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Original Research Paper

A Rare Case Report of Myxoma of Round Ligament at Labia Majora

General Surgery

KEYWORDS : myxoma, round ligament, processus vaginalis, labia majora

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ABSTRACT

Myxomas are a variety of benign tumors that occur in various locations, including the heart, bones, skin, subcutaneous and aponeurotic tissue, genitourinary tract and skeletal muscle.

We report a case of myxoma ofround ligament, presenting as swelling at right labia majora, of one month duration in a 29 year old multiparous female. On examination it was a soft, non-tender, fluctuant 3cms diameter swelling, which is reducible on supine head low position, expansile cough impulse was noted. Swelling was palpable through the right wall of vagina. Wide bore needle aspiration was done to rule out cystic nature. Plain Computerized Tomography revealed a patent processes vaginalis. Diagnostic laparoscopy was done to rule out obturator hernia. An incision was given directly over the swelling and excision was done. Grossly it was a gelatinous lesion resembling fetal umbilical cord andit histopathological examination revealed the diagnosis of myxoma

Case report:

A 29 year old multiparous female, homemaker, presented with complaint of swelling in right labia majora of one month duration. The swelling started as a small elevation in the right labia majora, slowly progressed in size to attain the present size. There was no history of pain at the swelling, chronic cough, fever or trauma. No history of discharge per vagina or pain during coitus. Sleep and appetite are unaltered. She attained menarche at the age of 15 years, has regular 5/30 days cycles. She gave birth to two children, last child birth was 10 months back and both were lower segment caesarian sections.

On examination, in lithotomy position a single, soft swelling of about 3x3 cms was noted in the right labia majora. On standing and straining, the swelling increased in size. The surface of the swelling appears smooth and the skin over the swelling is normal. Ther is no warmth or tenderness over the swelling. The swelling is reducible and cough impulse present. There was no regional or generalised lymphadenopathy. Howship Romberg sign was negative.

Figure 1: Clinical Picture showing the swelling and its relations.



Investigations

Routine blood tests were normal. Urine routine was under normal limits.

CT scan shows evidence of a well-defined ovoid hypodense area of fluid attenuation measuring 10x5cms posterior to urinary bladder extending to right labium majora. The superior aspect this area is leading to narrowed abrupt tapering (narrowed neck) with suspicious communication with perilabial area. The abdominal wall musculature and the bony skeleton in the scanned region appear within normal limits, leaving an impression of hydrocele of persistent Processus vaginalis (Hydrocele of Canal of Nuck).

Histopathology of excision biopsy specimen showed features suggestive of myxoma i.e., structures of fibrocollagenous tissue with sheets of myxoid stromal cells with background of myxomatous areas with uniform spindle cells with occasional vascular proliferations in between.

Surgery

Diagnostic laparoscopy was done to rule out pelvic/obturator hernia. Hernia orifices where normal and there was no extent of swelling into the abdominal cavity. An incision was given directly over the swelling and excision was done. Grossly it was a gelatinous lesion resembling fetal umbilical cord andit histopathological examination revealed the diagnosis of myxoma.

Figure 2: Intra Operative picture

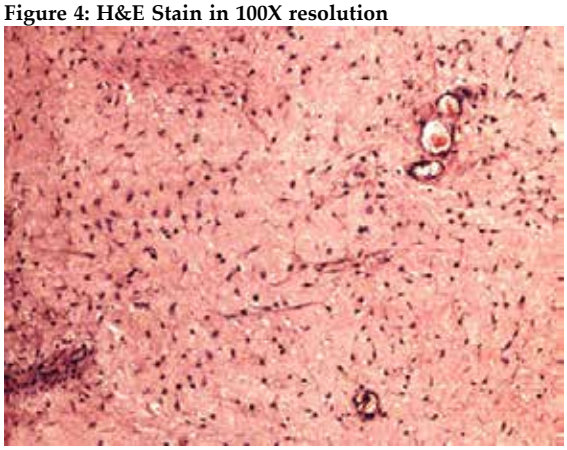


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Figure 3: Specimen and its cut section



Figure 4: H&E Stain in 100X resolution



Discussion:
Round ligament is embryologically a mesenchymal structure formed by the caudal portion of the mesonephric fold, equivalent to gubernaculum testis in males1,2. It begins at the cornua of the uterus and extends through the inguinal canal to the labia majora. The most commonly found tumors are leiomyomas, followed by endometriosis and mesothelial cysts3.

Myxomas are rare neoplasms (pseudo sarcomas) that are mucoid, slimy and extremely gelatinous commonly appearing as circumscribed lesions deep within the muscle, usually in an extremity4. Strangely enough for a nearly avascular lesion, an occasional myxoma may reach a considerable size of 10-13cms. They almost always occur in adults and are more common in females5. Myxomas develop as painlessmasses growing slowly or are stationary for long periods before suddenly enlarging.The duration of development of the lesion may vary from few weeks to as long as 10 years. There is no apparent relationship between the size of the myxomaand the duration of its presence.

A high proportion of myxomas arise within the skeletal muscle (intramuscular myxomas). They especially tend to involve large muscles like thigh, buttocks, upper arm and shoulder. Myxomas also occur in the head and neck, hand, leg pelvic girdle, chest wall, axilla, clavicular and scapular areas. Multiple intramuscular myxomas are seen in association with fibrous dysplasia of bones of the same extremity.Other important location is juxta articular(particularly knee6),subcutaneous or from fascial planes and neurovascular sheaths.Presence of multiple myxomas should arise the possibility of Carney complex7.

Grossly, myxomas are sharply circumscribed and may be encapsulated when in superficial soft tissue8. Intramuscular myxoma, lacks a true capsule, and histologically infiltrates the involved muscle8, 9, 10 and may also attach to the periosteum of adjacent bone or to the overlying fascia4, 11.

Microscopically a typical myxoma has a bland and hypocellular appearance. Mitotic activity is absent and blood vessels are extremely scanty (well documented with angiographic and microangiographic studies).Towards the periphery,an increased number of spindle cells abut on a lightly collagenized capsule. Centrally, a mucinous cyst or focal eosinophilic fibrinous quality can often be appreciated. These attributes suggest a reactive lesion; indeedmyxomas rarely recur. Focal aggregates of foamy histiocytes with neutral fat with the oil red O stain may be present (Not to be confused with lipoblasts of myxoidliposarcoma)4. The lesion consists of a slightly basophilic proteoglycan matrix. At high power, a few stellate or bipolar cells with oval nuclei are seen.

Immunohistochemically, myxoma shows no reactivity for S-100 protein, this being another difference with myxoid liposarcoma4. Desmin is absent, but vimentin is expressed12. The myxoid material is entirely digestible by hyaluronidase.

Differential diagnosis
Several tumors and tumor like conditions of soft tissues have a mucinous stroma resembling myxoma11,13. The differential diagnoses comprises of two groups of diseases14. The first is a group of neoplasms with myxoid change as prominent secondary feature like myxofibrosarcoma, aggressive angiomyxoma, myxoidliposarcoma, chondrosarcoma, leiomyosarcoma, embronal rhabdomyosarcoma, neurofibroma, and nerve sheath myxoma / neurothekeoma15. The second group of diseases are non-neoplastic disorders resulting in focal mucinous degeneration of the skin or soft tissues, such as, mucinous (myxoid) cyst, nodular fasciitis, localized myxedema, follicular mucinosis (alopecia mucinosa), papular mucinosis and cutaneous focal mucinosis16.

Myxoid degeneration of Leiomyoma of round ligament, bartholins cyst/ abscess, obturator hernia, endometriosis, fibromas of vulva, fibroadenoma of vulva and parasitic leiomyoma are other possible differential diagnoses. Vulvar leiomyomas are extremely rare tumors felt to arise from either smooth muscle within the vulval erectile tissue or by transmigration through the round ligament17. Based on clinical examination and duration of the lesion, myxomatous degeneration of leiomyoma, fibromas of vulva (Primarily found in labia majora and range in size from 0.6-8 cms) can be excluded. Fibroadenoma of vulva arises from embryonic breast tissue, the caudal end of the milk line.

Aggressive angiomyxoma is a locally aggressive myxoid tumor, histologically similar to a myxoma but contains a prominent vascular component. It occurs predominantly in women and involves the genital tract, pelvis, and perineum18,19 with a significant recurrence rate after excision. Thelesion is larger involving deeper structures. A counterpart located in the superficial soft tissue called cutaneous myxoma (su-

perficial angiomyxoma), has been described arising more commonly in middle-aged males.Some arise in the genital region20 and so may be confused with aggressive angiomyxoma.

Myxoidliposarcoma is more deeply located and larger than cutaneous myxoma, and it is characterized by a plexiform vasculature with scattered lipoblasts. The possibility of low grade myxofibrosarcoma should be considered if a myxoidtumour is hypercellular and hyper vascular. Myxofibrosarcoma has a greater degree of nuclear atypia and hyperchromasia as well as curvilinear vessels often lined by hyperchromatic tumor cells21.But a cellular variant of myxoma has been described which is more cellular and vascular than the ordinary type, but lacks the pleomorphism, nuclear atypia, and curvilinear vascular pattern of low grade myxofibosarcoma.

Cutaneous myxoid cyst is exclusively location on the fingers. Neurothekeoma has a more pronounced lobular growth pattern and is characterized by plumper cells, positive for S-100 protein. Myxoidneurofibroma is composed of cells with wavy or buckled nuclei that are also S-100 protein-positive. Focal cutaneous mucinosis lacks the lobular architecture, stromal neutrophils, and epithelial structures found in cutaneous myxoma.

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