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VAGINAL SCHWANNOMA: CASE REPORT

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ABSTRACT

39-year-old female, P2L2, presented with complaints of vaginal pain while sitting. No complaints of abnormal discharge or bleeding per vaginum. Bowel and bladder habits were normal. No history of similar complaints in other family members. Pelvic examination revealed a mass of 3x3cm size in the posterior vaginal wall. On palpation, uterus was of normal size. Hard mass with sharp edges was felt beneath the posterior vaginal wall, not freely mobile. Mass was felt 3cm from the introitus. Rectal mucosa was normal.
No other lumps or masses detected on the skin or mucosa.
Ultrasonography of pelvis revealed a round hypoechoic area of size 3x3cm in lower vagina. A decision was taken to excise the mass, which was done through a vertical incision on the posterior vaginal wall. The mass was enucleated. Histopathological examination revealed a benign neural tumour-Schwannoma.

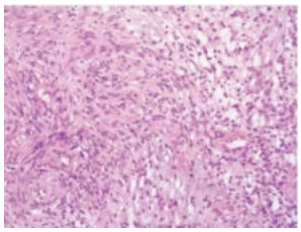
Vaginal Schwannoma: Case Report

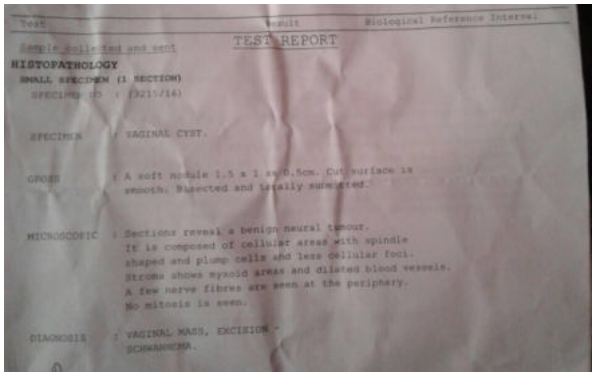
39-year-old female presented with complaints of vaginal pain while sitting. No complaints of abnormal discharge or bleeding per vaginum. Bowel and bladder habits were normal. Patient had previous two normal vaginal deliveries with uneventful antenatal and post-natal period. Last childbirth was 15 years ago. No menstrual complaints M/C 3-4/30days, normal flow, not associated with dysmenorrhea. Tubectomy surgery done 15 years back. No history of hypertension, diabetes, bronchial asthma or thyroid disorder. No history of similar complaints in other family members. Pelvic examination revealed a mass of 3x3cm size in the posterior vaginal wall. No ulceration or abnormal discharge noted. Cervix appeared normal.

On palpation, uterus was of normal size. Hard mass with sharp edges was felt beneath the posterior vaginal wall, not freely mobile. Mass was felt 3cm from the introitus. Rectal mucosa was normal.

No other lumps or masses detected on the skin or mucosa.
Ultrasonography of pelvis revealed a round hypoechoic area of size 3x3cm in lower vagina. Uterus and adnexa appeared normal.

A decision was taken to excise the mass, which was done through a vertical incision on the posterior vaginal wall. The mass was enucleated and sent for histopathological examination, which revealed a benign neural tumour- Schwannoma.





TEST REPORT

HISTOPATHOLOGY

SMALL SPECIMEN (1 SECTION)

SPECIMEN : VAGINAL CYST.

GROSS : A soft nodule 1.5 x 1 x 0.5cm. Cut surface is smooth. Bisected and totally submitted.

MICROSCOPIC : Sections reveal a benign neural tumour. It is composed of cellular areas with spindle shaped and plump cells and less cellular foci. Stroma shows myxoid areas and dilated blood vessels. A few nerve fibres are seen at the periphery. No mitosis is seen.

DIAGNOSIS : VAGINAL MASS, EXCISION - SCHWANNOMA.

Image: Histopathology confirming the diagnosis of vaginal schwannoma

Discussion:

Schwannoma is a benign nerve sheath tumour composed of Schwann cells, which produce the insulating myelin sheath covering peripheral nerves.

They are usually slow growing. Less than 1% becomes malignant known as neurofibrosacoma. Schwannomatosis is the third type of neurofibromatosis type 1 and type 2 characterised by multiple non-cutaneous schwannomas in the absence of bilateral vestibular schwannomas.

Incidence of primary nerve sheath tumours of central nervous system is 1.1 per 100,000 person-years (CBTRUS 1995-1999 and LACCSP 1995-1998). Incidence of multiple schwannomas is 5-10%.

Schwannomas of head, arms, neck and trunk are more

common(Biswas, 2007). It is more common in age group 20-50 years.

Schwannomas originating from clitoris, vulva, vaginal wall, cervix, uterus, fallopian tubes, ovaries, pelvic floor have been documented (S Kulkarni, 2005).

They sometimes occur in people suffering from neurofibromatosis which is due to gene mutation(Genetic Home reference December 2013). Germline mutations in tumour suppressor gene, SMARCB1 and LZTR1 appear to explain the predisposition to schwannomatosis(Smith MJ, 2012; Smith MJ, 2015). Most schannomas are not inherited. 20% of cases appear to be familial defined by one or more closely-related relatives with at least one schwannoma(Jacoby LB, 1999). Schwannoma usually occurs as a single tumour(Gonzalvo A et al, 2011). Most common complaints of genitourinary schwannomas are localised pain, discomfort, abnormal vaginal discharge or they may be asymptomtic. Vaginal schwannomas, depending upon their position, may cause pressure symptoms ie. bowel or bladder disturbances. And may cause disturbance in sexual function or dyspareunia.

Vaginal schwannomas are usually around 3-4cm in size.
Schwannoma ocurring in other parts of the body may present with abnormality of peripheral nervous system electrophysiology. The most important differential diagnosis in patients with multiple schwannomas is neurofibromatosis-2. Other differential diagnosis, include calcifying aponeurotic fibroma, fibrous histiocytoma, leiomyoma, leiomyosarcoma, appendiceal smooth muscle.

Diagnosis requires a detailed clinical and family history. Diagnosis may be aided by X-ray, ultrasonography, CT scan. High resolution MRI is useful in assisting in diagnosis. MRI with or without contrast is required. MRI with short T1 inversion recovery (STIR) sequences can help visualise nerve sheath tumours.

Diagnostic criteria — The diagnosis of schwannomatosis is based on clinical criteria first proposed in 2005 and modified in 2006(MacCollin M, 2012). According to the 2006 criteria, patients can be diagnosed with definite, possible, or segmental schwannomatosis.

A definite diagnosis of schwannomatosis can be made in either of the following circumstances:

(I) Age >30 years and all of the following:
- Two or more nonintradermal schwannomas, at least one with histologic confirmation
- Diagnostic criteria for NF2 not fulfilled
- No evidence of vestibular schwannoma on high-quality MRI scan
- No first-degree relative with Nf2
- No known constitutional NF2 mutation
- One pathologically confirmed nonvestibular schwannoma plus a first-degree relative who meets the above criteria
- Criteria for possible schwannomatosis are met in any of the following circumstances:

(II) Age <30 years and all of the following:
- Two or more nonintradermal schwannomas, at least one with histologic confirmation
- Diagnostic criteria for NF2 not fulfilled
- No evidence of vestibular schwannoma on high-quality MRI scan
- No first-degree relative with Nf2
- No known constitutional NF2 mutation

(III) Age >45 years; no symptoms of eighth cranial nerve dysfunction; and all of the following:
- Two or more nonintradermal schwannomas, at least one with histologic confirmation
- Diagnostic criteria for NF2 are not
- No first-degree relative with Nf2
- No known constitutional NF2 mutation
- Radiographic evidence of a schwannoma and first-degree relative meeting criteria for definite schwannomatosis

Biopsy of the tumour for confirmatory of diagnosis of schwannoma.

Histopathology: Macroscopic/Gross description of usually solitary; large tumours may be cystic; the nerve of origin present in the periphery.

The main microscopic feature being the presence of alternating areas of compact spindle cells arranged in compact fascicles called Antoni A areas, and less cellular and more disorganised areas called Antoni B areas (WHO classification of tumours of the Nervous systerm, 2007). Other histopathological features include biphasic compact hypercellular cells; myxoid hypocellular; narrow elongated cells, wavy cells with tapered ends interpersed with collagen fibres; nuclear pallisading; ill-defined cytoplasm with dense chromatin; characteristic luse bodies (long spaced collagen); foamy macrophages; nuclear atypia; amianthoid fibres/ collagenous spherules; rarely epitheloid glandular pigmented, plexiform rosettes; rare mitotic figures, axons where the nerve is attached.

Treatment of solitary genital schwannoma is by excision. Primary treatment for Schwanomatosis, is symptomatic approach. For multiple schwannomas, pain management with gabapentin and pregabalin, use of short acting opioids, non-steroidal antiinflammatory drugs. Non-pharmacologic therapy by following yoga, meditation mindfulness-based stress reduction (MBSR) can help deal with relaxation, stress reduction and training the mind to be less reactive to pain(Rosenzweig S, 2010). Malignant schwannomas may require chemotherapy or radiotherapy. Gene testing can be done for both SMARCB1 and LZTR1.

Prognosis: 40-60% do not enlarge, 4-12% of tumours involute during long term management. Recurrence rate: 1-2%.

Although vaginal schwannoma is a rare tumour, it should be considered in the differential diagnosis of vaginal lesions and proper knowledge of schwannomatosis will help us in complete treatment and avoid several diagnostic pitfalls.

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