Subcutaneous Scrotal Leiomyosarcoma Presenting as Pedunculated Multi-locular Cystic Growth in the Scrotum Mimicking a Sebaceous Cyst: A Case Report and Review of the Literature

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Abstract

Background
Scrotal leiomyosarcomas are extremely rare and hence the biological behaviour of these rare tumours may not be known by most medical practitioners. To the knowledge of the authors about 37 cases of leiomyosarcoma of scrotum have so far been reported.

Aims
To report a case of leiomyosarcoma of scrotum
To review the literature on leiomyosarcomas of scrotum

Case Report
A 66-years-old man underwent a day case procedure excision under local anaesthesia of a pedunculated scrotal lump which had been annoying him for a number of years. Clinically, the lump was considered to be a benign lump which pre-operatively was thought to have features of a sebaceous cyst. However during the procedure of excision of the lump it was felt that the lump was firm and was most likely a fibroma. Based upon the fact that the lump was considered to be benign the patient was discharged from follow-up only to be referred but by his General Practitioner in case he developed any post-operative wound problems. Histology of the excised scrotal lump was consistent with that of scrotal leiomyosarcoma. At follow-up, 29 months pursuant to excision of the lump, there was no evidence of recurrence.

Literature review indicated that:
- Only 37 cases of scrotal leiomyosarcoma had previously been reported.
- Wide excision of scrotal leiomyosarcomas with a clear margin of at least 10 mm was associated with better outcome in comparison with margin involvement or tumour clear margin which was less than 10 mm.
- Late local recurrence and distant metastases occurred in some cases therefore long-term follow-up was recommended.
- The problem of incomplete resection or margin involvement occurs and in such situations re-excision should be performed.
- Chemotherapy may be effective in selected group of patients who have refused surgery.
- Radiotherapy and chemotherapy have been used in some cases but there is no universal agreement on their use as adjuvant therapy in all cases in the treatment of leiomyosarcoma of scrotum.
- Radiotherapy / chemotherapy should supplement and should not substitute complete excision of the lesion.

Conclusion
Leiomyosarcoma of scrotum is rare and this should be treated by excision of the lesion with a clear margin of at least 10 mm. There is a place for Adjuvant therapy (radiotherapy / chemotherapy) in some cases.

Key Words: Leiomyosarcoma; scrotum; wide-excision; 10mm; clear-margin; spindle-cell; tumour; immunohistochemistry; Vimentin; SMA;

Introduction
Leiomyosarcoma (LMS) of scrotum is a rare tumour. Scrotal leiomyosarcomas (LMS) are slow growing tumours that present as firm rubbery non tender irregular masses [1]. They may arise from paratesticular or scrotal skin. Over 95% of all paratesticular leiomyosarcomas are located in the spermatic cord or epididymis, and their location in the scrotal skin or subcutaneous position in the scrotum is exceptionally rare. To date approximately 37 cases of scrotal leiomyosarcomas have been reported in the literature. We report a case of subcutaneous leiomyosarcoma which was presented as an annoying pedunculated scrotal lump with a review of the literature.

Case Report

A 66-years-old man was seen in the out-patients clinic complaining of an annoying lump which had been protruding on his scrotum for a long time. There was no associated pain. There was no sudden increase in size of the lump. His past medical history included diverticulosis; cervical spondylosis; chronic obstructive airway disease/asthma; hypertension and diabetes. His medications included: Metformin; Amlodipine;
Bendrofluazide; Salbutamol accuhaler; and Seretide accuhaler.

His general and systematic examinations were unremarkable but his scrotal examination revealed a pedunculated multi-locular cystic growth which in the opinion of the examining surgeon looked and felt like a sebaceous cyst; the lump was sub-cutaneous in position and was lying in the anterior aspect on both sides of the scrotum (extending to the left and right of the midline raphe). He was listed for removal of the pedunculated lump under local anaesthesia because the lump was amenable to excision under local anaesthesia instead of general anaesthesia in view of his diabetes and chronic obstructive airways disease/asthma and diabetes which were his co-morbidities.

He went on holidays for about six weeks and upon his return underwent excision of the scrotal lump as a day case procedure. The lump was completely excised with some normal surrounding tissue and during the procedure, it was noted that the pedunculated scrotal lump was located in the middle of the scrotum, large, mobile, and clinically felt firm. The features of the lump were suggestive of a benign scrotal lesion which was provisionally diagnosed as perhaps a fibroma or a dermoid cyst. The specimen was sent for histological examination. The wound was stitched using vicryl stitches. His post-operative recovery was unremarkable and he was discharged home with no routine follow-up appointment to be reviewed only if his histological examination revealed any un-usual pathology which was felt to be unlikely.

The histological examination by two pathologists was reported as follows:

- Macroscopic examination revealed a nodular portion of tissue 45 x 35 x 20 mm which on sectioning showed pale fibrous areas with occasional haemorrhagic spots and myxoid-change and no areas of necrosis.
- Microscopic examination revealed subcutaneous spindle cell tumour with focal necrosis, severe cytological atypia with some bizarre nuclei and high mitotic rate with atypical forms, in keeping with a malignant spindle cell tumour which was considered to be probably smooth muscle in nature (see illustrations [figures] 1 and 2). The features of the lesion were in the opinion of the pathologists consistent with malignant spindle cell tumour which required classification by means of immunohistological stains.
- Immunohistological examination, showed the tumour to have strong and diffuse positivity for SMA and Vimentin SMA (see illustrations [figures] 3 and 4) and negativity for S100 protein and CD34, in keeping with a leiomyosarcoma.

At follow-up 29 months after excision of the tumour the patient remained well without any evidence of any recurrence and would continue to be followed up by clinical examination and CT scan of thorax, abdomen and pelvis.

**Discussion**

Soft tissue Sarcomas constitute 1% of all malignancies. Leiomyosarcomas comprise of 10% to 20% of soft tissue sarcomas. Leiomyosarcomas arise most often from uterus, gastrointestinal tract and retroperitoneal region [1]. Subcutaneous leiomyosarcomas (LMS) comprise of 1% to 2% of all superficial soft tissue malignancies [2]. With regard to genitourinary sarcomas in adults, leiomyosarcomas are the most common type and they arise in the urinary bladder, kidney, or prostate. Leiomyosarcoma originating from the scrotum is exceptionally or extremely rare with only about 37 cases reported in the literature. Talikoti and associates [3] reported the 37th case of leiomyosarcoma of scrotum. Leiomyosarcomas are malignant mesenchymal line neoplasms whose tumour sizes vary from 2 cm to 9 cm, with an average size of 5 cm. Confirmation of the diagnosis of leiomyosarcoma is based upon histological examination of biopsy specimens. They typically show spindle cells with cigar shaped nuclei arranged in interweaving fascicles [1]. On immunohistochemistry leiomyosarcomas are positive for actin, desmin and CD 34 [4]. The mode of spread of leiomyosarcoma is primarily haematogenous to lung, liver, and bone. The prognosis of leiomyosarcoma depends upon the size, depth and grade of the tumour and evidence or no evidence of distant metastases.

It has been suggested that the rarity of leiomyosarcomas of the scrotum is such that a General Practitioner would usually see such a tumour once every 20 years [5]. In view of the fact that only about 37 cases of scrotal leiomyosarcomas have up till now been reported it would be argued that most practitioners would never come across a scrotal leiomyosarcoma in their lives. Leiomyosarcomas involving the tunica vaginalis or the spermatic cord are more frequent [4]. Clinically, paratesticular leiomyosarcomas manifest as painless scrotal lumps of several months to years duration occurring in patients with a mean age of 51 years [6]. Washecka and associates [7] stated that in leiomyosarcoma the diagnosis of malignancy is based on the mitotic rate of two to ten mitosis per high power field (2-10 mitosis /HPF), although the presence of nuclear pleomorphism, vascular invasion, tumour
In view of the rarity and heterogeneity of soft tissue sarcomas there have only been few studies done, which would explain the lack of a uniform follow-up strategy or post-operative management [8].

The need for proper clearance finds its root in the surgical management of scrotal leiomyosarcomas. Smooth muscle is seen in the skin in three locations, the arrectores pilorum muscles, the wall of vessels and the specialized muscles in genital skin (the dartos, vulvar, and mamillary muscles of the scrotum, labia majora, and the breast respectively [9].

Echenique and associates [10] stated that leiomyosarcomas of the scrotum usually present as firm, rubbery, non tender, irregular masses and scrotal leiomyosarcomas tend to be slow growing tumours that tend to be present for years. Johnson H Jr [11] reported the first case of leiomyosarcoma of scrotum in 1987. Kaushal and associates [2] stated that leiomyosarcoma of scrotum is often mistaken for a benign lesion and it is best treated by wide local excision. John and associates [12] stated that: owing to the small number of patients, well documented data regarding adjuvant therapy is limited; inguinal lymph node dissection is not advocated, unless a high degree of suspicion is present for lymph node metastasis.

Kaushal and associates [2] stated that:
- Radiation therapy has been of doubtful value in treatment of leiomyosarcoma except for palliation.
- Chemotherapy with gemcitabine, paclitaxel, vincristine, Doxorubicin-D, have been used with limited success.
- The recommended treatment of localized leiomyosarcoma of the scrotum is wide excision.
- Adjuvant treatment is not considered necessary however, locally aggressive tumours may recur.
- Chemotherapy may be effective in selected group of patients who refuse surgery.
- Long-term follow-up is essential, because of the risk of delayed local recurrence and distant metastasis. It has been stated that [13] the main problem associated with the treatment of leiomyosarcoma of scrotum is incomplete resection margin. It has also been stated that:
- The need for proper clearance finds its root in the pathology of soft tissue sarcomas.
- These grow by radial expansion, infiltrating the local tissues as they proliferate.
- They lack a surrounding capsule so that an excisional biopsy with a macroscopically clear margin will often leave microscopic tumour behind [6].

Catton and associates [14] reported a 27% microscopic residual tumour detection rate in cases of ‘completely excised’ tumours. Stojadinovic and associates [15] stated that studies at MSKCC had confirmed that the recurrence rate in the presence of a positive margin is increased 2.4 fold. Khatari and Goodnight [16] stated that failure of clearance of the margin or recurrence of the tumour necessitates further excision, which is fraught with problems. First of all, there is often a loss of anatomical planes as a result of the process of fibrosis which emanates shortly after surgery, which hampers the surgeon’s physical appreciation of the operating field. In addition, McKee and associates [17] stated that there is the risk of spillage or local spread of tumour through each operation. It has also been stated that inadequate clearance eventually causes a vicious cycle of repeat surgery and repeat spillage [13]. It has been clearly and generally accepted that the proper resection of a soft tissue sarcoma requires a surrounding margin to be excised. However, the actual size of an oncologically accepted safe margin has not been universally clearly defined, in view of the rarity of the tumour and the non availability of studies on those tumours. McKee and associates [17] depicted that the local recurrence-free interval at 5 years is higher with a margin of >10 mm (84%) than those of 0 mm (58%) or 1-9 mm (58%). The similar rates of recurrence-free intervals between the latter two groups would indicate that a less than optimal margin is as good as no margin at all. It may be conjectural but perhaps the similar recurrence-free intervals between the latter two groups could be explained by the ensuing three factors: (1) residual microscopic disease; (2) the result of spillage of tumour cells at the time of surgery; (3) the presence of discontinuous nests of tumour cells. McKee and associates [16] also stated that a surgical margin of

Whilst it is easy to understand and accept that a surgical margin of at least 10 mm is required to reduce the risk of local tumour recurrence, the problem with managing such cases is that usually the tumours have the clinical characteristics of benign lumps and hence a number of surgeons would inadvertently excise the lesions with surgical margins of less than 10 mm with the pre-operative assumption of benign lesions. This problem may be avoided by surgeons assuming that occasionally a lesion that was pre-operatively presumed to be benign could potentially be diagnosed post-operatively by histological examination as a malignant lesion therefore excision of all lesions assumed to be benign should be carried out with an excision margin of at least 10 mm. Such a suggestion
would perhaps be unlikely to be accepted by a number of surgeons in view of the fact that the subsequent finding of a malignant tumour in a presumed benign lesion is rare. It has been suggested that if a conservative excision of a scrotal lump has been made initially and a diagnosis of sarcoma is established unexpectedly, then a re-excision with a margin of 1 cm should be the next step [12]. Whilst this suggestion is easy to understand, re-excision with a surgical margin of more than 10 mm may not be easy to determine in view of the post-operative fibrosis at the site of the previous excision by the surgeons inability to clearly identify normal tissue in that case perhaps there is a place for frozen section histological examination to help the surgeon to determine that the new excision site is at least 10 mm away from tumour. Mondaini and associates [18] have shown in their studies that the best chance for a cure with this type of tumour resides with surgical excision. Rajkomar and associates [13] also stated that the role of radiotherapy has not been well defined. Bowden and Booher [19] stated that subcutaneous scrotal leiomyosarcomas are extremely rare. And malignant mesenchymal tumours such as leiomyosarcomas have a "pseudo-capsule" [1] Liae and associates [20] as well as Ballo and associates [21] suggested that there may be infiltration of tumour cells in the adjacent tissues; therefore it might prove difficult to precisely estimate the tumour margins. In the case of leiomyosarcomas of the spermatic cord due to the 'irregular' pattern of tumour growth and the anatomical constraints in the inguinal canal, wide circumferential resection margins may be difficult to achieve [20],[21]. Nevertheless, in view of leiomyosarcoma's high propensity for local recurrence if negative histological margins are not achieved during primary excision, re-excision should be considered mandatory, even if this should involve sacrificing some areas of normal anatomy. It has been stated that adjuvant radiotherapy has become an established method in the management of leiomyosarcomas of the spermatic cord [21], [14], [22], [23]. In view of the extreme rarity of leiomyosarcomas related to the scrotum it cannot be said categorically whether or not adjuvant radiotherapy would be mandatory taking into consideration the fact that leiomyosarcomas limited to the sub-scrotal areas are easily accessible to wide excision with removal of surrounding normal tissue in order to avoid local recurrence. Some authors recommend adjuvant radiotherapy only for high grade leiomyosarcomas or to those patients believed to be at a high risk of local recurrence. Nevertheless, in view of the tumours high propensity (about 50%) [4], [5], of local recurrence pursuant to surgery alone, [21], [24], there is increasing consensus that leiomyosarcomas of all grades and histology should receive adjuvant radiotherapy [21]. A number of studies have reported better results with combined modality treatment and the recurrence could be reduced to 10 – 20% by using adjuvant radiotherapy [21], [24],[25]. It has been emphasised that adjuvant radiotherapy should supplement rather than substitute radical excision and should be instituted only after complete clearance of the tumour is achieved surgically. The field of radiotherapy should include the inguinal canal, ipsilateral pelvic tissue [21] and scrotum in cases of leiomyosarcomas of cord [14]. There is no authoritative literature on the use of adjuvant radiotherapy in the case of localised pedunculated subcutaneous scrotal leiomyosarcoma to the knowledge of the authors. The role of pre-operative radiotherapy has not been clearly established and hence there is no evidence to suggest that radiotherapy prior to surgical excision of leiomyosarcoma reduces rate of recurrence or improves the over-all prognosis. At the moment there are no documented controlled studies that specifically address the role of adjuvant systemic chemotherapy in adult spermatic cord leiomyosarcomas [26], though they have a well-defined role in childhood rhabdomyosarcomas [27]. Adjuvant chemotherapy is currently not routinely used in the treatment of spermatic cord leiomyosarcomas although it has been suggested that it might have a role in abrogating the haematogenous metastatic potential in high grade sarcomas [21] and in patients with metastatic disease. Subcutaneous intrascrotal leiomyosarcomas are extremely rare and there is no documented controlled study on the use of chemotherapy pursuant to its surgical excision. The prognosis of patients with leiomyosarcomas is highly variable. Kyle in 1966 [28]suggested a probable five year survival of 10 to 15% but recently a five-year survival rate of 50 to 80% has been reported [22], perhaps or possibly reflecting the advances in diagnosis and management of these tumours. It has been suggested that the wide range in the five-year survival rate of leiomyosarcomas might be due to the variations in tumour grade and stage at the time of diagnosis as well as the diversity of therapies involved [29]. There is paucity of literature pertaining to subcutaneous scrotal leiomyosarcomas; even though it may be conjectural, it may be argued that in the case subcutaneous scrotal leiomyosarcomas, the position
of such lesions allows for complete excision of subcutaneous leiomyosarcomas with surrounding normal tissue does minimising the risk of local recurrence hence perhaps these tumours should have better prognosis in comparison with paratesticular / spermatic cord leiomyosarcomas; perhaps such sub-cutaneous tumours should be treated by wide local excision only and adjuvant radiotherapy and chemotherapy should be reserved for those sub-cutaneous tumours in which there is histological evidence of lympho-vascular permeation.

Our patient’s lump was considered by both the patient and clinicians pre-operatively to be consistent with an innocuous benign lump and the patient was discharged from further follow-up to remain under the care of his General Practitioner who should refer him back for review if there was any problem with the wound post operatively. However, he was called back for review upon receiving the histology report.

Conclusion

The experience gained in this case and literature review is sufficient for us to make the ensuing concluding statements:

-All surgically excised specimens should always be submitted for histological examination.

-The histology report of all surgically excised specimens that have been submitted for histological examination should be reviewed by the surgeon before finally discharging every patient even after day case surgery whether or not the specimens appear to be benign.

-On the whole surgical excision of a scrotal leiomyosarcoma with a clear margin of at least 10mm should be sufficient as initial treatment; if there is evidence of surgical margin involvement or a clear margin of less than 10 mm then re-excision would be required to provide a clear margin of at least 10 mm.

-Adjuvant therapy in the form of radiotherapy or chemotherapy should supplement radical excision with an acceptable tumour margin clear (at least 10 mm) and in general radiotherapy / chemotherapy alone should not be a substitute for radical surgical excision of a scrotal leiomyosarcoma.

-Because of the possibility of late local recurrence and distant metastases patients should be followed-up over a reasonably long period.

-There should be role for Chemotherapy / radiotherapy if there is evidence of lymphovascular permeation or distant metastasis.

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Illustrations

Illustration 1

Hematoxylin and Eosin staining X 10 Magnification Scrotal skin with underlying leiomyosarcoma

Illustration 2

Illustration 2 Hematoxylin and Eosin staining X 20 magnification illustrating leiomyosarcoma showing atypical spindle cells with mitotic figures
Illustration 3

Illustration 3 X 20 magnification showing positive immunohistological staining for SMA

Illustration 4

X 20 magnification Leiomyosarcoma showing positive immunohistological staining with Vimentin
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