T2FFE images. There was presence of right inguinal lymphadenopathy with tortuous vessels, enhancing on contrast study. Radical vulvectomy was done with removal of the mass measuring arising from right labia majora. On gross examination, the surface of the skin showed presence of an ulcer covered with scab. Cut surface had well formed yellow to brown nodules ranging from 1 to 3cm in size, against a white area in the background. There was a central necrotic area measuring 5x4cm with foci of haemorrhage (Figure 1). Microscopic examination showed a spindle cell neoplasm with arrangement of cells in short fascicles, whorls and characteristic wavy nuclei with nodular pattern. The tumor cells were concentrated around blood vessels and was infiltrating the overlying skin. Few neoplastic spindle cells displayed pleomorphic nuclei with hyperchromasia. Multinucleated giant cells were also present with foci of necrosis and haemorrhage. Mitotic rate was 3-4/10 high power field. On immunohistochemistry(IHC), spindle cells were strongly positive for S-100 with Ki67 of 5%. Cytokeratin (CK) was focally positive. The neoplastic cells were negative for smooth muscle antigen (SMA), desmin and epithelial membrane antigen (EMA). The endothelial cells of intervening blood vessels were positive for CD34.

Figure 1 Gross morphology showing the nodules on cut surface

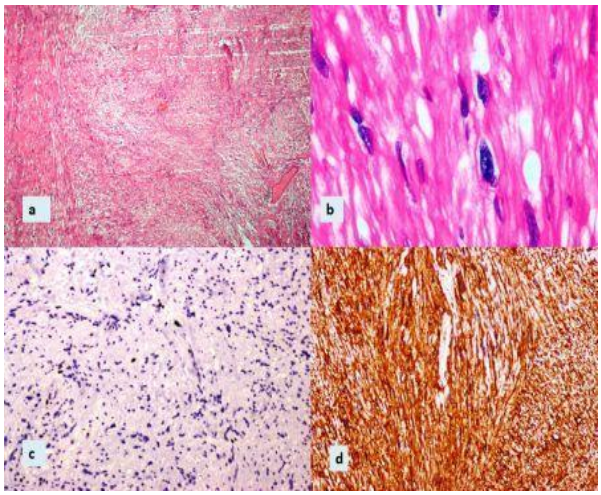

Fig. 2 microscopic findings; a) Plexiform schwannoma with typical multinodular architecture involving the dermis and subcutis (H&E, 40x). b) showing bizarre cells (400x). c) low Ki67(40x). d) Multiple tumour nodules consisting of S100 protein immunoreactive spindle cell(40x).

DISCUSSION

Schwannoma arises from the myelin sheath of peripheral nerves. It is considered to be benign tumor and is usually present as a single mass. These are encapsulated and slow growing tumors [1]. Head and neck, extremities, trunk, posterior mediastinum and retroperitonium represent the usual sites of occurance for this tumor [2,3]. The schwannoma arising from female genital tract is rare, and vulva in particular is even rarer with only few case reports in literature. The median age of presentation varies widely from 5 to 84 years although it most commonly occurs between 20 to 40 years of age [4,5]. The clinical features depend on which nerve is involved, but usually these tumors present with painless, immobile and non tender mass. Most of schwannoma of female external genitalia are benign with malignant variant representing less than 1% [6,7]. Bartholin's cyst, labial cyst, and mesenchymal tumors such as lipoma, liposarcoma, fibrosarcoma, and angiosarcoma are the diferential diagnosis and should be excluded from schwannoma [8]. Vulvar schwannoma can be misdiagnosed as neurosarcoma, neurofibrosarcoma or malignant schwannoma, hence a proper biopsy with histopathological confirmation is important for deciding upon appropriate treatment [9]. The presence of alternating Antoni type A and Antoni type B areas is considered hallmark of schwannoma. A tightly packed sheath of spindle cells arranged in pallisade and swirling patterns comprises of Antoni type A tissue whereas Antoni type B tissue is composed of loosely packed spindle cells and cells with small round nuclei. There is presence of cystic degeneration, with Verocay bodies occurring as the tumor grows. On microscopic examination, the tumor is encapsulated with presence of two types of Antoni areas, Antoni type A and Antoni type B. A strong IHC staining for S-100 protein distinguishes schwannoma from neurofibromas [10]. The most common subtype of vulvar schwannoma is the conventional type although there are reports of plexiform and ancient schwannoma in literature [2,8,11]. IHC findings are the most accurate method to establish diagnosis of vulvar schwannoma. There is a strong positive staining of S-100 in the benign form of schwannoma. The malignant counterpart usually are composed of dedifferentiated cells and do not synthesize S-100 protien [12].

Complete excision of the mass remains the treatment of choice for benign schwannoma. Prognosis is excellent although few reports of relapse after excision have been documented. These may represent result of incomplete excision in a large tumor [13]. In our case the clinical and radiological findings were suggestive of a varicocele but on proper histopathological examination with IHC the diagnosis of plexiform schwannoma was confirmed. After reviewing the literature extensively, we could find only five case reports of plexiform schwannoma arising in the external female genitalia. None of the reported patients were associated with neurofibromatosis. Summary of these findings is shown in table 1.

Table 1

No.	Author	Year	Age (years)	Neurofibromatosis	Site	Recurrence
1	Woodruff *et al*.[14]	1983	26	-	Clitoris	Twice
2	Santos *et al*.[11]	2001	5	-	labia minora	-
3	Agaram *et al*.[15]	2005	26	-	labia minora	-
4	Chuang *et al*.[16]	2007	41	-	Clitoris	-
5	Yegane *et al*.[17]	2008	6	-	Clitoris	-

In conclusion, the ancient or benign schwannoma of vulva is a rare occurance and it should be considered in the differential diagnosis of a vulvar mass. Accurate diagnosis should be made on histopathological and IHC findings for optimizing the treatment.

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