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# Inguinal swellings with diagnostic challenge - 2 Case Reports

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**Abstract**

The aim of the case report is to depict inguinal swellings with diagnostic challenge through 2 case reports.

**Case report-1:** Angiomyofibroblastoma (AMFB) is a benign mesenchymal tumour most commonly found in the female genital tract of premenopausal women. Although rare, AMFB is an important consideration in the differential diagnosis of inguino-scrotal swellings in both the sexes. We are describing a case of left inguinal region angiomyofibroblastoma which was treated surgically and was confirmed histopathologically.

**Case report- 2:** Round ligament tumours are rare which includes leiomyomas, mesothelial cysts and some cases of endometriosis. Leiomyomas are the commonest of them.

These usually present as an inguinal swelling mimicking an inguinal hernia or a lymph node.

We are describing a case of a 47-year-old lady who presented with a right sided labial swelling with lower abdominal pain and had surgical intervention for the same.A round ligament leiomyoma was confirmed by histopathology.

**Keywords:** Angiomyofibroblastoma,Mesenchymal tumours, round ligament leiomyoma, Inguinoscrotal swellings, Desmin, Labial swellings.

**Case report-1**

**Introduction**

Angiomyofibroblastoma (AMFB) commonly occurs during the fifth to eighth decades of life and mainly involves the inguinoscrotal region. It is derived from perivascular stem cells and has the capacity of lipoid and myofibroblastic differentiation. Angiomyofibroblastoma (AMFB) predominantly occurs in the female genital tract, such as the vulva, perineum, vagina and pelvis. In 1992, Fletcher et al [1] first described a rare, benign tumour that occurs in the reproductive system of middle-aged women, known as AMFB. Thereafter, in 1998, Laskin et al [3] reported 11 cases of similar entities in males and suggested the term AMFB tumour (also known as cellular angiofibroma). The study reported uncharacterized mesenchymal tumours resembling female AMFB which arise in the inguinoscrotal region of adult men [3]. This entity in male genitalia is exceedingly rare and has been described as its female analogue or under the name of male AMFB tumour [3, 4] and are known to occur in regions such as the inguinal area, scrotum and perineum. AMFB tumours have been described only in isolated case reports. Clinically, the tumour has asymptomatic, well-circumscribed and slow growing characteristics. The current case report presents a case of AMFB tumour in the inguinal region.

A 46-year-old male presented to General Surgery OPD with a painless mass in the left inguinal region with an USG Abdomen report which revealed findings suggestive of obstructed hernia. However, on clinical examination sonographic findings were not correlating. The patient had observed that the swelling had gradually increased in size during recent months.

On physical examination at the time of admission, a firm, painless mass was palpated in the left inguinal region and his laboratory data were within normal limits. The preoperative diagnosis was of a left inguinal mass of undetermined origin.

An inguinal exploration was planned and upon opening the inguinal canal a well-encapsulated, pleomorphic tumour, measuring 7.5 × 4.0 cm postero-medial to spermatic cord. Microscopically, the tumour was a well circumscribed lesion with prominent vascular component and a loose oedematous stroma. The vessels were small to medium sized, thin walled and ecstatic. Larger, dilated vessels were seen in between. The component cells had scanty cytoplasm and round to ovoid nuclei. Plasmacytoid cells and many scattered mast cells are seen. Focally, paler and hypocellular appearing areas were present. There was no mitotic activity or necrosis. Fibro fatty tissues with congested, thick-walled vessels were seen outside the lesion. By immunostaining, the tumour cells stained positive for Desmin& CD34 and negative for S-100, smooth muscle actin (SMA) &Estrogen Receptor (ER).

Figure 1:

Figure 2:

Figure 3:

[The left inguinal mass: postero-medial to left spermatic cord (held by Babcock's forceps)]

**Discussion**

Mesenchymal neoplasms of the genital tract occur predominantly in the vulva, perineum, and pelvis of women but have also been described in men associated with the spermatic cord, inguinal hernias, scrotum, and perineum. Some of these neoplasms occur with or without myofibroblastic differentiation or as a spectrum of spindle cell tumours [12,14]. Although subtle morphologic and immunohistochemical differences between these lesions do occur, they are Histo-genetically related lesions arising from a common precursor cell [8]. One of the mesenchymal neoplasms is AMFB that was described by Fletcher CD et al. in 1992 [1]

Male genital AMFB share many clinical and pathologic features comparable to those described in their female counterparts. These lesions almost exclusively involve the inguinal region and scrotum. To date, a total of only 15 cases of the male AMFB have been documented in the literature, including 9 cases in the scrotum and 6 cases in the inguinal region [1,3,5,6]. Based on the analysis of 11 cases of male AMFB, Laskin et al [3] proposed the view that the male AMFB tumours, including spindle cell lipoma, encompass a broad spectrum of soft-tissue neoplasms, presumably originating from perivascular stem cells that may potentially undergo myofibroblastic differentiation. Although the exact nosologic position of AMFB still remains unknown, we conclude that our case represents male AMFB, based on its conventional histopathologic and immunophenotypic features.

The immunoprofile of the male neoplasm, although still consistent with myofibroblastic differentiation, varied slightly from the female AMFB. Desmin expression was noted in 37.5% whereas the figure is 94% for the female AMFB reported in the literature. 50% reacted with SMA, whereas this figure is only 15% for all female AMFB. The mean estrogen and progesterone receptor scores were substantially higher in the female AMFB [1]. The immunoreactivity of neoplastic cells for desmin

is a common finding in Angiomyofibroblastoma; it is occasionally desmin negative, especially in examples arising in male patients [2,14]. Histo-genetically, Laskin WB et al. believes that the male AMFB tumour, like the female AMFB, is derived from a perivascular stem cell with a capacity for fatty and myofibroblastic differentiation governed by hormonal, microenvironmental, and growth factor/cytokine-related influences [3]. This progenitor cell may be related to the CD34-positive fibroblast-like cell that normally resides around vessels. A CD34-positive fibroblast-like cell composes the spindle cell element of spindle cell lipoma. In the male AMFB tumour, this cell may theoretically lose its ability for CD34 expression with myofibroblastic differentiation [1,4].

**Conclusion**

Though extremely rare, in a patient with an inguino-scrotal swelling, it is important to consider AMFB among the differential diagnoses, particularly because of the importance of distinguishing AMFB from aggressive Angiomyxoma. However postoperative histopathological examination is needed for definitive diagnosis.

**Case report- 2**

**Introduction**

Leiomyomas are the most common gynecological tumours with a prevalence of 20 -30% among women and are most commonly found in the uterus [1,2]. The round ligament in females is the embryological equivalent of gubernaculum testis in males and is composed of mainly smooth muscle fibres and it extends from its origin at the uterine horns, in the parametrium and passes through the inguinal canal to the labia majora [3]. Leiomyomas of the round ligament is rare, commonly seen in premenopausal women and it usually presents as an inguinal swelling. Round ligament leiomyomas at abdominal or vulvar locations has also been reported [1, 4]. The Canal of Nuck is a small evagination of the parietal peritoneum, which is attached to the uterus by the round ligament through the internal inguinal ring into the inguinal canal. This structure is homologous to the processus vaginalis of the males [5]. Hydrocele of the canal of Nuck is a rare disease that occurs in the inguinal area or labium as a painless edema usually before adolescence [6]. Unilateral labial swellings can be a Bartholins cyst, furuncle, carcinoid and rarely adenomyoma of round ligament or a patent processsus vaginalis with ascites [7].

47-year-old lady presented with a lower abdominal pain and painful swelling of the right labia majora for 4 years which was gradually increasing in size. The pain was dragging in nature. There were no gastrointestinal or urinary symptoms or any history of associated fever or weight loss. On examination there was a healthy lower abdominal scar of TAH along with a 8 x 7 cm tender right groin swelling extending to the right labia majora. The patient was evaluated with an MRI scan which showed a thin walled enhancing cystic mass herniating through right inguinal ring to labium majus – suggestive of hydrocele of canal of Nuck or patent processsus vaginalis and a small bulge at the right inguinal ring.

She underwent right inguinal exploration and was found to have a 10 x 6 cm mass arising from the round ligament extending to the labia majora along with a direct inguinal hernia. The mass was excised and right inguinal hernioplasty was done and the specimen was sent for histopathological examinations. The histopathology reports were suggestive of round ligament leiomyoma with degenerative changes and immunohistochemistry showed positive for Desmin.

Figure 4: External oblique fibres are seen before opening inguinal canal

Figure 5: Forceps showing direct inguinal hernia

Figure 6:

**Discussion**

Round ligament, which is the embryological equivalent of gubernaculum testis in males, is composed of smooth muscle fibres and extends from the parametrium of the uterus to the labia majora and mons pubis passing through the inguinal canal. It is responsible for descent of ovary from the posterior abdominal wall to the uterus and is composed of connective tissues, smooth muscle fibres, vessels and nerves with a mesothelial coating. Estrogen and progesterone receptors are present on the surface of these smooth muscles. Round ligament tumours are rare and leiomyomas are the commonest among them followed by endometriomas and mesothelial cysts. These most commonly arise from the extraperitoneal end of the round ligament and are usually unilateral and common on the right side [16,17]. Leiomyomas of the round ligament usually presents as an inguinal swelling, but in our case the patient has presented with a labial swelling. Only one case of transformation of round ligament leiomyomas to leiomyosarcomas has been reported in the modern era [18].

Imaging modalities like an MRI or a CT can aid in the diagnosis. Surgical exploration and excision of the mass is the treatment of choice and gives the most accurate diagnosis which is confirmed by histopathological examination. Immunohistochemistry studies will show positive for desmin.

**Conclusion**

The round ligament leiomyomas presenting a labial swelling is a very rare condition. However it should be kept as a differential diagnosis with clinical radiological and pathological correlation. Surgical excision of the mass is considered as the most appropriate way of treatment. An inguinal approach would be sufficient as

the tumour can be delivered through the surgical wound and hernioplasty can be attempted, if necessary, like in our case. An MRI can aid us in the diagnosis but the accurate diagnosis is revealed intraoperatively and confirmed by Histopathological examinations.

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**Abbreviations**

AMFB – Angiomyofibroblastoma

SMA- Smooth Muscle Actin

ER- Estrogen Receptor

CD 34- Cluster of Differentiation -34 (Transmembrane phosphoglycoprotein)

TAH – Total Abdominal Hysterectomy