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Case Report

Schwannoma of the clitoris: a case report

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Schwannoma (neurilemoma or neurinoma) is a tumor that develops from the Schwann cells in the myelin sheath of the peripheral nerves. It is most commonly observed on the head, neck, upper and lower extremities, posterior mediastinum and retroperitoneum. Schwannomas are rarely found in the external female genitalia. To the best of our knowledge, only 6 cases of clitoral schwannoma have been reported in medical literature. We currently report the case of a 46-year-old woman who was presenting with a 3 × 2 cm, painless mass in her clitoris for the past two years. She has no history of neurofibromatosis. Surgical excision of the mass was undertaken. Histopathology and immunohistochemistry revealed schwannoma of the clitoris. No evidence of local recurrence was noted after 6 months of follow-up. We are reporting the current patient because of the rare location of schwannoma. In the present case report, clinical features, treatment guidelines, diagnostic features and the immunohistochemical characteristics of this tumor are reviewed.

Keywords

Clitoris; Schwannoma

1. Introduction

Schwannoma (neurinoma, neurilemoma) is the encapsulated solitary tumor of the peripheral nervous sheath and it is generally benign, with < 1% of tumors presenting as malignant [1–3]. Schwann cells assist in the conduction of nerve impulses. Schwannoma can either occur in patients with neurofibromatosis or sporadically. It is most common between 20-40 years of age in female population [4–7]. Schwannomas are generally observed on the head, neck, upper and lower extremities, retroperitoneum and posterior mediastinum. They are rarely located in the female genitalia [1, 2, 5, 8]. Surgical removal is sufficient to treat the tumor and results in an excellent postoperative prognosis with rare instances of recurrence [5, 8]. In the present study, we describe clinical features, treatment guidelines, diagnostic features and the immunohistochemical characteristics of a benign schwannoma.

2. Case

A 46-year-old woman, gravida 4, para 2, presented with a painless vulvar swelling which had been present for two years. She had no symptoms of irritation or bleeding. She had a history of two dilatation and curettages for abortion and two cesarean sections. Her vulvar mass had gradually increased in size.

The patient was in good general health and had no significant weight loss. Her previous menstrual history was regular. She had a history of a right upper extremity operation due to a radial bone fracture eight years ago. Her past medical and family history were negative for any serious disease except hypertension, for which she was also using hypertensive medication.

The patient initially attended our gynecology clinic on November 2019. She had a nontender, mobile solid mass in her clitoris. She was referred to our gynecological oncology clinic for further evaluation and excision of the mass.

Upon physical examination, we observed an approximately 2 × 3 cm in diameter, palpable, subcutaneous, mobile, non-tender, oval, well-circumscribed and solitary mass lesion located at her clitoris. The mass was attached to the vulvar skin, but not fixed to any deep structures. There was no associated pain, bleeding and ulceration. The cervix, uterus, and bilateral attachments did not exhibit any abnormalities upon palpation. Pelvic, abdominal, urological examinations were all within normal limits. Upon physical examination, no other abnormalities were observed except the vulvar swelling (due to the lesion) and there was no inguinal lymphadenopathy. There were no clinical signs indicative of neurofibromatosis 1 and 2 or hyperandrogenism.

A solid mass of 29 × 24 mm was measured by ultrasonographic examination and transvaginal ultrasonography findings were normal. There were no abnormal findings during the pre-operatory laboratory tests and chest radiograph.

She was operated two weeks after the initial examination. A urinary catheter was inserted into the bladder. The solid mass was removed for diagnosis and treatment, with clear margins and without any perioperative complications. The tumor was resected via sharp dissection with some additional, marginal normal tissue around it. The resected material was sent to the histopathology department. Macroscopically, the resected specimen was nodular, measuring 3 × 2 × 2 cm. in size and weighed 8 grams. The tumor section revealed a mass, yellow in color and containing cystic areas. The immunohistochemical staining confirmed a histopathological diagnosis of benign schwannoma.

The tumor was well circumscribed and, upon microscopic examination of the excisional biopsy by hematoxylin eosin (Fig. 1a), commonly displayed a foci of cyst formation. The Antoni A areas illustrated show nuclear palisading with Verocay bodies (Fig. 1b). Differentiated Schwann cells evidencing S-100 protein were also observed in the schwannoma (Fig. 2).

There has been no recurrence of the tumor during the follow-up period of 6 months. Written, informed consent was obtained from the patient for the use of her data.

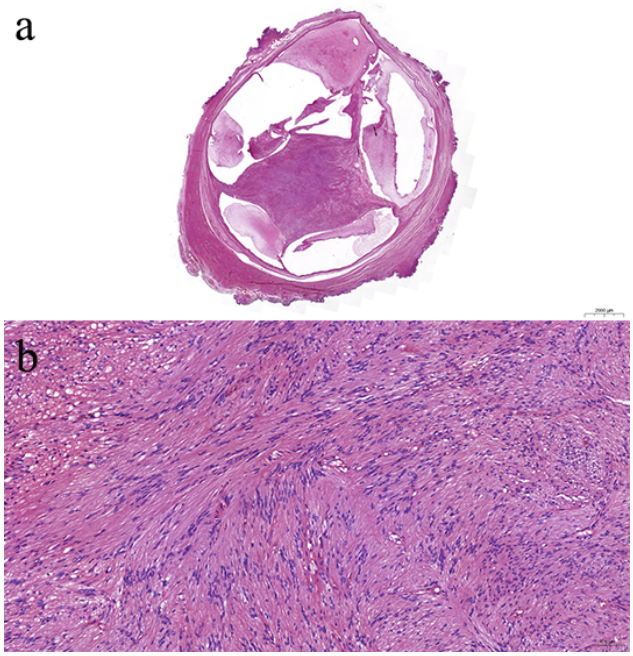

Fig. 1. (a) Tumors are well circumscribed and commonly display foci of cyst formation; excisional biopsy (H&E ×100). (b) Antoni A areas illustrating nuclear palisading with Verocay bodies. High power view shows nuclear palisading (H&E ×400).

3. Discussion

Schwannoma is typically an encapsulated, solitary and usually benign tumor that has arisen from Schwann cells [5]. Schwannoma can arise from neurofibromatosis or it can occur sporadically [5, 9]. Schwannoma slowly increases in size before becoming symptomatic and presents with nonspecific symptoms. The size of the tumor and the nerves affected are important factors for clinical symptoms. As mostly painless, nontender mass lesions are observed, small lesions are usually discovered by chance [5, 9, 10]. The patient in the current case had no symptoms of irritation or bleeding except a painless vulvar swelling which had been present for two years.

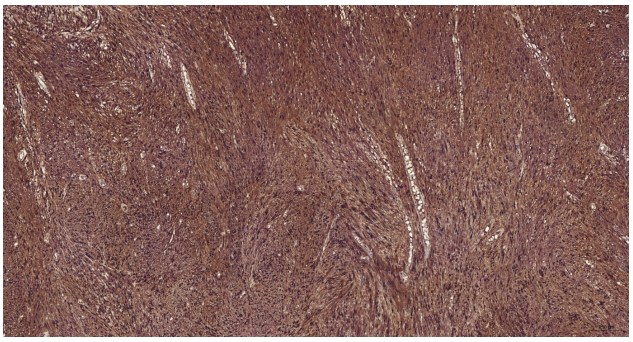

Fig. 2. Differentiated Schwann cells expressing S-100 protein in a schwannoma; (IHC ×400).

Schwannomas are generally located on the head, neck, flexor surface of the limbs or the posterior mediastinum [5, 7]. They infrequently occur in the vulva and the other female genital organs [2, 5, 7]. The clitoris has a rich nerve supply, however schwannoma of the clitoris is extremely rare. To the best of our knowledge only 6 cases of clitoral schwannoma have been reported in medical literature from 1983 to 2019 [1–3, 11, 12]. The age of the patients with vulvar schwannoma ranges between 5-84 years, mostly observed between 20-40 years in female population [4–7]. In the current case, the patient was a perimenopausal middle-aged woman.

In practice, schwannomas must be differentiated from other important neural tumors in this location, such as neurofibromas, granular cell tumors and malignant tumors, such as neurosarcoma, neurofibrosarcoma andmalignant schwannoma [5]. The differential diagnosis of schwannoma includes a bartholin cyst, lipoma, fibroma, epidermoid cysts, a labial cyst, liposarcoma, fibrosarcoma, angiosarcoma, and mesenchymal tumors [2, 7, 10, 13]. A wide spectrum of mesenchymal tumors can occur in the vulva and vagina of adults. As for prepubertal patients, the relative frequency of these tumors has not been precisely reported since such tumors are extremely rare in young girls [14].

Considering subtypes of schwannomas, a conventional variant of the vulvar schwannoma, such as a uninodular tumor, is most commonly observed histologically [5, 9]. A plexiform or a multinodular pattern has been reported in about 5% of schwannomas, and most plexiform schwannomas are sporadic tumors [11].

In terms of the resources for diagnosis, radiological imaging is useful for therapeutic planning, as it may provide information about the tumor's size, location, and possible invasion of other structures that enable the discrimination of schwannoma from other soft tissue tumors; as well as differentiating

the benign forms from the malignant ones. In the current case, the patient had a solid mass measuring 29 × 24 mm. By conducting a transvaginal ultrasonographic examination, it was clearly seen that the mass was not fixed to the urethra or any other structures. Hence, we did not need to utilize any further radiological imaging methods. In most cases, the preoperative diagnosis of schwannoma is difficult and a biopsy is often needed to exclude a malignant neoplasm and to determine an appropriate treatment method [2].

In the field of gynecology, schwannoma and neurofibroma are the most common types of benign peripheral nerve sheath tumors [1]. A combination of clinical, pathological, and immunohistochemical studies can confirm the diagnosis [15]. Microscopic observation followed by hematoxylin eosin staining can also predict the differential diagnosis by the characteristics found in the histologic morphology. Tumor cells may present basal lamina components including laminin and type IV collagen [1]. They may also contain vimentins or KP1 (CD68) and glial fibrillary acidic proteins. Histologically, Antoni type A and B, spindle cells, encapsulation, and diffuse immunostaining for S-100 proteins characterize benign schwannomas and distinguish them from neurofibromas [1, 15]. In such cases, the cells are positively stained for S-100 protein in benign schwannomas. The dedifferentiated Schwann cells in malignant schwannomas are believed to have lost their capacity to synthesize S-100 protein [2].

For benign schwannomas, complete excision is sufficent and the decisive treatment modality is surgical removal [5]. As previously stated, the majority of schwannomas are benign, slow-growing tumors with less than 1% of them being malignant. The prognosis for patients is generally good and recurrence is rare. Recurrences have been detected in a few patients following incomplete removal. A clear resection of the margins is important for the prevention of the recurrence [10]. Again, in our case, a painless, nontender mass was palpated upon physical examination of our patient. A complete, wide surgical resection was performed and there has been no recurrence in the patient during the follow-up period of 6 months.

4. Conclusions

With this paper, we have presented an exceptional case of clitoral schwannoma. Schwannoma is a benign encapsulating neoplasm, arising mostly in the head, neck, and trunk regions. To the best of our knowledge, schwannoma is rarely located in the clitoris, with only 6 cases having been previously reported in medical literature [1, 2, 11, 12]. Ultrasonography can be used to differentiate between solid and cystic tumors. A CT scan and MRI are also helpful in determining size, location, local involvement and distant spread. After surgical excision, histopathology and immunohistochemical determination results in a definitive diagnosis. As the surgical resection is curative, the probability of a malignant schwannoma or the risk of malignant progression and/or recurrence is very low in such cases. As such, schwannomas should be included among the differential diagnoses of benign vulvar tumors.

Author contributions

Alper Kocak is the assistant operator in the present case, analyzed the data, wrote the paper, Belgin Selam analyzed the data, wrote the paper, Eylem Akar analyzed the data, Serkan Erkanli is the operator in the present case, conceived and designed the manuscript

Ethics approval and consent to participate

All subjects gave their informed consent for inclusion before they participated in the study.

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Conflict of interest

None of the authors have any relationships with the companies and financial interest in the information contained in the manuscript. The authors declare no competing interests.

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